National Guideline Alliance

Version 1.0

# End of life care for infants, children and young people: planning and management

## Full Guideline

*NICE Guideline*

*Methods, evidence and recommendations*

*1st July 2016*

*Draft for Consultation*

*Commissioned by the National Institute for Health and Care Excellence*

**Disclaimer**
Healthcare professionals are expected to take NICE clinical guidelines fully into account when exercising their clinical judgement. However, the guidance does not override the responsibility of healthcare professionals to make decisions appropriate to the circumstances of each patient, in consultation with the patient and/or their guardian or carer.



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# 1 Guideline summary

## 1.1 Guideline Committee membership, NGA staff and acknowledgements

### 1.1.1 Guideline Committee members

| Name | Role |
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| Karen Brombley | Nurse Consultant for Children and Young People's Palliative Care, Helen Douglas House Hospices<br>Honorary Nurse Consultant, Oxford University Hospital NHS Trust |
| Lucy Coombes | Caroline Menez Research Sister and Clinical Nurse Specialist, Children and Young People's Outreach and Symptom Care Team, Royal Marsden NHS Foundation Trust |
| Stacey Curzon (from November 2015) | Youth Worker, Rainbows Hospice for Children and Young People |
| Bobbie Farsides | Professor of Clinical and Biomedical Ethics, Brighton and Sussex Medical School |
| Jane Green | Patient and carer member |
| Satbir Jassal | General Practitioner, Bridge Street Medical Practice |
| Emily Harrop (Interim Chair from April 2016) | Consultant in Paediatric Palliative Care,<br>Helen & Douglas House Hospices<br>Honorary Consultant, Oxford University Hospital NHS Trust |
| Paul Nash | Senior Chaplain,<br>Birmingham Children's Hospital NHSFT and Co-convenor Paediatric Chaplaincy Network (GB&I) |
| Fauzia Paize (from May 2015) | Neonatologist, Liverpool Women's NHS Foundation Trust |
| Zoe Picton-Howell | Patient and carer member |
| David Vickers (Chair) | Consultant Paediatrician, Cambridgeshire Community Services NHS Trust |
| Amy Volans | Lead Clinical Psychologist and Systemic Family Psychotherapist, Diana Children's Community Palliative Care Team East London NHS Foundation Trust |
| Claire Wensley | Consultant Paediatrician, York Teaching Hospital NHS Foundation Trust |
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| Elissa Coster (until September 2015) | Children and Families Worker, The Brain Tumour Charity |
| Co-opted members | |
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| Name | Role |
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| | Young People |
| Afia Manaf | Pharmacist, Paediatric Critical Care, Royal Manchester Children's Hospital |
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### 1.1.2 National Guideline Alliance

| Name | Role |
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| Kate Coles (from December 2014) | Project Manager |
| Rishiraj Caleyachetty (December 2014 to March 2015) | Research Fellow |
| Katharina Dworzynski (from March 2015) | Senior Research Fellow and Guideline Lead |
| Yelan Guo | Interim Senior Research Fellow |
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| Jessica Sims (until December 2014) | Project Manager |
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#### 1.1.2.1 Acknowledgements

- Additional support was received from:
  - Development:
    - Ebenezer Ademisoye
    - Norin Ahmed
    - Caroline Cannon
    - Tina Chignoli
    - Taryn Krause
    - Ferruccio Pelone
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    - Dr Linda Maynard
    - Jane McHugh
  - Focus group report:
    - Together for Short Lives: Jane Aldridge, Lizzie Chambers, Johanna Taylor and all the researchers who helped develop this report

Thank you to the children and young people who took part in the Together for Short Lives report referred to in this guideline.

### 1.1.3 Dedication

In memory of Adam Bojelian and Callum David Miller, whose mothers ensured that their voices were heard during the development of this guideline.

## 1.2 What is a NICE clinical guideline?

National Institute for Health and Care Excellence (NICE) clinical guidelines are recommendations for the care of individuals in specific clinical conditions or circumstances within the NHS – from prevention and self-care through primary and secondary care to more specialised services. We base our clinical guidelines on the best available research evidence, with the aim of improving the quality of healthcare. We use predetermined and systematic methods to identify and evaluate the evidence relating to specific review questions.

NICE clinical guidelines can:

- provide recommendations for the treatment and care of people by healthcare professionals
- be used to develop standards to assess the clinical practice of individual healthcare professionals
- be used in the education and training of healthcare professionals
- help patients to make informed decisions
- improve communication between patients and healthcare professionals.

While guidelines assist the practice of healthcare professionals, they do not replace their knowledge and skills.

We produce our guidelines using the following steps:

- The guideline topic is referred to NICE from the Department of Health.
- Stakeholders register an interest in the guideline and are consulted throughout the development process.
- The scope is prepared by the National Collaborating Centre for Women and Children's Health (NCC-WCH).
- The NCC-WCH establishes a Guideline Committee.
- A draft guideline is produced after the group assesses the available evidence and makes recommendations.
- There is a consultation on the draft guideline.
- The final guideline is produced.

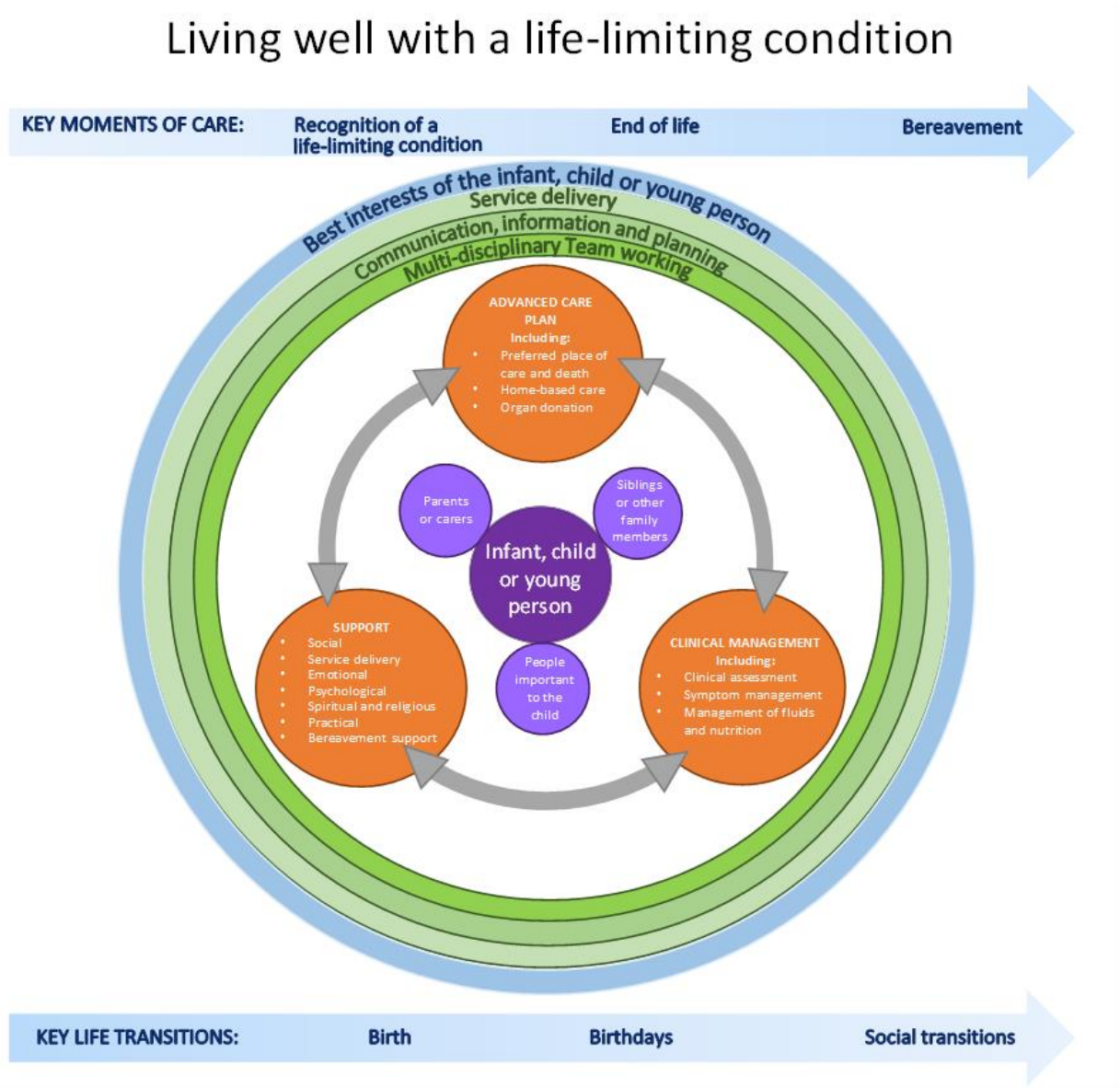
The NCC-WCH and NICE produce a number of versions of this guideline:

- The 'full guideline' contains all the recommendations, together with details of the methods used and the underpinning evidence.
- The 'short guideline' lists the recommendations, context and recommendations for research.
- 'Information for the public' is written using suitable language for people without specialist medical knowledge.
- NICE Pathways brings together all connected NICE guidance.

## 1.3 Supportive framework

The Committee agreed to present the content of the guideline graphically but was aware that the topic does not lend itself to an algorithm. Figure 1 represents a framework in which the guideline topics are placed to provide context.

**Figure 1: Supportive framework**



## 1.4 Recommendations

1. Be aware that most children and young people with life-limiting conditions and their parents or carers want to be fully informed about the condition and its management, and they value information that is:
    - specific to the child's or young person's individual circumstances
    - clearly explained and understandable
    - consistent
    - up-to-date
    - provided orally and in writing.
2. Be aware that some children and young people and parents or carers may be anxious about receiving information about their condition.
3. Ask how children and young people and their parents or carers would like to discuss the life-limiting condition. For example:
    - Ask which topics they feel are important and would particularly want information on
    - Ask whether there are topics they don't want detailed information on, and discuss their concerns
    - If appropriate ask parents or carers whether they think their child understands their condition and its management, and which professional their child would like to talk to about it.
    - If appropriate, ask parents or carers what they think their child should be told about their condition
    - Discuss with the child or young person and their parents or carers their right to confidentiality and how information about their condition will be shared
    - Review these issues with them regularly, because their feelings and need for information may change over time or if their circumstances change.
4. When talking to children and young people and their parents or carers:
    - be sensitive, honest and realistic
    - give reassurance when appropriate
    - discuss any uncertainties about the condition and treatment.
5. Be alert for signs or situations that the child or young person or their parents or carers need more information or discussions, for example if:
    - they are more anxious or concerned
    - the child or young person's condition deteriorates
    - a significant change to the treatment plan is needed.
6. Provide children and young people and their parents and carers with the information they need on:
    - their role and participation in Advance Care Planning (see 6.1)
    - the membership of their multidisciplinary team and the responsibilities of each professional (see 7.1)

- the care options available to them, including specific treatments, preferred place of care and place of death (see 6.2)
- any relevant resources or support available to them.

7. When difficult decisions must be made about end of life care, give children and young people and their parents or carers enough time and opportunities for discussions.
8. Think about how to provide information for children and young people with life-limiting conditions, taking into account their age and level of understanding. When appropriate, use formats such as:
    - one-to-one discussion
    - play, art and music activities
    - written materials and pictures
    - digital media, for example social media.
9. When deciding how best to communicate with the individual child or young person and their parents or carers, focus on their views and take account of:
    - their personal and family situation
    - their religious, spiritual and cultural beliefs and values
    - any special needs, such as communication aids or the need for interpreters.
10. Ask children and young people with life-limiting conditions and their parents or carers:
    - if there are other people important to them (such as friends, boyfriends or girlfriends, teachers, or foster parents) who they would like to be involved, and if so
    - how they would like those people to provide a supporting role.
11. Think about how best to communicate with each child or young person and their parents or carers:
    - when the life-limiting condition is first recognised
    - when reviewing the Advance Care Plan
    - if their condition worsens
    - when they are approaching the end of life.
12. Ensure that all parents or carers are given the information and opportunities for discussion that they need.
13. When deciding which healthcare professional should lead on communication at a particular stage in a child or young person's illness, take account of:
    - their expertise and ability to discuss the topics that are important at that time
    - their availability, for example if frequent discussions are needed during an acute illness or near the end of life
    - the views of the child or young person and their parents or carers.

14. When a life-limiting condition is first diagnosed, tell the child or young person (if appropriate) and their parents or carers about the condition and what it may mean for them.
15. Be aware of the importance of talking about dying, and if appropriate discuss with children and young people:
    - whether they want and are able to talk about dying
    - whether they or their parents or carers would like support in talking to each other about this.
16. When a child or young person is likely to die within hours or days, support them and their parents or carers by:
    - listening to any fears or anxieties they have and
    - showing empathy and compassion.
17. If a child or young person is likely to die within hours or days, explain to them and their parents or carers:
    - why you think this is likely, and any uncertainties
    - what clinical changes can be expected
    - whether you think the treatment plan should be changed.
18. Be aware that children and young people may have difficulty asking directly if they are going to die or are dying. Explore and discuss their concerns if you think they want to talk about this.
19. Be aware that parents or carers may have difficulty asking directly if a child or young person is dying. Explore and discuss their concerns if you think they want to talk about this.
20. Recognise that children and young people with life-limiting conditions and their parents or carers have a central role in decision-making and care planning.
21. Regularly ask children and young people and their parents or carers how they want to be involved in making decisions about their care, because this varies between individuals, at different times, and depending on what decisions are being made.
22. Explain to children and young people and to their parents or carers that their contribution to decisions about their care is very important, but that they do not have to make decisions alone and the multidisciplinary team will be involved as well.
23. Manage transition from children's to adult's services in line with the NICE guideline on transition from children's to adult's services.
24. Develop and record an Advance Care Plan for the current and future care of each child or young person with a life-limiting condition. The Advance Care Plan should include:
    - demographic information about the child or young person and their family
    - up-to-date contact information for:
        - the child or young person's parents or carers and
        - the key professionals involved in care
    - a statement about who has responsibility for giving consent
    - a summary of the life-limiting condition

- an agreed approach to communicating with and providing information to the child or young person and their parents or carers
- a statement covering what information about the child or young person and their parents or carers will be shared, and with whom
- an outline of the child or young person's life ambitions and wishes, for example on:
  - family and other relationships
  - social activities and participation
  - education
  - how to incorporate their religious, spiritual, and cultural beliefs and values into their care
- a record of significant discussions with the child or young person and their parents or carers
- agreed treatment plans and objectives
- education plans, if relevant
- a record of any discussions and decisions on
  - parallel planning of end of life care and medical care that is specifically for the underlying condition
  - the preferred place of care or place of death
  - organ and tissue donation (see 1.1)
  - management of life-threatening events, including plans for resuscitation or life support
  - specific wishes, for example on funeral arrangements and care of the body
- a distribution list for the Advance Care Plan.

25. Begin discussing an Advance Care Plan with parents during the pregnancy if there is an antenatal diagnosis of a life-limiting condition.
26. Develop and regularly review Advance Care Plans:
    - with relevant members of the multidisciplinary team and
    - in discussion with the child or young person and their parents or carers.
27. Advance Care Plans should take account of the child's or young person's life as a whole.
28. When developing the Advance Care Plan, take account of the beliefs and values of the child or young person and their parents or carers.
29. Explain to children and young people and their parents or carers that Advance Care Planning should:
    - help them be involved in planning their care and give them time to think about their views carefully
    - help them to understand the life-limiting condition and its management
    - ensure that relevant professionals can plan, develop and implement a management plan for now and the future

- help to prepare for possible future difficulties or complications
- support continuity of care, for example if there are changes in the professionals involved or in the care setting (such as a hospital admission or discharge).

30. Share the Advance Care Plan with the child or young person and their parents or carers, and with relevant professionals and services involved in their care, such as:
    - GPs
    - hospital consultants
    - hospices
    - respite centres
    - community nursing services
    - their school and other education services
    - ambulance services.
31. Update the advance care plan when needed, for example if:
    - new professionals become involved
    - the care setting changes (for example, hospital admission or discharge)
    - the child or young person and their parents or carers move home.

    Discuss the changes with the child or young person (if appropriate) and their parents or carers.
32. Share the Advance Care Plan with everyone involved each time it is updated.
33. When making an Advance Care Plan, discuss with the child or young person and their parents or carers:
    - the nature of their life-limiting condition, its likely consequences and its prognosis
    - the expected benefits and possible harms of the management options.
34. Be aware that all children and young people with life-limiting conditions should have an Advance Care Plan in their medical record, and that this should not be confused with a do-not-resuscitate plan.
35. Be aware that any existing resuscitation plan for a child or young person may need to be changed in some circumstances, for example if they are undergoing general anaesthesia.
36. Never assume that there is a do-not-resuscitate plan in place for a child or young person unless this is explicitly stated in their record.
37. Be aware that discussing the Advance Care Plan can be distressing for children and young people who are approaching the end of life and their parents or carers, and they may:
    - be reluctant to think about end of life care
    - have difficulties discussing end of life care with the professionals or with one another
    - have differences of opinion about the care plan.

38. When making or reviewing the Advance Care Plan for a child or young person approaching the end of life, talk to the parents or carers about the care and support they can expect when the child or young person dies. Discuss their personal needs and feelings about this.
39. When a child or young person is approaching the end of life, think about and discuss with them and their parents or carers their specific support needs. Review these needs regularly.
40. Discuss with children and young people with life-limiting conditions and their parents or carers where they would prefer to be cared for and where they would prefer to die.
41. Agree the preferred place of care and place of death with children and young people and their parents or carers, taking into account:
    - their wishes, which are personal and individual
    - their religious, spiritual and cultural values
    - the views of relevant and experienced healthcare professionals
    - safety and practicality.
42. If possible, services should ensure that children and young people can be cared for at their preferred place of care and die at their preferred place of death.
43. Explain that the place of care or place of death may change, for example:
    - if the child or young person and their parents or carers change their minds or
    - for clinical reasons or
    - due to problems with service provision.
44. Discuss with the child or young person and their parents or carers whether or not they are eligible to donate organs or tissue.
45. Involve the organ donation service if needed. If organ or tissue donation is not possible, explain why.
46. If the child or young person is eligible to donate organs or tissue, discuss this with them and their parents or carers when they are ready and as part of Advance Care Planning, and:
    - provide written information leaflets if needed
    - discuss how deciding to donate could affect their care, for example by changing their place of care and place of death
    - explain the practical policies and procedures involved.
47. If the child or young person does not have the capacity to decide about organ and tissue donation, ask their parents or carers to make the decision.
48. For further information on organ donation, including donor identification and consent, see the NICE guideline on organ donation for transplantation.
49. Children and young people with life-limiting conditions should be cared for by a defined multidisciplinary team.
50. As the child or young person's circumstances change (for example if they change from having care primarily to manage their condition to having end of life care), the membership of the multidisciplinary team should be adjusted accordingly.

51. Depending on the needs of the child or young person, the multidisciplinary team may include:
    - healthcare professionals from primary, secondary or tertiary services, including those with specialist expertise in palliative care
    - social care practitioners
    - education professionals
    - spiritual or religious advisors
    - hospice professionals.
52. Explain to children and young people and their parents or carers:
    - who the multidisciplinary team members are and how they are involved in their care.
    - how the multidisciplinary team membership will change if the care that is needed or the care setting changes.
53. Think about involving children and young people and their parents or carers in multidisciplinary team meetings (when appropriate).
54. Think about having a named individual from the multidisciplinary team to act as a first point of contact and coordinate care for the child or young person and their parents or carers.
55. For children and young people with life-limiting conditions who are approaching the end of life and are having home care, services should provide (when needed):
    - specialist medical advice at any time (day and night), for example telephone advice
    - paediatric nursing care at any time (day and night)
    - home visits by a healthcare professional with expertise in palliative care, for symptom management
    - practical support and equipment for interventions including oxygen, enteral nutrition, and subcutaneous and intravenous therapies
    - anticipatory prescribing for children and young people who are likely to develop symptoms.
56. Services should have agreed strategies and processes to support children and young people who are approaching the end of life and are having home care. These services should be based on established clinical networks, and should collaborate on care planning and service delivery.
57. If it is suspected that a child or young person may die soon and they are not in their preferred place of death, think about whether rapid transfer is possible and in their best interest. Discuss this with them and their parents or carers.
58. When planning rapid transfer to the preferred place of death, review and if necessary update the Advance Care Plan in discussion with the child or young person and their parents or carers and with the healthcare professionals who will be involved following the transfer. The updated Advance Care Plan should include a record of:
    - any intended changes to care and when they should happen
    - care plans that cover:

- the final hours or days of life
- what will happen if the child or young person lives longer than expected
- support for the family after the child or young person dies
- care of the child's or young person's body after death.

- the professionals who will be involved and their responsibilities
- the professionals who will help with the practical and administrative arrangements after the death.

59. When planning rapid transfer of a child or young person to their intended place of death:
    - be aware that the course of their condition may be unpredictable, and that they may die sooner or later than expected
    - discuss any uncertainties about the course of their condition and how this could affect their care with them and their parents or carers
    - ensure that relevant changes to the Advance Care Plan are implemented.
60. Think about using the rapid transfer service to allow the child or young person to be in their preferred place of death when withdrawing life-sustaining treatments, such as ventilation.
61. Before rapid transfer, agree with the parents or carers where the child's or young person's body will be cared for after their death.
62. In collaboration with local hospitals, hospices, and community, primary care and ambulance services, establish a rapid transfer service for children and young people with life limiting conditions to allow urgent transfer to the preferred place of death (for example from the intensive care unit to their home, or other locations [such as a children's hospice]).
63. When discussing possible places of care or places of death with children and young people and their parents or carers, provide information about:
    - the various care settings (for example home, hospice or hospital care)
    - the care and support available in each setting
    - practical and safety issues.
64. If the child or young person and their parents or carers prefer home care, take into account and discuss the practical considerations with them, such as the possible need for:
    - home adaptations
    - changes to living arrangements
    - equipment and support.
65. Services for children and young people who are approaching the end of life and are being cared for at home should be able to support parenteral drug administration (for example, continuous subcutaneous opioid or anticonvulsant infusions).
66. Be aware that children and young people with life-limiting conditions and their parents or carers may have:
    - emotional and psychological distress and crises

- relationship difficulties
- mental health problems.

67. Be aware that children and young people and their parents or carers may need support, and sometimes expert psychological intervention, to help with distress, coping, and building resilience.
68. Be aware that children and young people may experience rapid changes in their condition and so might need emergency interventions and urgent access to psychological services.
69. Be aware of the specific emotional and psychological difficulties that may affect children and young people who have learning difficulties or problems with communication.
70. Provide information to children and young people and their parents or carers about the emotional and psychological support available and how to access it.
71. Regularly discuss emotional and psychological wellbeing with children and young people and their parents or carers, particularly at times of change such as:
    - when the life-limiting condition is diagnosed
    - if their clinical condition deteriorates
    - if their personal circumstances change
    - if there are changes to their nursery care, school or college arrangements, or their employment
    - if there are changes to their clinical care, for example if their care changes focus from treating the condition to end of life care.
72. Be aware that continuity of care is important to children and young people and their parents or carers. If possible, avoid frequent changes to the healthcare professionals caring for them.
73. Be aware that children and young people with life-limiting conditions and their parents or carers have varied social and practical support needs, and that those needs may change during the course of their condition. This may include:
    - material support, for example housing or adaptations to their home, or equipment for home drug infusions
    - practical support, such as access to respite care
    - technical support, such as training and help with administration of drug infusions at home
    - education support, for example from hospital school services
    - financial support.
74. Discuss with parents or carers the practical arrangements that will be needed after the death of their child, and provide this information in writing. This should cover matters such as:
    - the care of the body
    - relevant legal considerations, including
        - the involvement of the child death overview panel
        - the involvement of the coroner

- registration of the death
- funeral arrangements
- post-mortem examination (if this is to be performed).

75. When a child or young person is approaching the end of life, discuss the bereavement support available with their parents or carers and provide them with written information.
76. When a child or young person is approaching the end of life, talk to their parents or carers about available psychological bereavement support groups.
77. Offer bereavement support to the parents or carers both before and after the death of a child or young person.
78. When planning bereavement support for parents or carers:
    - talk to them about the support that is available and explore with them what they would find helpful and acceptable
    - think about what support different professionals could provide, for example:
        - their GP
        - healthcare professionals who know the child or young person and are involved in their care
    - think about the role of individual healthcare professionals in providing specific aspects of support
    - inform the multidisciplinary team about the support plan.
79. When making a bereavement support plan with parents or carers, discuss possible options with them such as:
    - opportunities to talk to the professionals caring for the child or young person, to:
        - discuss memories and events
        - answer any concerns or questions they may have
    - home visits from the healthcare professionals caring for the child or young person
    - bereavement support groups.
80. Give professionals involved in the care of the child or young person opportunities to talk about and explore their thoughts and feelings:
    - when the child or young person is approaching the end of life and
    - after the child or young person has died.
81. Following the death of a child or young person, ensure that relevant healthcare and other professionals are informed in a timely manner.
82. Update relevant documents and databases after the death of a child or young person (to avoid, for example, clinical appointments being offered by mistake).
83. Ensure that healthcare professionals providing bereavement support have the necessary expertise.
84. In all discussions with children and young people and their parents or carers explore with them whether, based on their beliefs and values,

there are any aspects of care about which they have particular views or feelings.

85. Ask children and young people with life-limiting conditions and their parents or carers if they want to discuss the beliefs and values (for example religious, spiritual or cultural) that are important to them, and how these should influence their care. Be aware that they may need to discuss their beliefs and values more than once.

86. Take account of the beliefs and values of children and young people and of their parents and carers in all discussions with them and when making decisions about their care.

87. Be aware that:

- some children and young people and their parents or carers find discussions about their beliefs and values difficult or upsetting
- others find these discussions reassuring and helpful.

88. Be aware that children and young people may feel differently to their parents, carers, or healthcare professionals about how their beliefs and values should influence their care. If there is disagreement, try to make a mutually acceptable care plan, and if necessary involve the chaplaincy service or another facilitator.

89. When thinking about the possibility of treatment withdrawal for a child or young person who is approaching the end of life, take into account their beliefs and values and those of their parents or carers.

90. Take account of the beliefs and values of children and young people and their parents or carers when thinking about funeral arrangements and the care of the child or young person's body after death.

91. When a child or young person is approaching the end of life, discuss with their parents or carers what would help them, for example:

- important rituals
- recording or preserving memories (for example with photographs, hair locks or hand prints).

92. When assessing and managing pain, be aware that various factors can contribute to it, including:

- causative factors, for example musculoskeletal disorders or constipation
- environmental factors, such as an uncomfortable or noisy care setting
- psychological factors, such as anxiety and depression
- social, emotional, religious, spiritual or cultural considerations.

93. When assessing pain in children and young people:

- use an age-appropriate approach that takes account of their stage of development and ability to communicate
- try to identify what is causing or contributing to their pain, and be aware that this may not relate to the life-limiting condition
- take into account the following causes of pain and distress that might have been overlooked, particularly in children and young people who cannot communicate:
  - neuropathic pain (which can be associated with cancer)

- gastrointestinal pain (which can be associated with diarrhoea or constipation)
- bladder pain (which can be caused by urinary retention)
- bone pain (which can be associated with metabolic diseases)
- pressure ulcers
- headache (which can be caused by raised intracranial pressure)
- musculoskeletal pain (particularly if they have neurological disabilities)
- dental pain.

94. Be aware that pain, discomfort and distress may be caused by a combination of factors, which will need an individualised management approach.

95. For children and young people who have pain or have had it before, regularly reassess for its presence and severity even if they are not having treatment for it.

96. Think about non-pharmacological interventions for pain management, such as:

- Changes that may help them to relax, for example:
  - environmental adjustments (reducing noise)
  - music
  - physical contact such as touch, holding or massage
- local hot or cold applications to the site of pain
- comfort measures, such as sucrose for neonates.

97. When tailoring pain treatment for an individual child or young person, take into account their views and those of their parents or carers on:

- the benefits of pain treatment
- the following possible side effects of analgesia for moderate to severe pain (such as opioids):
  - unwanted sedation
  - reduced mobility
  - constipation.

98. Consider using a stepwise approach to analgesia in children and young people, based on pain severity and persistence:

- For mild pain, consider paracetamol or ibuprofen sequentially, and then in combination if needed.
- For moderate to severe pain, consider one of the following options:
  - paracetamol or ibuprofen sequentially, and then in combination if needed **or**
  - low-dose oral opioids (such as morphine), **or**
  - transmucosal opioids **or**
  - subcutaneous opioids **or**

- intravenously infused opioids (if a central venous catheter is in place).

99. If treatment with a specific opioid does not give adequate pain relief or if it causes unacceptable side effects, think about trying an alternative opioid preparation.

100. When using opioids, titrate treatment to find the minimal effective dose that will relieve and prevent pain.

101. Titrate treatment to provide continuous background analgesia, and prescribe additional doses for breakthrough pain if this occurs.

102. In addition to background analgesia, consider giving anticipatory doses of analgesia for children and young people who have pain at predictable times (for example when changing dressings, or when moving and handling). Do not include anticipatory doses when calculating the required daily background dose of analgesia.

103. Calculate opioid dosages for children and young people who are approaching the end of life using weight rather than age, because they may be underweight for their age.

104. If you suspect neuropathic pain and standard analgesia is not helping, consider a trial with other medicines, such as:

- gabapentin **or**
- a low-dose tricyclic antidepressant (for example amitriptyline ) **or**
- an anti-NMDA agent (for example ketamine or methadone), used under guidance from a specialist.

105. Be aware that as children and young people with life-limiting conditions approach the end of life they may:

- become agitated, shown by restlessness, irritability, aggressive behaviour, crying or other distress
- show signs of delirium, such as confusion, disrupted attention, disordered speech, hallucinations and agitation.

106. If a child or young person who is approaching the end of life becomes agitated or delirious, make sure that they are safe from physical injury.

107. If a child or young person becomes agitated as they are approaching the end of life, look for causes and factors that may be contributing to this, including:

- medical disorders and conditions such as pain, hypoxia, anaemia, dehydration, urinary retention or constipation
- psychological factors such as fear, anxiety or depression
- adverse effects from medication.

108. For children and young people with a neurological disability who are approaching the end of life, be aware that the symptoms and signs of agitation or delirium can be mistaken for the signs and symptoms of seizures or dystonia.

109. If a child or young person who is approaching the end of life needs treatment for agitation:

- identify and if possible treat any medical or psychological conditions that may be contributing to it
- think about non-pharmacological interventions, such as:

- calm speaking, reassurance, distraction, and physical contact such as holding and touch
- changes to the environment to make it more comfortable, calm and reassuring, to reduce noise and lighting, to maintain a comfortable room temperature, and to provide familiar objects and people and relaxing music
- religious and spiritual support if this is wanted and helpful

- think about pharmacological interventions (beginning with low doses and increasing if necessary). Drugs to think about using include:
  - benzodiazepines, such as midazolam, diazepam or lorazepam
  - neuroleptics, such as haloperidol or levomepromazine.

110. If a child or young person is approaching the end of life and has respiratory distress, breathlessness or noisy breathing, think about and if possible treat the likely causes or contributing factors. If it is likely to be caused by:
    - Anxiety:
      - discuss why they are anxious
      - reassure them and manage the anxiety accordingly
      - consider breathing techniques and guided imagery.
    - Physical discomfort – think about what could be causing the discomfort (for example their position) and help them with it if possible.
    - Environmental factors – think about environmental changes such as changing the temperature.
    - Accumulated airway secretions – think about repositioning, airway suctioning, physiotherapy or anti-secretory drugs.
    - Medical disorders (for example pneumonia, heart failure, sepsis or acidosis) – use appropriate interventions (according to their Advance Care Plan) such as:
      - anti-secretory agents
      - bronchodilators
      - nebulised saline
      - sedatives or anxiolytic agents
      - opioids
      - oxygen.
111. For children and young people who are approaching the end of life and have respiratory distress, breathlessness or noisy breathing that needs further assessment, consider referral to an appropriate specialist (for example, a respiratory or cardiac specialist).
112. If a child or young person is approaching the end of life and has respiratory distress, breathlessness or noisy breathing:
    - explain to them and to their parents or carers that these symptoms are common
    - discuss the likely causes or contributing factors

- discuss any treatments that may help.

113. If a child or young person is approaching the end of life and has a seizure, look for and if possible treat or remove any potential causes, triggers or contributing factors, for example:
    - fever
    - electrolyte disturbances
    - drug reactions
    - sleep deprivation
    - pain
    - excessive environmental stimulation.
114. If a child or young person is thought to be at increased risk of seizures, include seizure management in their Advance Care Plan. Think about the benefits and drawbacks of specific seizure treatments and:
    - take into account how any decisions could affect the choices available for place of care and place of death and
    - discuss this with the child or young person and their parents or carers.
115. For children and young people who are approaching the end of life, be aware that abnormal movements (such as dystonic spasms) might be mistaken for seizures. If in doubt seek specialist advice.
116. If a child or young person is approaching the end of life and is thought to be at increased risk of seizures (for example because they have had seizures before or because of an existing brain disorder), explain to them and their parents or carers:
    - how likely it is that they may have a seizure
    - what they might notice if a seizure happens
    - that seizures can be frightening or upsetting
    - what parents or carers should do if a seizure happens at home (for example, placing the child or young person in a safe position).
117. Ensure that parents or carers who have been provided with anticonvulsive therapy (such as buccal midazolam) know how and when to use it if the child or young person has a seizure at home.
118. If a child or young person with a life-limiting condition is approaching the end of life or is dying, discuss how to manage their fluid needs with them and their parents or carers.
119. If a child or young person is dying, encourage and support them to drink if they want to and are able.
120. If a child or young person is dying, continue to provide them with lip and mouth care.
121. If a child or young person is dying and cannot drink, discuss with them (as appropriate) and their parents or carers whether starting or continuing enteral tube or intravenous fluids is in their best interests.
122. Be aware that enteral tube and intravenous fluids may have a significant effect on care, may be a burden for children and young people, and may mean the place of care and place of death need to be changed.

123. If a child or young person is given enteral or intravenous fluids, review this decision regularly to make sure it continues to be in their best interests.

124. If a child or young person is approaching the end of life or is dying, discuss how to manage their nutritional needs with them and their parents or carers.

125. If a child or young person with a life-limiting condition is dying, encourage and support them to eat if they want to and are able.

126. If a child or young person is dying and they are receiving enteral tube feeding or intravenous nutrition:

- discuss with them (as appropriate) or their parents or carers whether continuing this is in their best interest and
- review this decision regularly.

127. For children and young people with life-limiting conditions, be aware that:

- there are various symptoms and signs (individually or in combination) that indicate they may be likely to die within hours or days **and**
- the wider clinical context is also relevant **and**
- there is often some uncertainty about this.

128. When assessing whether a child or young person is likely to die within hours or days, be aware that the following signs are common in the last hours or days of life, and monitor these non-invasively as far as possible:

- a change of breathing pattern (for example noisy, laboured or irregular breathing)
- impaired peripheral perfusion (which can be indicated by a pale or grey appearance, or a prolonged capillary refill time), including temperature instability
- loss of interest in or ability to tolerate drinks or food
- a marked and unexplained fall in urine output
- an altered level of awareness (for example reduced consciousness, alertness or responsiveness, excessive sleeping, or confusion)
- intractable seizures that keep occurring even with optimal management
- new onset of profound weakness
- increasing pain and need for analgesia.

129. When assessing symptoms and signs to decide whether a child or young person is likely to die within hours or days, take into account the wider clinical context, including:

- their normal clinical baseline
- past clinical events (such as previous episodes of temporary deterioration)
- the overall progression of their condition.

130. When assessing whether a child or young person is likely to die within hours or days, take into account the clinical judgement of healthcare professionals experienced in end of life care.

131. If the child or young person or their parents or carers feel that they are likely to die within hours or days:
    - be aware that they may be correct
    - discuss their concerns with them.
132. When a child or young person is likely to die within hours or days:
    - be aware that they or their parents or carers may not express their feelings openly, and may:
        - have intense and varied feelings such as fear, hopelessness or anger **or**
        - become more accepting of the inevitability of death
    - give them and their parents or carers opportunities to talk.
133. When children and young people become seriously ill and are likely to die within hours or days, provide care as specified in their Advance Care Plan and review if needed.
134. If a child or young person may be approaching the end of life and they or their parents or carers want to be involved in making decisions about their care, discuss and review their Advance Care Plan with them.
135. When a child or young person is approaching the end of life, discuss with them and their parents or carers and with relevant healthcare professionals:
    - any available invasive treatments that might be in their best interest
    - any interventions they are currently receiving that may no longer be in their best interest.
136. If withdrawing a treatment for a child or young person who is dying, explain to them and to their parents or carers that it is often difficult to tell if or how this may affect them, or when they will die.
137. When a child or young person is likely to die within hours or days, ensure that they can have private time with their parents or carers.

## 1.5 Key research recommendations

1. When planning and managing end of life care, what factors help children and young people with life-limiting conditions and their parents or carers to decide where they would like end of life care to be provided and where they prefer to die?
2. Do protocols for rapid transfer of children and young people with life-limiting conditions help ensure that they are able to die in their preferred place of death?
3. What emotional support do children and young people with life-limiting conditions and their parents or carers need, and how would they like these needs to be addressed?
4. What is the acceptability, safety, and effectiveness of different types of opioid analgesia for breakthrough pain in children and young people with life-limiting conditions who are having end of life care in the community?

5. What signs and symptoms indicate that a child or young person with a life-limiting condition is likely to die within hours or days?

## 1.6 Research recommendations

1. When planning and managing end of life care, what factors help children and young people with life-limiting conditions and their parents or carers to decide where they would like end of life care to be provided and where they prefer to die?
2. Do protocols for rapid transfer of children and young people with life-limiting conditions help ensure that they are able to die in their preferred place of death?
3. What is the effectiveness of a home-based package of care as opposed to hospital or hospice care?
4. What emotional support do children and young people with life-limiting conditions and their parents or carers need, and how would they like these needs to be addressed?
5. What are children's, young people's and their families' perceptions and attitudes about chaplaincy in paediatric end of life care and when would they like to access religious and spiritual support?
6. What is the acceptability, safety, and effectiveness of different types of opioid analgesia for breakthrough pain in children and young people with life-limiting conditions who are having end of life care in the community?
7. What is the acceptability, safety and effectiveness of oral / trans-mucosal opioids or benzodiazepines in the management of acute breathlessness in the context of end of life care?
8. What is the acceptability, safety and effectiveness of delivering different subcutaneous infusions of anti-epileptic medication during the out of hospital management of persistent seizures close to the end of life?
9. What signs and symptoms indicate that a child or young person with a life-limiting condition is likely to die within hours or days?

## 1.7 Other versions of the guideline

- The 'short guideline' lists the recommendations, context and recommendations for research.
- 'Information for the public' is written using suitable language for people without specialist medical knowledge.

## 1.8 Schedule for updating the guideline

For the most up-to-date information about guideline reviews, please see the latest version of the NICE guidelines manual available from the NICE website.

# 2 Introduction

**In this guideline:**

**'Children and young people' refers to everyone under 18 years old. This includes neonates and infants when applicable even when these groups are not specifically mentioned (as is the case in in the recommendations).**

**'Parents or carers' refers to the people with parental responsibility for a child or young person. If the child or young person or their parents or carers (as appropriate) wish, other family members or people important to them should also be given information and be involved in discussions about care.**

In modern Western Society, the death of a child is not expected by the family or carers, and thus has wide and devastating consequences. Society recognises orphans and widow(er)s, but there is no name given for those who have been bereaved of a child. Parents reasonably expect that they will die before their children, and fortunately the death of a child or young person is an uncommon event in the UK. There has been a particularly significant change in recent years, as the infant mortality rate in 2012 in England and Wales was the lowest ever recorded, at 4 deaths per 1000 live births, which can be partly explained by improvements in neonatal intensive care. As recently as 1982 the infant mortality rate was 10.8 deaths per 1000 live births.

Complications of pre-term birth, particularly respiratory and cardiovascular conditions, account for about half of infant deaths. Congenital anomalies account for about a further third.

The 2014 report, Why children die: death in children and young people in the UK, noted that, despite improving mortality rates, in 2012 more than 2,000 children and young people aged between 1 and 19 years died in England and Wales. For children and young people aged between 1 and 15 years, cancer, neurodevelopmental, respiratory, cardiovascular and congenital conditions (which tend to be chronic and progressive) account for about 60% of deaths. For young people aged over 15 years, external causes (such as accidents) are more common, accounting for 42% of deaths. The proportion of young people aged over 15 years who die from chronic conditions falls to about 30%, although cancer and neurodevelopmental conditions continue to be common causes of death in young people.

It is estimated that almost 50,000 children and young people aged 19 years or under in the UK (40,000 of these in England) are living with a life-limiting condition at any time, and may therefore need end of life care. They may have widely varying needs, as there are over 300 conditions that could be classed as life-limiting or life-threatening in this age group (Fraser 2012). Some of these children and young people also have severe disabilities and multiple complex health and care needs, in addition to end of life care needs. The importance of support for children and young people with life-limiting conditions is an area that these guidelines try to emphasise.

There is wide regional variation in paediatric end of life care practice, particularly in how services are delivered, combining a broad range of health and other care services, including hospitals, hospices, primary care and community professionals, ambulance services, dedicated palliative care teams, and other support providers. Specialist end of life care services for children may be delivered in a variety of settings. Consultant-led teams may be found within some children's hospices, as well as in tertiary children's hospitals, and also within community-based services. Hospices and community services offering a specialist service will often offer 'in-reach' to local hospitals, as well as supporting end of life care in hospice, schools and at home. Services thus span statutory and charitable sectors (for example hospices). Because of this, good communication, care coordination, and effective

networking are essential to providing good end of life care. Children and young people are likely to need different services at different stages of their illness and they will get the best care possible when services communicate with and support each other. Core end of life care skills exist in most local community teams, among children's community nurses and general paediatricians/general practitioners.

End of life care for adults is a well-established discipline, with evidence that if it starts early it can both enhance and even prolong life when facing a life-limiting illness (Temel, 2012). Paediatric end of life care generally lasts for a longer time frame and for a wider range of life-limiting conditions than for adults (Spathis, 2012). It begins when a life-limiting condition is diagnosed (potentially in the antenatal stage), and continues even if a child is having treatment for the underlying condition (WHO, 1998) and will, in the event of the death of a child of young person, continue to the immediate bereavement support of their family. Young people may continue to have end of life care after they turn 18 years, and it may remain part of the transition to adult care (see the NICE guidance: Transition from children's to adult's' services).

Children, young people and their parents, families or carers may have varied and differing ideas about what represents good end of life care. They may also have differences of opinion with each other and what is a priority for them, and at various stages, over time, their priorities may change.

This guideline covers the physical, emotional, social, and spiritual elements of end of life care, and focuses on improving the child or young person's quality of life and supporting their family and carers. There are, for instance, recommendations on managing distressing symptoms and providing care and bereavement support after death. Recommendations have also been made about how services should be delivered. The guideline is aimed at all providers of paediatric end of life care, whatever their level of practise, and also for children and young people with life-limiting conditions and their parents or carers.

The guideline covers children and young people with a life-limiting condition. It does not make recommendations for children or young people who die suddenly and unexpectedly (for example, accidental death).

## 2.1 For whom is this guideline intended

- All children and young people with life-limiting conditions (conditions that are expected to result in an early death, either for everyone with the condition or for a specific person)
- Families, carers and other people who are important to children and young people with life-limiting conditions
- Professionals who provide end of life care for children and young people
- Commissioners of end of life care services for children and young people.

## 2.2 Related NICE guidance

- Acutely ill patients in hospital (2007) NICE guideline CG50
- Antenatal and postnatal mental health: clinical management and service guidance (2014) NICE guideline CG192
- Antisocial behaviour and conduct disorders in children and young people: recognition and management (2013) NICE guideline CG158
- Attention deficit hyperactivity disorder: diagnosis and management (2008) NICE guideline CG72
- Autism in under 19s: recognition, referral and diagnosis (2011) NICE guideline CG128
- Autism in under 19s: support and management (2013) NICE guideline CG170
- Bipolar disorder: assessment and management (2014) NICE guideline CG185

- Borderline personality disorder: recognition and management (2009) NICE guideline CG78
- Challenging behaviour and learning disabilities: prevention and interventions for people with learning disabilities whose behaviour challenges (2015) NICE guideline NG11
- Common mental health problems: identification and pathways to care (2011) NICE guideline CG123
- Depression in adults: recognition and management (2016) NICE guideline CG90
- Depression in children and young people: identification and management (2015) NICE guideline CG28
- Eating disorders in over 8s: management (2004) NICE guidelines CG9
- Generalised anxiety disorder and panic disorder in adults: management (2011) NICE guideline CG113
- Improving outcomes in children and young people with cancer (2005) NICE cancer service guidance CSG7
- Improving supportive and palliative care for adults with cancer (2004) NICE cancer service guidance CSG4
- Neuropathic pain – pharmacological management (2013) NICE guideline CG173
- Opioids in palliative care (2012) NICE guideline CG140
- Organ donation for transplantation (2011) NICE guideline CG135
- Patient experience in adult NHS services (2012) NICE guidance CG138
- Pressure ulcers (2014) NICE guideline CG179
- Post-traumatic stress disorder: management (2005) NICE guidelines CG26
- Psychosis and schizophrenia in adults: prevention and management (2014) NICE guideline CG178
- Psychosis and schizophrenia in children and young people: recognition and management (2013) NICE guideline CG155
- Psychosis with substance misuse in over 14s: assessment and management (2011) NICE guideline CG120
- Self-harm in over 8s: long-term management (2011) NICE guideline CG133
- Self-harm in over 8s: short-term management and prevention of recurrence (2004) NICE guideline CG16
- Social anxiety disorder: recognition, assessment and treatment (2013) NICE guideline CG159

## 2.3 Remit

NICE received the remit for this guideline from the Department of Health and then commissioned the NGA to produce the guideline.

The Department of Health has asked NICE: ‘To prepare a clinical guideline on the End of life care for infants, children and young people’.

# 3 Guideline development methodology

This chapter describes the methods used to review the evidence and generate the recommendations presented in subsequent chapters. This guidance was developed in accordance with the methods outlined in the NICE guidelines manual 2012 for the scoping phase, and the NICE guidelines manual 2014 from the development phase.

## 3.1 Developing the review questions and protocols

Review questions were developed according to the type of question:

- intervention reviews – in a PICO framework (patient, intervention, comparison and outcome)
- reviews of diagnostic test accuracy – using population, index tests, reference standard, and target condition
- qualitative reviews – using population, area of interest and themes of interest
- prognostic reviews – using population, presence or absence of a risk factor, and outcome.

These frameworks guided the literature searching process, critical appraisal and synthesis of evidence and facilitated the development of recommendations by the Committee. The review questions were drafted by the NGA technical team, then refined and validated by the Committee. The questions were based on the key clinical areas identified in the scope (Appendix A).

A total of 20 review questions were identified (see Table 1).

Full literature searches, critical appraisals and evidence reviews were completed for all the specified review questions.

**Table 1: Description of review questions**

| Chapter or section number | Type of review | Review questions | Outcomes |
|---|---|---|---|
| 4 | Qualitative | What information and information type (written or verbal) is perceived as helpful and supportive by children and young people (if appropriate), and their family or carer before and after an infant, child or young person dies including managing practical arrangements, and care of the body? | Themes will be identified from the literature, for example:<br>• use of jargon and terminology<br>• uncertainty around likelihood of death<br>• methods of information provision (tools to facilitate). |
| 5 | Qualitative | What are the barriers and facilitators to effective communication between the child or young person, the family or carer and the healthcare professionals about the life-limiting condition and likelihood of imminent death? | Themes will be identified from the literature, for example:<br>• empathy and rapport (cultural and religious considerations)<br>• timing (when to initiate)<br>• resources (time spent with individuals and place of communication; that is, privacy in hospital)<br>• families' acceptance of prognosis. |
| 6.1 | Qualitative | What are the barriers and facilitators to the infant, child or | Themes will be identified from the literature, for example: |

| Chapter or section number | Type of review | Review questions | Outcomes |
|---|---|---|---|
| | | young person, the family or carer and the multidisciplinary team in being involved in decision-making to inform the development, assessment and reviews of personalised, parallel and Advance Care Planning (including if appropriate decisions about continuing or stopping life-sustaining treatment and attempting cardiopulmonary resuscitation)? | • timing of planning<br>• need for regular reviews<br>• assessments of needs<br>• professional roles<br>• cultural, religious and ethical differences. |
| 6.2 | Qualitative | What preferences do children and young people with a life-limiting condition and their family members or carers have for place of care and for place of death, and what determines those preferences? | Themes will be identified from the literature, for example:<br>• circumstances that facilitate or hinder availability of choices (personal, social, practical)<br>• characteristics of acceptable place for care or to die<br>• dynamic changes (trajectory of care). |
| 6.3 | Qualitative | What aspects of communication and information provision facilitate or hinder discussions between children and young people with a life-limiting illness and their family members or carers with healthcare professionals to make decisions on organ or tissue donation? | Themes will be identified from the literature, for example:<br>• bereavement experience (consolation)<br>• altruism<br>• organ and tissue donation as part of the care plan<br>• religious or spiritual beliefs. |
| 7.3 | Intervention | What services have to be in place to make rapid transfer available to take infants, children and young people with a life-limiting illness to their preferred place of care in their last days of life as part of service delivery?<br>*Note*. As an integrated part of the rapid transfer programme, particular consideration will be given to infants, children and young people (ICYP) who need compassionate extubation (including all life-sustaining treatment, for example non-invasive ventilation) in the preferred place (what services should be in place to facilitate) | Outcomes will include:<br>• Quality of life of the child or young person or/and their families/carers – for example, pain of the ICYP, release of distressing symptoms of the ICYP, and anxiety of the ICYP and their parents or carers.<br>• Quality of death.<br>• Successful transfer to preferred place of care/death (fulfilment of the transfer plan).<br>• Satisfaction of the child or young person and their families/carers with the care.<br>• Time taken to achieve transfer<br>• Unexpected hospital re-admission.<br>• Access of parents or carers to the patient in both settings. |
| 7.2 | Intervention | What is the effectiveness of day and night specialist telephone healthcare professional support (or parents/carers support), day and | Outcomes will include:<br>• Satisfaction with the care on the part of the child or young |

| Chapter or section number | Type of review | Review questions | Outcomes |
|---|---|---|---|
| | | night community nursing support, and the combination of the 2 for the needs of infants, children and young people with life-limiting conditions, and for the needs of their family members and carers during this time and after death as part of service delivery? | person and/or their families/carers<br>• Change in health resources utilisation (for example, reduction in unintended hospital re-admission rates, reduction of hospitalisation, reduction in length of hospital stay).<br>• Change in level of distressing symptoms such as pain, agitation.<br>• Change in home visits by nurses (mainly relevant to day and night specialist advice support). |
| 7.1 | Intervention | What is the clinical and cost effectiveness of a defined multi-disciplinary team (MDT) of a particular composition compared with one of a different composition and compared with care without a defined MDT? | Outcomes will include:<br>• Prevention of unplanned hospital admissions.<br>• Discharge time.<br>• Quality of life of the child, young person.<br>• Quality of life of the parent, carer.<br>• Satisfaction of the child or young person.<br>• Satisfaction of the parent or carer with the CYP's care (for example, level of care and improved communication).<br>• Control of symptoms (pain, dyspnoea, nausea/vomiting). |
| 7.4 | Intervention | What is the clinical and cost effectiveness of a home-based programme of care compared with care in other settings? | Outcomes will include:<br>• Unplanned/precipitous admission to hospital.<br>• Family or care giver stress and distress.<br>• ICYP satisfaction/comfort.<br>• Parent/carer satisfaction/comfort.<br>• Control of symptoms (pain, dyspnoea, nausea/vomiting).<br>• Health related quality of life (levels of comfort, lack of distress). |
| 8.1 | Mixed - intervention and qualitative | Are psychological interventions effective for infants, children and young people with life-limiting conditions and what factors influence the attitudes of children and young people and the family's involvement and decisions about | Quantitative outcomes:<br>• Psychological well-being of ICYP (for example resilience, depression, fear, anxiety, mood change).<br>• Quality of life of ICYP.<br>• Satisfaction of ICYP. |

| Chapter or section number | Type of review | Review questions | Outcomes |
|---|---|---|---|
| | | choices of those interventions? | • Pain- and child illness-related symptoms.<br>• Distressing symptoms (restlessness, agitation).<br><br>For qualitative outcomes, themes will be identified from the literature, for example:<br>• Unmet needs.<br>• Individual attitudes towards therapies based on for instance cultural differences.<br>• The skill and experience of therapists. |
| 8.1 | Mixed – intervention and qualitative | Are psychological interventions (including short-term bereavement therapies) effective for family members and carers of infants, children and young people and what factors influences their attitudes about those interventions before and after the death of an infant, child or young person with a life-limiting condition? | Quantitative outcomes:<br>• Psychological well-being (for example resilience, depression, fear, anxiety, mood change) of parents, families and carers before and after the ICYP's death.<br>• Quality of life of parents, families and carers before and after the ICYP's death.<br>• Satisfaction of parents, families and carers before and after the ICYP's death.<br>• Coping of parents, families and carers before and after the ICYP's death.<br>• Activities of daily living and parenting.<br>• Family function before and after the ICYP's death.<br><br>For the qualitative review, themes will be identified from the literature, for example:<br>• Bereavement of parents, families and carers after the ICYP's death<br>• Individual attitudes towards therapies based on, for example, cultural differences.<br>• Unmet needs. |
| 8.2 | Mixed - intervention and qualitative | What factors of social and practical support (including care of the body) are effective in end of life care of infants, children and young people with life-limiting conditions and their family members or carers and what influences attitudes about these before and after death? | Quantitative outcomes:<br>• ICYP well-being, including psychological well-being, common mental disorder or death distress, coping.<br>• The coping of parents or carers. |

| Chapter or section number | Type of review | Review questions | Outcomes |
|---|---|---|---|
| | | | • ICYP quality of life.<br>• Parents and carers' quality of life.<br>• Family functioning.<br>• ICYP health service use.<br><br>For the qualitative outcomes, themes will be identified from the literature, for example:<br>• Family functioning.<br>• ICYP health service use.<br>• Financial stress.<br>• Provision of equipment.<br>• Time spent on caregiving activities. |
| 8.3 | Mixed - intervention and qualitative | What factors of spiritual or religious support (including care of the body) are effective in end of life care of infants, children and young people with life-limiting conditions and their family members or carers and what influences attitudes about these before and after death? | Quantitative outcomes:<br>• ICYP well-being, including psychological well-being, common mental disorder or death distress.<br>• ICYP physical symptoms, such as pain, fatigue, hypersomnia and breathlessness.<br>• ICYP quality of life.<br>• Quality of life of parents, families and carers.<br>• ICYP health service use.<br>• ICYP satisfaction.<br>• Parents' or carers' satisfaction.<br><br>For the qualitative outcomes, themes will be identified from the literature, for example:<br>• Relationship with self and others<br>• Relationship with nature and music<br>• Hope<br>• Meaning and purpose in life/ meaning making. |
| 9.2 | Intervention | What pharmacological and non-pharmacological (excluding psychological) interventions are effective for the management of pain in ICYP with a life-limiting condition? | • Pain (measured by a validated scale, such as FLACC, NIPS).<br>• ICYP levels of distress.<br>• Parent, family and carer levels of distress.<br>• Adverse events, particularly opioid related, such as: |

| Chapter or section number | Type of review | Review questions | Outcomes |
|---|---|---|---|
| | | | ○ constipation<br>○ nausea / vomiting<br>○ itching<br>○ urinary retention<br>○ fatigue<br>○ confusion<br>○ respiratory depression<br>○ unwanted levels of sedation.<br>• Quality of life for ICYP and their parents, families and carers (using validated instruments, such as PedQL).<br>• Control of other distressing symptoms (including agitation and breathlessness).<br>• Proportion of ICYP taken home/re-admission to hospital/admission to hospice. |
| 9.3 | Intervention | What pharmacological and non-pharmacological (excluding psychological) interventions are effective for the management of agitation in ICYP with a life-limiting condition? | • Reduction of agitation.<br>• ICYP's levels of distress alleviated.<br>• Family or carers' levels of distress alleviated.<br>• ICYP's (health-related) quality of life.<br>• Family or carers' quality of life.<br>• ICYP satisfaction.<br>• Family or caregiver satisfaction (also retrospective)<br>• Adverse effects. |
| 9.4 | Intervention | What pharmacological and non-pharmacological (excluding psychological) interventions are effective for the management of respiratory distress in ICYP with a life-limiting condition? | • Objective and subjective signs of respiratory distress alleviated.<br>• ICYP levels of distress alleviated.<br>• Parent, family or carer levels of distress alleviated.<br>• ICYP (health-related) quality of life.<br>• Parent, family or carer quality of life.<br>• ICYP satisfaction.<br>• Parent, family or carer satisfaction (also retrospective).<br>• The number of different types of interventions (including varying doses and types of anticholinergics) needed to |

| Chapter or section number | Type of review | Review questions | Outcomes |
|---|---|---|---|
| | | | change noise intensity.<br>• Adverse effects. |
| 9.5 | Intervention | What pharmacological and non-pharmacological (excluding psychological) interventions are effective for the management of seizures in ICYP with a life-limiting condition? | • Reduction of seizures.<br>• ICYP levels of distress alleviated.<br>• Parent, family or carer levels of distress alleviated.<br>• ICYP (health-related) quality of life.<br>• Parent, family or carer quality of life<br>• ICYP satisfaction.<br>• Parent, family or carer satisfaction (also retrospective)<br>• Adverse effects. |
| 10.1 | Intervention | What is the effectiveness of medically assisted hydration in infants, children and young people during end of life care? | • Comfort or distress of the ICYP (or relevant proxy outcomes).<br>• Satisfaction of parents, family or carers.<br>• Adverse events including vomiting, respiratory distress, abdominal pain. |
| 10.2 | Intervention | What is the effectiveness of medically assisted nutrition in infants, children and young people during end of life care? | • Comfort or distress of the ICYP (or relevant proxy outcomes).<br>• Satisfaction of parents, family or carers.<br>• Adverse events including vomiting, respiratory distress, abdominal pain. |
| 11 | Mixed – prognostic, diagnostic and qualitative | What signs and symptoms, individually or in combination, help to recognise the infants, children or young people are likely to be in the last days of life and which of them are considered most informative by healthcare professionals? | For the quantitative outcomes:<br>• For diagnostic information:<br>○ sensitivity<br>○ specificity<br>○ positive predictive value<br>○ negative predictive values<br>○ positive likelihood ratios<br>○ negative likelihood ratios.<br>• If thresholds are established/pre-defined or for prognostic information:<br>○ relative risk (RR) or odds ratio (OR) (and ultimately risk difference) for patient outcomes listed above for those in higher or lower risk groups.<br>For the qualitative outcomes:<br>• Healthcare professionals' |

| Chapter or section number | Type of review | Review questions | Outcomes |
|---|---|---|---|
| | | | views on which signs and symptoms, prognostic tools, scores or indices, and laboratory or biological information are most useful. |

## 3.2 Searching for evidence

### 3.2.1 Clinical literature search

During the scoping stage, a search was conducted for guidelines and reports available on the websites of organisations which were relevant to the topic, and all references suggested by stakeholders during the scope consultation were considered for inclusion.

Systematic literature searches were undertaken to identify all published clinical evidence relevant to the review questions.

Databases were searched using relevant medical subject headings and free-text terms. Due to the large number of life-limiting conditions, it was considered appropriate to search primarily using terms related to end of life care. Where possible, searches were restricted to retrieve only English-language articles. Where appropriate, study type filters were applied. All searches were conducted in MEDLINE, Embase and The Cochrane Library. Where appropriate, certain searches were also conducted in PsycINFO, CINAHL or AMED. All searches were updated on 10 April 2016. Studies added to the databases after this date (even if they were published prior to this date) were not included unless specifically stated in the text.

Search strategies were quality-assured by cross-checking reference lists of key studies, analysing search strategies from other systematic reviews, and asking the Committee members to identify key studies. All search strategies were also quality-assured by a second Information Scientist working at the NGA, who had not created the strategies. Details of the searches, including study filters that were applied and databases that were used, can be found in Appendix E.

Grey and unpublished literature were not included in the searches, and searches for electronic, ahead-of-print publications were not routinely undertaken unless a particular study was identified by the Guideline Committee. Studies published in languages other than English were not reviewed.

### 3.2.2 Health economic literature search

A systematic literature search was undertaken to identify health economic evidence relevant to any review question. The evidence was identified by conducting a broad search relating to end of life care in the NHS Economic Evaluation Database (NHS EED) and the Health Technology Assessment (HTA) database with no date restrictions. Additionally, the same broad search was run on Medline, the Cochrane Central Register of Controlled Trials (CCTR) and Embase, with an economic filter applied. Where possible, searches were restricted to articles published in English and studies published in languages other than English were not reviewed. The titles and abstracts of records retrieved by the broad search were sifted for relevance, and full-text copies of potentially relevant publications were obtained. These were assessed using the inclusion criteria specified in the protocol for each review question. The search strategies for the health economic literature search are included in Appendix F. All searches were updated on 10 April 2016. Any studies added to the

databases after this date (even those published prior to this date) were not included unless specifically stated in the text.

## 3.3 Reviewing and synthesising the evidence

The evidence was reviewed following the steps shown schematically in Figure 2.

- Potentially relevant studies were identified for each review question from the relevant search results by reviewing titles and abstracts. Full papers were then obtained.
- Full papers were reviewed against pre-specified inclusion and exclusion criteria to identify studies that addressed the review question in the appropriate population, as outlined in the review protocols (review protocols are included in Appendix D).
- Relevant studies were critically appraised using the appropriate checklist as specified in the NICE guidelines manual (NICE, 2014).
- Key information was extracted on the study's methods, according to the factors specified in the protocols and results. These were presented in summary tables (in each review chapter) and evidence tables (in Appendix G).
- Summaries of evidence were generated by outcome (included in the relevant review chapters) and were presented in Committee meetings(details of how the evidence was appraised is described in section 3.3.4 below): :
    - Randomised studies: meta-analysis was carried out where appropriate and results were reported in GRADE profiles (for intervention reviews).
    - Observational studies: data were presented as a range of values in GRADE profiles.
    - Prognostic studies: data was presented as a range of values, usually in terms of the relative effect as reported by the authors.
    - Diagnostic studies: data were presented as measures of diagnostic test accuracy (sensitivity, specificity, positive and negative predictive value).
    - Qualitative studies: each study was summarised by theme and meta-synthesis was carried out where appropriate to identify an overarching framework of themes and subthemes.

For quality assurance of study identification, either whole study selections or a sample of the study selection results were double checked by a second reviewer as follows: service delivery (whole search for both rapid transfer and 24/7 service delivery), psychological interventions (for children and for adults 10% of the search) and pain and agitation symptom management (all 10%). A sample of all evidence tables was also quality assured and all write-ups of reviews were checked by a second reviewer. Any discrepancies were resolved by discussion between the 2 reviewers.

**Figure 2: Step-by-step review of evidence in the guideline**


### 3.3.1 Inclusion and exclusion criteria

The Committee was consulted about any uncertainty regarding inclusion or exclusion. The inclusion and exclusion of studies was based on the review protocols, which can be found in Appendix D. Excluded studies by review question (with the reasons for their exclusion) are listed in Appendix H.

In addition to the review protocols, there were particular inclusion and exclusion criteria which have been highlighted here for the following areas of the scope:

#### Guideline population

The guideline population was defined as children and young people with a life-limiting condition. As stated in the introduction, there are over 300 conditions that can be classed as life-limiting or life-threatening. Because of this high number of conditions, it was not possible to use all conditions as search terms. However, the focus of the guideline is on end of life care rather than on the specific treatment of each condition and therefore terms related to end of life and palliative care were used to identify the guideline population (see Appendix E). In the absence of evidence in the population of interest, it was discussed with the Committee whether indirect evidence would be relevant. In some instances evidence was identified that included a mixed population (for example, children likely to die from acute rather than life-limiting conditions, or from studies with children and young people up to the age of 21 rather than 18 as long as the average age and standard deviation was at a lower end). Evidence

from mixed populations was included in the following topics: planning; religious, spiritual and cultural support needs.

**Recognising the signs and symptoms of dying (mixed methods review)**

Another noteworthy inclusion in the qualitative review on the topic of 'signs and symptoms of dying' (chapter 11) was that Delphi consensus studies were also deemed acceptable for this topic (even though not strictly speaking qualitative in design). This was included to add other applicable expert consensus to the consensus of the Committee. A larger group of people (those in the Delphi panel as well as the Committee) agreeing on signs and symptoms would provide more weight to the selected signs and symptoms and therefore add robustness to the recommendations.

Furthermore, in the quantitative section of this review we aimed to identify pre-specified signs and symptoms that were independently related to recognising that a child or young person is in the last days of life; that is, independent of other characteristics. Therefore, the focus of the evidence was on studies using multivariable analysis.

**Other qualitative reviews (information, communication, planning, organ and tissue donation, social/practical support, spiritual/religious support, psychological interventions)**

Delphi and other descriptive surveys (such frequency of people who responding to closed-ended questions) were not included in the other qualitative reviews, for which rich qualitative data such as studies using interviews, focus groups, or surveys with open-ended options were considered most appropriate. For these reviews, if sufficient applicable evidence (in way of context and setting) was available, this was preferred over and above other possible included studies. This was the case in the review of communication and information provision where we looked for evidence from different perspectives on the barriers and facilitators that they were encountering; that is, the child or young person with life-limiting condition, their parents or carers and healthcare professionals. There was a large evidence base for these topics, therefore the evidence was restricted to the most applicable studies. This took into account to cover the issue from different perspectives of, for example, parents, healthcare professionals or children and young people. This issue is re-visited in section 3.3.2 on combining evidence from qualitative studies.

**Qualitative review for preferred place of care and preferred place of death**

Quantitative survey data was included in the preferred place of care and preferred place of death (see section 6.2) review. The Committee wanted to assess the percentage of people with a particular preference as well as their attitudes and reasoning about why they made this choice.

**Intervention reviews (for example, symptom management)**

Randomised trials, non-randomised trials, and observational studies were included in the evidence reviews as appropriate. For the intervention studies, both randomised and non-randomised comparative studies were included because the evidence base of randomised controlled trials in this particular study population was low.

**Other general study type inclusions / exclusions**

Conference abstracts were not automatically excluded from the review but were initially assessed against the inclusion criteria and then considered for inclusion only if no other full publication was available for that review question, in which case the authors of the selected abstracts were contacted for further information. None of the reviews included evidence from conference abstracts.

Literature reviews, posters, letters, editorials, comment articles, unpublished studies and studies not in English were excluded.

The review protocols are presented in Appendix D.

### 3.3.2 Methods of combining clinical studies

When planning reviews (protocols) the following approaches for data synthesis were discussed and agreed with Committee. However, insufficient evidence was identified to pool data for intervention reviews (and no evidence at all for prognostic or diagnostic components of the 'recognition of dying' protocol).

#### 3.3.2.1 Data synthesis for intervention reviews

It was planned to conduct meta-analyses where possible to combine the results of studies for each review question using Cochrane Review Manager (RevMan5) software.

Fixed-effects (Mantel-Haenszel) techniques were used to calculate risk ratios (relative risk) for binary outcomes, such as rate of adverse events or rate of people with symptom improvements.

For continuous outcomes, measures of central tendency (mean) and variation (standard deviation) would be required for meta-analysis. Data for continuous outcomes (such as number of episodes of vomiting) were planned to be analysed using an inverse variance method for pooling weighted mean differences and, where the studies had different scales, standardised mean differences were used. A generic inverse variance option in RevMan5 is used if any studies reported solely the summary statistics and 95% confidence interval (95% CI) or standard error; this included any hazard ratios reported. However, in cases where standard deviations were not reported per intervention group, the standard error (SE) for the mean difference is calculated from other reported statistics (p values or 95% CIs); meta-analysis was then undertaken for the mean difference and SE using the generic inverse variance method in RevMan5. When the only evidence was based on studies that summarise results by presenting medians (and interquartile ranges), or only p values were given, this information was assessed in terms of the study's sample size and was included in the GRADE tables without calculating the relative or absolute effects. Consequently, aspects of quality assessment such as imprecision of effect could not be assessed for evidence of this type. However, the limited reporting of this outcome was classified as a risk of bias in study limitations.

Stratified analyses were predefined for some review questions at the protocol stage when the Committee identified that these strata are different in terms of biological and clinical characteristics and the interventions were expected to have a different effect.

Statistical heterogeneity was assessed by visually examining the forest plots, and by considering the chi-squared test for significance at $p<0.1$ or an I-squared inconsistency statistic (with an I-squared value of more than 50% indicating considerable heterogeneity). Where considerable heterogeneity was present, we carried out predefined subgroup analyses. For instance in the pharmacological management of distressing symptoms, causes leading to the symptom would be a subgroup. The guideline group also considered that, for instance, route of administration, delivery system, and drug class could also be possible reasons for heterogeneity in results. In case of unexplained heterogeneity sensitivity analysis was planned to be carried out based on the quality of studies eliminating studies at overall high risk of bias (randomisation, allocation concealment and blinding, missing outcome data).

Assessments of potential differences in effect between subgroups were based on the chi-squared tests for heterogeneity statistics between subgroups. If no sensitivity analysis was found to completely resolve statistical heterogeneity then a random-effects (DerSimonian and Laird) model was employed to provide a more conservative estimate of the effect.

#### 3.3.2.2 Data synthesis for prognostic factor reviews

Signs and symptoms that indicate a child or young person is likely to die hours or days could be construed as a characteristic that predicts death occurring. This would be classified as a prognostic/predictive factor. In this respect odds ratios (ORs), risk ratios (RRs) or hazard ratios (HRs), with their 95% confidence intervals (95% CIs) for the effect of the prespecified prognostic factors were extracted from the papers when reported. Evidence came from observational studies because signs and symptoms that may indicate that someone is in the last days of life are not factors that could ever be randomised. For this topic, we looked for studies that took into account possible key confounders as reported in multivariable analyses. The reported measures were therefore adjusted to take into account other characteristics less likely to be actual signs and symptoms of being in the last days of life. Studies did this in a pre-specified manner or used statistical methods that included variables that were likely signs and symptoms related to dying and modelled them using statistical methods (such as multivariable logistic regressions) which then indicated which characteristics were the most likely independent prognostic factors rather than a factor only spuriously related.

#### 3.3.2.3 Data synthesis for diagnostic test accuracy reviews

**Data and outcomes**

Recognising dying could also be considered as being like a diagnostic process in which the child or young person either displays recognised signs or does not. Following death, children or young people can be identified as having had the sign or not. We therefore anticipated that studies would report there having been a particular sign, that could be assessed by a value, above or below a threshold value (for instance they might have had tests for a continuously measured characteristic, such as kidney function tests for renal signs and symptoms).

There are a number of diagnostic test accuracy measures. The area under the curve (AUC) of receiver operating characteristics (ROC) shows true positive rate (sensitivity) as a function of false positive rate (1 minus specificity). Sensitivity, specificity, positive and negative predictive values, and positive and negative likelihood ratios were reported.

The threshold of a diagnostic test is defined as the value at which the test can best differentiate between those with and without the target condition (for instance, a particular serum creatinine value) and, in practice, it varies among studies. For this particular question (recognising that a child or young person may be dying) specificity was regarded as particularly important. When specificity is high, a positive test rules in the diagnosis and when sensitivity is high, a negative test rules out the diagnosis – researchers have created the mnemonic SoPin/SnNout[a] for this (Sackett 1992). In other words in the case of high specificity with low sensitivity someone who has this sign/symptom (that is, akin to testing positive) would be likely to die within the next few days whereas for those who do not have the sign/symptom (akin to having a negative test) we are uncertain about when they may die. Sensitivity (ruling out), however, was also recognised as being important in order not to miss people who may be dying in the next few days.

a If a sensitive (Sn) test is negative (N), rule the diagnosis ‘out’; if a specific (Sp) test is positive (P), rule the diagnosis ‘in’.

**Data synthesis**

Diagnostic paired sensitivity-specificity forest plots were produced for each sign/symptom, using RevMan5. In order to do this, 2×2 tables (the number of true positives, false positives, true negatives and false negatives) were extracted.

Area under the ROC curve (AUC) data for continuous test results (such as serum creatinine for instance as a proxy for a sign of kidney function or failure) were given as AUC values with 95% confidence intervals. The Committee agreed on the following criteria for AUC:

- ≤0.50: worse than chance
- 0.50–0.60: very poor
- 0.61–0.70: poor
- 0.71–0.80: moderate
- 0.81–0.92: good
- 0.91–1.00: excellent or perfect test.

#### 3.3.2.4 Data synthesis for qualitative reviews

Where possible a meta-synthesis was conducted to combine qualitative study results. The main aim of the synthesis of qualitative data was to produce a description of the topics that may influence the experience of the person who is dying, those people important to them and healthcare professionals involved in their care, rather than build new theories or reconceptualise the topic under review. Whenever studies identified a qualitative theme, this was extracted and the main characteristics were summarised. When all themes were extracted from studies, common concepts were categorised and tabulated. This included information on how many studies had contributed to an identified overarching theme.

In qualitative synthesis the more a theme is reported by different studies does not necessarily mean that it would be more important than other themes. The aim of qualitative research is to identify new perspectives on a particular topic. Study type and population in qualitative research can differ widely, meaning that themes identified by just 1 or a few studies can provide important new information for a given topic. Therefore, for the purpose of the qualitative reviews in this guideline, we did not add further studies when they reported the same themes that had already been identified from the same perspectives (i.e. children or young people, parents or carers, or healthcare professionals) because the emphasis was on conceptual robustness rather than the quantitative completeness of evidence. This has implications for the types and numbers of studies that are included in the qualitative reviews. Study inclusion continued until no new relevant data could be found regarding a topic that would add to or refute it, a concept referred to in the literature as ‘theoretical saturation’ (Dixon-Wood 2005).

The most relevant evidence in this respect would originate from studies set in the target context of the UK NHS setting. Therefore when the evidence base was particularly large, we were able to focus first on studies in the most relevant context, but widened the study inclusion criteria when important perspectives were either not covered or were insufficiently covered. The final selection of included or excluded studies from those identified in the literature search was carried out by at least 2 researchers. Themes from individual studies were then integrated into a wider context and, when possible, overarching categories of themes with sub-themes were identified. Themes were derived from data presented in individual studies based directly on quotes from interviewees. When themes were extracted, theme names derived from the studies that provided it, such as ‘ready to die and go to heaven’, to take into account the influence of religious beliefs on care planning (see 6.1.5). The names of overarching themes, however, were named by the systematic reviewers, for instance ‘interpersonal/interactive communication’ (see 5.5.1).

Emerging themes were then placed into a thematic map that would present the relationship between themes and subthemes. The purpose of the map was to show relationships between overarching themes and their subthemes. The mapping part of the review was drafted by 1 member of the technical team, but the final framework of themes was further shaped and, when necessary, re-classified through discussion with at least 1 other member of the technical team. The Committee could then draw conclusions from each theme in each setting / country and how they may help in forming recommendations.

### 3.3.3 Type of studies

For most intervention reviews in this guideline, parallel randomised controlled trials (RCTs) were prioritised because they are considered the most robust type of study design that could produce an unbiased estimate of the intervention effects. The Committee expected there to be limited evidence of this type (due the study population being children or young people with life-limiting conditions), therefore non-randomised studies were also considered. This included consideration of uncontrolled studies (also called before-and-after studies without a control group - Higgins 2008).

For diagnostic reviews, cross-sectional and retrospective studies were considered for inclusion. For prognostic reviews, prospective and retrospective cohort studies were included. Case–control studies were not considered for inclusion.

In the qualitative reviews, studies using focus groups, structured or semi-structured interviews were considered for inclusion. Survey data or other types of questionnaires were only included if they provided analysis from open ended questions, but not if they reported descriptive quantitative data only.

Where data from observational studies were included, the Committee decided that the results for each outcome should be presented separately for each study and meta-analysis was not conducted.

### 3.3.4 Appraising the quality of evidence using 'Grading of Recommendations Assessment, Development and Evaluation' (GRADE)

#### 3.3.4.1 Elements of GRADE

For intervention reviews the evidence for outcomes from the included RCTs and observational studies, were evaluated and presented using GRADE developed by the international GRADE working group. Modified GRADE assessments were also carried out for outcomes per risk factor in prognostic reviews, for accuracy measures in diagnostic reviews and themes in qualitative reviews.

The software developed by the GRADE working group (GRADEpro) was used to assess the quality of each outcome, taking into account individual study quality factors and the meta-analysis results. This software is used mainly for intervention reviews, but can also be used for prognostic reviews. It is not presently designed to assess evidence from diagnostic and qualitative reviews. Therefore the modified GRADE approach for diagnostic and qualitative evidence was carried out without the software but using similar tables and concepts which are described below. Results were presented in GRADE profiles ('GRADE tables'), which consist of 2 sections: the 'Clinical evidence profile' table includes details of the quality assessment, while the 'Clinical evidence summary of findings' table includes pooled outcome data, and where appropriate, an absolute measure of intervention effect and the summary of the quality of evidence for that outcome. In this table, the columns for intervention and control indicate summary measures and measures of dispersion (such as mean and standard deviation or median and range) for continuous outcomes, and frequency of events (n/N: the sum across studies of the number of patients with events divided by sum of the number of completers as well as 95% confidence intervals) for binary outcomes. Reporting or

publication bias was only taken into consideration in the quality assessment and included in the 'Clinical evidence profile' table if it was apparent.

The evidence for each outcome was examined separately for the quality elements listed and defined in Table 2 for intervention, Table 3 for prognostic, Table 4 for diagnostic, and Table 5 for qualitative reviews. Each element was graded using the quality levels listed in Table 6. The main criteria considered in the rating of these elements are discussed below (see Section 3.3.4.2 Grading of the quality of clinical evidence). Footnotes were used to describe reasons for grading a quality element as having serious or very serious problems. The ratings for each component were summed to obtain an overall assessment for each outcome, see Table 7.

The GRADE toolbox is currently designed for randomised trials and observational studies only, but for this guideline the quality assessment elements and outcome presentation were adapted for all other review types (diagnostic, prognostic and qualitative studies).

**Table 2: Description of the elements in GRADE used to assess the quality of intervention studies**

| Quality element | Description |
|---|---|
| Risk of bias ('Study limitations') | Limitations in the study design and implementation may bias the estimates of the treatment effect. High risk of bias for the majority of the evidence decreases confidence in the estimate of the effect |
| Inconsistency | Inconsistency refers to an unexplained heterogeneity of results |
| Indirectness | Indirectness refers to differences in study population, intervention, comparator and outcomes between the available evidence and the review question, or recommendation made, such that the effect estimate is changed |
| Imprecision | Results are imprecise when studies include relatively few patients and few events and thus have wide confidence intervals around the estimate of the effect. Imprecision results if the confidence interval includes the clinically important threshold |
| Publication bias | Publication bias is a systematic underestimate or an overestimate of the underlying beneficial or harmful effect due to the selective publication of studies |

For evidence from diagnostic studies with regard to recognising signs and symptoms of dying, an adapted GRADE approach was planned. This looked at whether the identification of a particular sign or symptom could accurately indicate ('diagnose') that a child or young person was in the last days of life.

**Table 3: Description of the elements in GRADE and how they are used to assess the quality for diagnostic accuracy reviews**

| Quality element | Description |
|---|---|
| Risk of bias ('Study limitations') | Limitations in the study design and implementation may bias the estimates of the diagnostic accuracy. High risk of bias for the majority of the evidence decreases confidence in the estimate of the effect. Diagnostic accuracy studies are not usually randomised and therefore would not be downgraded for study design from the outset and start as high level evidence. |
| Inconsistency | Inconsistency refers to an unexplained heterogeneity of test accuracy measures such as sensitivity and specificity between studies. |
| Indirectness | Indirectness refers to differences in study population, differences in index tests across studies, reference standard and outcomes between the available evidence and the review question. |
| Imprecision | Results are considered imprecise when studies include relatively few patients and the probability to be diagnosed correctly in this group is low. Accuracy measures would therefore have wide confidence intervals around the estimate of |

| Quality element | Description |
|---|---|
| | the effect. |

For prognostic factors (that is, signs and symptoms which are risk factors for entering the last days of life), an adapted GRADE approach was conducted. This looked at the body of the evidence for each risk factor across studies for 1 outcome (in the case of this guideline, the outcome would be death occurring within 14 days).

**Table 4: Description of the elements in GRADE and how they are used to assess the quality for prognostic reviews**

| Quality element | Description |
|---|---|
| Risk of bias ('Study limitations') | Limitations in the study design and implementation may bias the estimates interpretation of the effect of the prognostic risk factor. High risk of bias for the majority of the evidence decreases confidence in the estimate of the effect. Prognostic studies are not usually randomised and therefore would not be downgraded for study design from the outset and start as high level evidence. |
| Inconsistency | Inconsistency refers to an unexplained heterogeneity between studies looking at the same sign or symptom resulting in wide variability between ORs, RRs, or HRs with little or no overlap in confidence intervals. |
| Indirectness | Indirectness refers to any departure from the review protocol, for instance differences in study population or risk factor that may affect how results can be generalised from the reviewed evidence. |
| Imprecision | Results are considered imprecise when studies include relatively few patients and also when the number of patients is too low for a multivariable analysis (as a rule of thumb a number of 10 participants per variable). This was assessed by looking at the confidence interval and where it lies in relation to the point estimate of the study. |

For qualitative studies an adapted GRADE-CERQual (Lewin 2015) approach was used. CERQual stands for Confidence in the Evidence from Reviews of Qualitative research. This looked at the quality of evidence by theme. These themes may have originated from an individual study or may have been identified through a number of individual themes or components of themes across a number of included studies.

**Table 5: Description of the elements in the adapted GRADE-CERQual approach used to assess qualitative evidence by theme**

| Quality element | Description |
|---|---|
| Risk of bias ('Study limitations') | Limitations in the study design and implementation may bias the estimates of the diagnostic accuracy. High risk of bias for the majority of the evidence decreases confidence in the estimate of the effect. Qualitative studies are not usually randomised and therefore would not be downgraded for study design from the outset and start as high level evidence. |
| Coherence of findings | The extent to which different individual themes or components of themes from studies fit into a wider network of overarching themes. For example, many components (relationship and rapport, clinical experience, information provision) can contribute to an overarching theme of healthcare professional factors in shared decision-making. Even though each individual study may not mention each factor the overall theme is coherent. |
| Applicability (or relevance) of evidence | The extent to which the evidence supporting the review finding is applicable to the context specified in the review question. In the case of this guideline qualitative evidence from the UK was prioritised over and above data from other contexts. |
| Theme | Theme saturation or sufficiency refers to a similar concept in qualitative |

| Quality element | Description |
|---|---|
| saturation / sufficiency | research. This refers to whether a theoretical point of theme saturation was achieved at which point no further citations or observations would provide more insight or suggest a different interpretation of this theme. Individual studies that may have contributed to a theme or subtheme may have been conducted in a manner that by design would have not reached theoretical saturation on an individual study level. |

The main criteria considered in the rating of these elements are discussed below (see Section 3.3.4.2 Grading of evidence). Footnotes were used to describe reasons for grading a quality element as having serious or very serious problems. The ratings for each component were summed to obtain an overall assessment for each outcome (Table 7).

#### 3.3.4.2 Grading the quality of clinical evidence

After data were synthesised, the overall quality of evidence was assessed for each outcome (in intervention or prognostic reviews), by diagnostic sign and symptom, or qualitative theme. The following procedure was adopted when using GRADE:

- An initial quality rating was assigned, based on the study design. RCTs start as High in intervention reviews, observational studies as Low, and uncontrolled case series as Low or Very low. In diagnostic, prognostic and qualitative reviews, evidence from non-randomised studies start as High.
- The rating was then downgraded for the specified criteria: risk of bias (study limitations), inconsistency, indirectness, imprecision and publication bias. These criteria are detailed below. In intervention reviews, evidence from observational studies (which had not previously been downgraded) was upgraded if there was: a large magnitude of effect, and/or a dose–response gradient, and/or if all plausible confounding would reduce a demonstrated effect or suggest a spurious effect when results showed no effect. Each quality element considered to have 'serious' or 'very serious' risk of bias was rated down by 1 or 2 points respectively.
- The downgraded or upgraded marks were then summed and the overall quality rating was revised. For example, all RCTs started as High and the overall quality became Moderate, Low or Very low if 1, 2 or 3 points were deducted respectively.
- The reasons or criteria used for downgrading were specified in the footnotes.
- For qualitative reviews a quality assessment of 'unclear' was added to the list of possible GRADE-CERQual levels. Together with the Committee it was decided that in qualitative reviews one 'unclear' rating did not mean an automatic downgrade of the evidence for this theme. However, 2 'unclear' ratings were downgraded by 1 and 3 'unclear' ratings downgraded by 2. Footnotes were not used for the CERQual tables.

**Table 6: Levels of quality elements in GRADE**

| Level | Description |
|---|---|
| None | There are no serious issues with the evidence |
| Serious | The issues are serious enough to downgrade the outcome evidence by 1 level |
| Very serious | The issues are serious enough to downgrade the outcome evidence by 2 levels |

**Table 7: Overall quality of outcome evidence in GRADE**

| Level | Description |
|---|---|
| High | Further research is very unlikely to change our confidence in the estimate of effect |
| Moderate | Further research is likely to have an important impact on our confidence in the estimate of effect and may change the estimate |

| Level | Description |
|---|---|
| Low | Further research is very likely to have an important impact on our confidence in the estimate of effect and is likely to change the estimate |
| Very low | Any estimate of effect is very uncertain |

The details of the criteria used for each of the main quality elements are discussed further in the following Sections 3.3.4.2.1 to 3.3.4.2.5.

#### *3.3.4.2.1 Risk of bias*

**Intervention studies**

Bias can be defined as anything that causes a consistent deviation from the truth. Bias can be perceived as a systematic error, for example, if a study was to be carried out several times and there was a consistently wrong answer, the results would be inaccurate.

The risk of bias for a given study and outcome is associated with the risk of over- or underestimation of the true effect.

The domains of risks of bias are listed in Table 8.

A study with a poor methodological design does not automatically imply that there is a high risk of bias; the bias is considered individually for each outcome and it is assessed whether this poor design will impact on the estimation of the intervention effect.

**Table 8: Domains of risk of bias in randomised controlled trials**

| Risk of bias | Explanation |
|---|---|
| Lack of allocation concealment | Those enrolling patients are aware of the group to which the next enrolled patient will be allocated (this is a major problem in 'pseudo' or 'quasi' randomised trials with, for example, allocation by day of week, birth date, chart number) |
| Lack of blinding | Patient, caregivers, those recording outcomes, those adjudicating outcomes, or data analysts are aware of the arm to which patients are allocated |
| Incomplete accounting of patients and outcome events | Missing data not accounted for and failure of the trialists to adhere to the intention-to-treat principle when indicated. Bias is suspected when the missing data is higher than the event rate and particularly when there is differential missing data between the groups in a trial (a difference of >10% was used). |
| Selective outcome reporting | Reporting of some pre-specified outcomes and not others (in particular if only significant results are reported) |
| Other risks of bias | For example:<br>Stopping the trial early for benefit observed in randomised trials, in particular in the absence of adequate stopping rules<br>Use of unvalidated patient-reported outcomes (for example, rating scales for noise intensity of respiratory secretions)<br>Recruitment bias in cluster-randomised trials |

**Diagnostic studies**

For diagnostic accuracy studies, the Quality Assessment of Diagnostic Accuracy Studies version 2 (QUADAS-2) checklist was used (see Appendix H in the NICE guidelines manual 2014). Risk of bias and applicability in primary diagnostic accuracy studies in QUADAS-2 consists of 4 domains:

**Table 9: Risk of bias for typical diagnostic accuracy studies (according to QUADAS-2)**

| Risk of bias | Explanation |
|---|---|

| Risk of bias | Explanation |
|---|---|
| Patient selection | It is assessed whether all the patients underwent all index tests or whether the index tests were appropriately randomised among the patients. |
| Index test (or sign/symptom) | Bias could be introduced when thresholds are not pre-specified, because this directly affects the sensitivity / specificity estimate for the study. |
| Reference standard | Usually this would be assessed by how well the reference standard is conducted. However, in the context of recognising dying this was not considered to be an appropriate factor. |
| Flow and timing | This is with regard to the timing of when the sign and symptom occurred in relation to when the person died. |

### Prognostic studies

For prognostic studies, quality was assessed using the checklist for prognostic studies (Appendix H in the NICE guidelines manual 2014).

This risk of bias for each risk factor across studies was derived by assessing the risk of bias across 6 domains for each study: selection bias, attrition bias, prognostic factor bias, outcome measurement bias, control for confounders and appropriate statistical analysis, with the last 4 domains being assessed for each outcome. A summary table on the quality of prognostic studies is presented at the beginning of each review to summarise the risk of bias across the 5 domains. More details about the quality assessment for prognostic studies are shown below:

**Table 10: Risk of bias for prognostic factor studies**

| Risk of bias | Explanation |
|---|---|
| Patient selection | Selection bias would be suspected if the allocation to groups directly leads to differences in baseline characteristics. If only 1 risk factor is considered, risk of bias may be introduced when there was no attempt to achieve roughly comparable groups, and/or there is evidence of biased selection. If 2 or more risk factors are considered, the same may not apply for patient selection issues and then the study would have to have controlled for confounders. |
| Prognostic factor bias (or sign/symptom) | This refers to any biases that could directly be linked to the validity of the prognostic factor under investigation, such as how the signs or symptoms were assessed or measured. |
| Attrition bias | This is assessed by whether there are similar numbers of people who were followed up in groups who have or have not got the particular sign or symptom. |
| Outcome measurement bias | This usually refers to whether or not the outcome has been measured on a validated scale or was otherwise reliably assessed. However, for the purpose of the 'recognising dying' review this was not considered to be an appropriate factor to assess. |
| Control for confounders / statistical analysis | This domain is an assessment of whether confounders have been adequately accounted for. Confounders would be signs and symptoms that may be related to dying but that are not under direct investigation. For instance age is related to dying, but we would not assess age in general as a sign or symptom of dying. We therefore wanted to assess whether signs and symptoms were independent predictors regardless of other non-related factors. |

### Qualitative studies

For qualitative studies, quality was assessed using a checklist for qualitative studies (as suggested in Appendix H in the NICE guidelines manual 2014). This was based on the Critical Appraisal Skills Programme (CASP) checklist for qualitative studies. The quality rating for risk of bias (low, high and unclear) was derived by assessing the risk of bias across 6 domains. The evidence was then GRADE across studies by theme as described above and labelled (no limitations, minor limitations, major limitations and unclear):

**Table 11: Risk of bias for qualitative studies**

| Risk of bias | Explanation |
|---|---|
| Aim and appropriateness of qualitative evidence. | This refers to an assessment of whether the aims and relevance of the study were clearly described and whether qualitative research methods were appropriate for investigating the research question. |
| Rigour in study design or validity of theoretical approach | This domain assesses whether the study approach has been clearly described and is based on a theoretical framework (for example, ethnography or grounded theory). This does not necessarily mean that the framework has to be explicitly stated, but that at least a detailed description is provided which makes it transparent and reproducible. |
| Sample selection | The background, the procedure, and reasons for the chosen method of selecting participants should be stated. It should also be assessed whether there was a relationship between the researcher and the informant, and if so, how this may have influenced the findings that were described. |
| Data collection | Consideration was given to how well the method of data collection (in-depth interviews, semi-structured interviews, focus groups or observations) was described, whether details were provided and how the data were collected (who conducted the interviews, how long did they last, and where did they take place). |
| Data analysis | For this criterion it is assessed whether sufficient detail is provided about the analytical process and whether it is in accordance to the theoretical approach. For instance if a thematic analysis was used it is assessed whether there was a clear description of how the theme was arrived at. Data saturation is also part of this section. This refers to whether a theoretical point of theme saturation was achieved at which point no further citations or observations would provide more insight or suggest a different interpretation of this theme. This could be explicitly stated or it may be clear from the citations presented that it may have been possible to find more themes. |
| Results | In relation to this section the reasoning about the results are important, for instance whether a theoretical proposal or framework is provided rather than being restricted to citations / presentation of data. |

#### *3.3.4.2.2 Risk of bias for evidence from the Delphi consensus study*

For the evidence from the Delphi study (included in Recognising Dying) we did not assess the quality using GRADE methodology, because it does not fall into a strict quantitative or qualitative category of data. An exception was made and for this type of evidence the quality was assessed by study quality only. There are published criteria for the assessment of Delphi studies and these were applied to see whether the methodology was used in a robust manner (Diamond 2014).

#### *3.3.4.2.3 Inconsistency / coherence of findings*

Inconsistency refers to unexplained heterogeneity of results. When estimates of the treatment effect, prognostic risk factor or diagnostic accuracy measures vary widely across studies (that is, there is heterogeneity or variability in results), this suggests true differences in underlying effects.

Heterogeneity in meta-analyses was examined, and if present, sensitivity and subgroup analyses were performed as prespecified in the protocols (Appendix D).

When heterogeneity existed (chi-squared $p<0.1$, I-squared inconsistency statistic of >50%, or from visually examining forest plots), but no plausible explanation could be found (for example, duration of intervention or different follow-up periods), the quality of the evidence was downgraded in GRADE by 1 or 2 levels, depending on the extent of inconsistency in the results. For diagnostic and prognostic evidence, this was assessed visually according to the

differences in point estimates and overlap in confidence intervals on the sensitivity / specificity forest plots. In addition to the I-squared and chi-squared values and examination of forest plots, the decision for downgrading was also dependent on factors such as whether the uncertainty about the magnitude of benefit (or harm) of the outcome showing heterogeneity would influence the overall judgment about net benefit or harm (across all outcomes).

For qualitative research, a similar concept to inconsistency is coherence, which refers to the way findings within themes are described and whether they make sense. This concept was used in the quality assessment across studies for individual themes. This does not mean that contradictory data was downgraded automatically, but that it was highlighted and presented, and that reasoning was provided. As long as the themes, or components of themes, from individual studies fit into a theoretical framework they do not necessarily have to have the same perspective. It should, however, be possible to explain these by differences in context (for example, the views of healthcare professionals might not be the same as those of family members, but they could contribute to the same overarching theme). Coherence was graded across studies with the following labels (coherent, incoherent, unclear).

#### *3.3.4.2.4 Indirectness / applicability or relevance of findings*

For quantitative reviews directness refers to the extent to which the populations, intervention/risk factor/index test, comparisons and outcome measures are similar to those defined in the inclusion criteria for the reviews. Indirectness is important when these differences are expected to contribute to a difference in effect size, or may affect the balance of harms and benefits considered for an intervention.

Relevance of findings in qualitative research is the equivalent of indirectness for quantitative outcomes, and refers to how closely the aims and context of the studies contributing to a theme reflect the objectives outlined in the review protocol of the guideline question.

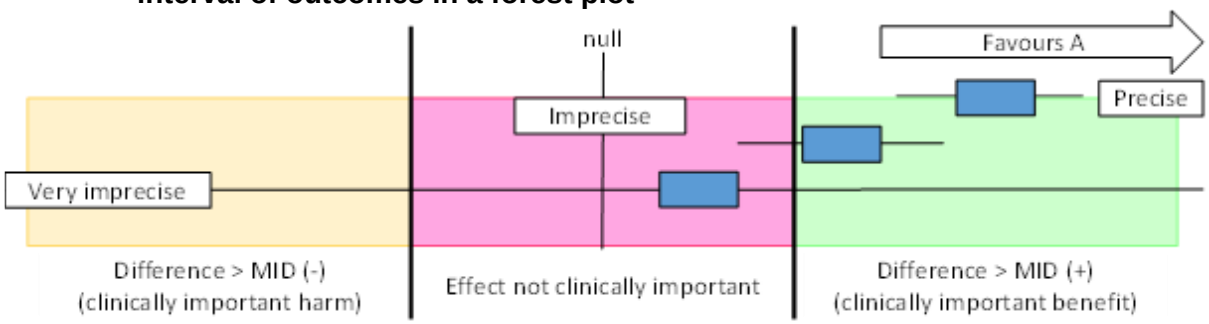
#### *3.3.4.2.5 Imprecision / theme saturation or sufficiency*

For quantitative reviews imprecision in guidelines concerns whether the uncertainty (confidence interval) around the effect estimate means that it is not clear whether there is a clinically important difference between interventions or not (that is, whether the evidence would clearly support 1 recommendation or appear to be consistent with several different types of recommendations). Therefore, imprecision differs from the other aspects of evidence quality because it is not really concerned with whether the point estimate is accurate or correct (has internal or external validity); instead, it is concerned with the uncertainty about what the point estimate actually is. This uncertainty is reflected in the width of the confidence interval.

The 95% confidence interval (95% CI) is defined as the range of values that contain the population value with 95% probability. The larger the trial, the smaller the 95% CI and the more certain the effect estimate.

Imprecision in the evidence reviews was assessed by considering whether the width of the 95% CI of the effect estimate was relevant to decision-making, considering each outcome in isolation. This has been explained in Figure 3, which considers a positive outcome for the comparison of treatment A versus treatment B. Three decision-making zones can be identified, bounded by the thresholds for clinical importance (minimal important difference – MID) for benefit and for harm. The MID for harm for a positive outcome means the threshold at which drug A is less effective than drug B by an amount that is clinically important to patients (favours B).

**Figure 3: Illustration of precise and imprecise outcomes based on the confidence interval of outcomes in a forest plot**




When the confidence interval of the effect estimate is wholly contained in 1 of the 3 zones (for example, clinically important benefit), we are not uncertain about the size and direction of effect (whether there is a clinically important benefit, or the effect is not clinically important, or there is a clinically important harm), so there is no imprecision.

When a wide confidence interval lies partly in each of 2 zones, it is uncertain in which zone the true value of effect estimate lies, and therefore there is uncertainty over which decision to make (based on this outcome alone). The confidence interval is consistent with 2 possible decisions and so this is considered to be imprecise in the GRADE analysis and the evidence is downgraded by 1 level ('serious imprecision').

If the confidence interval of the effect estimate crosses into 3 zones, this is considered to be very imprecise evidence because the confidence interval is consistent with 3 possible clinical decisions, and there is therefore a considerable lack of confidence in the results. The evidence is therefore downgraded by 2 levels in the GRADE analysis ('very serious imprecision').

Implicitly, assessing whether the confidence interval is in, or partially in, a clinically important zone, requires the Committee to estimate an MID or to say whether they would make different decisions for the 2 confidence limits.

The literature was searched for established MIDs for the selected outcomes in the evidence reviews such as symptom measurement tools. However none were identified for our guideline population. In addition, the Committee was asked whether they were aware of any acceptable MIDs in the clinical community. Finally, the Committee considered whether it was clinically acceptable to use the GRADE default MID to assess imprecision: for binary outcomes a 25% relative risk reduction or relative risk increase was used, which corresponds to clinically important thresholds for a risk ratio of 0.75 and 1.25 respectively. This default MID was used for all the binary outcomes in the interventions evidence reviews as a starting point and decisions on clinical importance were then considered based on the absolute risk difference. For continuous outcomes default MIDs were also used. These use half of the median standard deviation of the control group.

The same principle was used for prognostic factors, for example using the default MID as a starting point for the Committee discussion, to assess whether the size of the outcome effect would be large enough to be meaningful in clinical practice.

In diagnostic accuracy measures it was first of all considered whether sensitivity or specificity (or AUC for continuous variables) was going to be given more weight in the decision-making process. If 1 measure was given more importance than the other, then imprecision was rated on this statistical measure. It was not possible to pool the diagnostic data in this guideline. Therefore imprecision was assessed on individual study results. For the purpose of the 'recognising dying' review the focus was on specificity. A specificity value of above 90% was

considered by the Committee a good indicator of a sign or symptom that if found positive would be associated with death in the next days (that is, 90% or above of people who were classified positive as having this sign / symptom). This was then used in the same manner as an MID described above. A specificity value would be described as imprecise if it crossed this 90% threshold, and very imprecise if it also crossed the chance value of 50%.

Theme saturation or sufficiency refers to a similar concept in qualitative research. This refers to whether a theoretical point of theme saturation was achieved at which point no further citations or observations would provide more insight or suggest a different interpretation of this theme. As already highlighted in a previous section on qualitative reviewing methods it is not equivalent to the number of studies contributing to a theme, but rather to the depth of data and whether sufficient quotes / observations were provided that could underpin these findings.

#### *3.3.4.2.6 Assessing clinical significance (of intervention effects)*

The Committee assessed the evidence by outcome in order to determine if there was, or potentially was, a clinically important benefit, a clinically important harm or no clinically important difference between interventions. To facilitate this, where possible, binary outcomes were converted into absolute risk differences (ARDs) using GRADEpro software: the median control group risk across studies was used to calculate the ARD and its 95% CI from the pooled risk ratio. For continuous outcomes the mean difference between the intervention and control arm of the trail was calculated. This was then assessed in relation to the default MID (0.5 times the median control group standard deviation).

The assessment of clinical benefit, harm, or no benefit or harm was not based on the default MID of the relative risk which was only used as a starting point, but on the point estimate of the absolute effect, taking into consideration the precision around this estimate.

This assessment was carried out by the Committee for each critical outcome, and an evidence summary table (this additional table was used in the Committee meetings but is not presented in this guideline) was produced to compile the Committee's assessments of clinical importance per outcome, alongside the evidence quality and the uncertainty in the effect estimate (imprecision). In instances where the Committee decision differed from the default assessment decisions were captured in the Linking Evidence to Recommendations sections.

#### *3.3.4.2.7 Assessing clinical significance (of prognostic, diagnostic or qualitative findings)*

Absolute risk differences were not calculated for prognostic findings in this guideline. The Committee considered the size of the relative effects and whether this was large enough to constitute a sign or symptom predicting whether someone would die within the next few days.

In a similar manner this was carried out for diagnostic accuracy statistics to interpret how likely the size of the effect reflects a clinically meaning association between people having a sign or symptom and whether or not they die in the next few days.

For themes stemming from qualitative findings, clinical importance was decided upon by the Committee taking into account the generalisability of the context from which the theme was derived, and whether it was convincing enough to support or warrant a change in current practice as well as the evidence quality.

### 3.3.5 Evidence statements

Evidence statements are summary statements that are presented after the GRADE profiles, summarising the key features of the clinical evidence presented. The wording of the evidence statements reflects the certainty or uncertainty in the estimate of effect. The

evidence statements are presented by outcome or theme and encompass the following key features of the evidence:

- The quality of the evidence (GRADE rating)
- the number of studies and the number of participants for a particular outcome
- a brief description of the participants
- an indication of the direction of effect (for example, if 1 treatment is beneficial or harmful compared with the other, or whether there is no difference between the tested treatments)

### 3.3.6 Evidence of cost effectiveness

The Committee is required to make decisions based on the best available evidence of both clinical and cost effectiveness. Guideline recommendations should be based on the expected costs of the different options in relation to their expected health benefits (that is, their 'cost effectiveness') rather than the total implementation cost. Thus, if the evidence suggests that a strategy provides significant health benefits at an acceptable cost per patient treated, it should be recommended even if it would be expensive to implement across the whole population.

Evidence on cost effectiveness related to the key clinical issues being addressed in the guideline was sought, and a systematic review of the published economic literature was undertaken.

#### 3.3.6.1 Literature review

The health economist:

- Identified potentially relevant studies for each review question from the economic search results by reviewing titles and abstracts. Full papers were then obtained.
- Reviewed full papers against pre-specified inclusion and exclusion criteria to identify relevant studies (see below for details).
- Critically appraised relevant studies using the economic evaluations checklist as specified in the NICE guidelines manual
- Studies initially considered eligible but which were then excluded can be found in Appendix H with reasons for exclusion explained.

#### 3.3.6.2 Inclusion and exclusion criteria

Full economic evaluations (studies comparing costs and health consequences of alternative courses of action (cost–utility, cost-effectiveness, cost–benefit and cost–consequences analyses) and comparative costing studies that addressed a guideline review question in the relevant population were considered potentially includable as economic evidence.

Given the sparsity of full economic evaluations in the search, rigid exclusion criteria were not applied and articles were considered for inclusion if there was a significant resource content in a context relevant to a review question in the guideline.

### 3.3.7 Undertaking new health economic analysis

As well as reviewing the published economic literature for guideline review questions, as described above, new economic analysis was undertaken by the health economist in selected areas. Priority areas for new health economic analysis were agreed by the Committee after formation of the review questions and consideration of the available health economic evidence. Owing to a lack of clinical or effectiveness evidence, these new analyses focused on costing aspects of service delivery.

### 3.3.8 Cost effectiveness criteria

It was recognised in the scope that the use of QALYs was difficult in the context of end of life care for children and young people. The problems include the difficulties of eliciting health state utilities in this population, the often limited duration of life which means that any QALY gains will typically be very small and ethical issues around using conventional NICE cost-effective decision rules.

NICE's report 'Social value judgements: principles for the development of NICE guidance' sets out the principles that the Committee s should consider when judging whether an intervention offers good value for money but also that that cost-effectiveness is not the sole criterion for making decisions and for the above mentioned reasons this is especially the case for this guideline

In general, an intervention was considered to be cost effective if either of the following criteria applied (given that the estimate was considered plausible):

- the intervention dominated other relevant strategies (that is, it was both less costly in terms of resource use and more clinically effective compared with all the other relevant alternative strategies), or
- the intervention provided clinically significant benefits at an acceptable additional cost when compared with the next best strategy.

### 3.3.9 In the absence of economic evidence

When no relevant published studies were found, the Committee made a qualitative judgement about cost effectiveness by considering expected differences in resource use between options and relevant UK NHS unit costs, alongside the results of the clinical review of effectiveness evidence and their expert opinion.

The costs reported in the guideline are those that were presented to the Committee and were correct at the time recommendations were drafted. They may have changed subsequently before the time of publication but, we have no reason to believe they have changed substantially.

## 3.4 Involving children and young people with life-limiting conditions in this guideline – focus group research

### 3.4.1 Background

An integral part of guideline development process was the involvement of people with direct experience of the condition and the services available to them. The Committee included 2 mothers of children who had died because of a life-limiting condition. They contributed as full guideline members, developing review questions, highlighting sensitive issues and terminology, and bringing the experience of parents to the attention of the rest of the Committee. However, as part of the scoping process it was identified that there was limited evidence directly from the children and young people's perspective. For this topic it was considered crucial that the experiences, perspectives and opinions of children and young people would be incorporated in the guideline. The topics which were prioritised to benefit particularly from the input of children and young people with life-limiting conditions were the following:

- information: the information given to them with regard to their condition and its management
- communication: how that information should be made available, for example 1-to-1 discussion

- place of care: their views on where they would ideally like to receive care – and the factors that influence their thoughts on this
- care planning: how they would like to be involved in planning their own care
- psychological and other support needs: what kind of emotional and other support they consider to be important and helpful to them in living with their condition.

Additionally, the Committee also wanted to know what children and young people with life-limiting conditions thought about their current care in terms of:

- What areas of care were currently being 'done well' and where was the care less satisfactory
- If they could change one thing about their care, what would it be?

Focus groups with children and young people with life-limiting conditions were conducted for this guideline. The findings of this research was used as direct evidence in Chapters 4 'Providing information' and 5 'Communication, and in Sections 6.1 and 8.18.1. The details of this primary research project can be found in Appendix L.

This work was carried out by Together for Short Lives, an organisation representing the needs of children and young people with life-limiting conditions.

### 3.4.2 Methods with regard to the focus group

The details of the focus group methodology are described in Appendix L. For the purpose of this methodological chapter a short description of the method is provided in this section. The organisation conducted 3 focus groups: 1 in the North of England (Yorkshire), 1 in Bristol (where Together for Short Lives is based) and 1 centrally, in London, in order to ensure broad representation of participants across the UK. A total of 14 young people took part (7 male, 7 female), ranging in age from 12 to 18 years. Conditions included spinal muscular atrophy, cancer, cystic fibrosis, and other rare degenerative and life-threatening conditions. Key findings were shared with all participants; feedback received from 7 young people was used to help interpret the findings of the focus group.

### 3.4.3 Drawing on children's and young people's views to inform recommendations

A member of the research team from Together for Short Lives presented the findings from the focus group at a Committee meeting and the full report was circulated. The themes that emerged were presented to the Committee and together with any other identified evidence for the topics, were taken into consideration when the recommendations were drafted. This was the most applicable evidence for a number of the topics covered by the guideline, and therefore influenced the recommendations directly. The Committee therefore decided to highlight the contributions of the children and young people in a specific section in the 'Evidence to Recommendations' sections of the guideline, which provide the rationale for the recommendations.

## 3.5 Developing recommendations

Over the course of the guideline development process, the Guideline Committee was presented with:

- evidence tables of the clinical and economic evidence reviewed from the literature (all evidence tables are in Appendix G)
- summaries of clinical and economic evidence and quality assessment (as presented in Chapters 5 to 11)
- forest plots, when applicable (Appendix I)

- a description of the methods and results of the cost-effectiveness analysis undertaken for the guideline (Appendix K).

Recommendations were drafted on the basis of the Committee's interpretation of the available evidence. For intervention studies this would mean taking into account the balance of benefits, harms, and costs between different courses of action. This was either done formally, in an economic model, or informally. Firstly, the net benefit over harm (clinical effectiveness) was considered in discussion with the Committee, focusing on the critical outcomes. When this was done informally, the Committee took into account the clinical benefits and harms when 1 intervention was compared with another. The assessment of net benefit was moderated by the importance placed on the outcomes (the Committee's values and preferences), and the confidence the Committee had in the evidence (evidence quality). Secondly, the Committee assessed whether the net benefit justified any differences in costs.

When clinical and economic evidence was of poor quality, conflicting or absent, the Committee drafted recommendations based on their expert opinion. The considerations for making consensus-based recommendations include the balance between potential harms and benefits, the economic costs or implications compared with the economic benefits, current practices, recommendations made in other relevant guidelines, patient preferences and equality issues. The Committee also considered whether the uncertainty was sufficient to justify delaying making a recommendation and to await further research.

The wording of recommendations was agreed by the Committee and focused on the following factors:

- the actions healthcare professionals need to take
- the information readers need to know
- the strength of the recommendation (for example the word 'offer' was used for strong recommendations and 'consider' for weak recommendations)
- the involvement of people with the condition (and their parents or carers if needed) in decisions about treatment and care
- consistency with NICE's standard advice on recommendations about drugs, waiting times and ineffective interventions.

In cases of qualitative evidence, the Committee considered the themes that had been identified from the meta-synthesis or from the focus group (for instance barriers and facilitators for effective care planning), and assessed whether they were generalisable to the NHS context. This included an interpretation of how a concept originating from a named theme from the literature could apply to clinical practice. For example, in the 'Religious, spiritual and cultural support (in chapter 8.3) review the theme of 'ready to die and go to heaven' may highlight that clinicians should be aware of the impact of religious, spiritual and cultural beliefs on end of life care planning.

The main considerations of the Committee specific to each recommendation are outlined in the 'Recommendations and link to evidence' sections within each chapter.

### 3.5.1 Research recommendations

When areas within reviews were identified for which good evidence was lacking, the Committee considered making recommendations for future research. Decisions about inclusion were based on factors such as:

- the importance to patients or the population
- national priorities
- potential impact on the NHS and future NICE guidance
- ethical and technical feasibility.

### 3.5.2 Validation process

This guidance is subject to a 6-week public consultation and feedback as part of the quality assurance and peer review of the document. All comments received from registered stakeholders are responded to in turn and posted on the NICE website when the pre-publication check of the full guideline occurs.

### 3.5.3 Updating the guideline

Following publication, and in accordance with the NICE guidelines manual, NICE will regularly undertake a review of whether the evidence base has progressed significantly to alter the guideline recommendations and warrant an update.

### 3.5.4 Disclaimer

Healthcare providers need to use clinical judgement, knowledge and expertise when deciding whether it is appropriate to apply guidelines. The recommendations cited here are a guide and may not be appropriate for use in all situations. The decision to adopt any of the recommendations cited here must be made by practitioners in light of individual child or young person patient circumstances, the wishes of the patient and their parents or carers, clinical expertise and resources.

The National Guideline Alliance (NGA) disclaims any responsibility for damages arising out of the use or non-use of these guidelines and the literature used in support of these guidelines.

### 3.5.5 Funding

The NGA was commissioned by NICE to undertake the work on this guideline.

# 4 Providing Information

## 4.1 Review question

**What information and information type (written or verbal) is perceived as helpful and supportive by children and young people (if appropriate), and their family or carer before and after an infant, child or young person dies including managing practical arrangements, and care of the body?**

## 4.2 Introduction

The provision of information to parents and families around the time of a child's death is thought to be an essential component of palliative care. Such information needs to cover a wide range of detail, from specific matters such as the practical management of their child's symptoms, information to support difficult and complex decision-making about the care of their child, through to practical day to day issues such as provision of car parking or accommodation at a care facility. Most parents have never faced end of life decisions before, and feel that they are in completely unknown territory, needing some insight into what to expect.

Throughout a child or young person's life-limiting illness, there is a need for the parents or carers to understand medical facts, including diagnoses and prognoses. Therefore, what information and how it is provided is critically important. Good information provision can also build trust between parents or carers and healthcare professionals, and can promote the emotional wellbeing of the family or carers after their child or young person's death. After a child or young person has died, their parents or carers may need information about a number of practical decisions and formal matters that need to be addressed.

There is a need to determine what information parents require, what such material should contain and how it is best presented. This review seeks to explore what information, and what information type, is felt to be most helpful to parents and carers around the time of their child's death.

## 4.3 Description of clinical evidence

The aim of this review was to identify the content and type of information that is experienced as helpful and supportive or a hindrance, by the children or young people and their families or carers. It related to the periods before and after a child or young person dies and covered the life-limiting condition, likelihood of death and practical arrangements, and care of the body.

Qualitative studies were selected for inclusion for this review. Studies that collected data using qualitative methods (such as by using semi-structured interviews, focus groups, and surveys with open-ended questions) and analysed data qualitatively (including thematic analysis, framework thematic analysis, content analysis and so on) were looked for. Survey studies restricted to reporting descriptive data that were analysed quantitatively were excluded.

One meta-synthesis (Xafis 2015) on parents' information needs when facing end of life decisions for their child was retrieved during the re-runs stage (the second review of the published literature during the last months of the guideline development phase when searches are updated) of the guideline development. Relevant individual studies of this review were cross-checked and many of them have been included for this review. Because most of the themes identified and reported in this meta-synthesis have also been identified and reported in our review, the metasynthesis itself was not included in this review.

Given the nature of qualitative reviews, findings/themes have been summarised from the literature and were not restricted to those identified as likely themes by the Committee (which were use of jargon and terminology, uncertainty around the likelihood of death, method of information provision, choices and options, and direct practical information).

For full details see review protocol in Appendix D.

A total of 21 studies were identified for inclusion in this review. Of them:

- 15 studies focused on the perspectives of parents or carers whose child had died due to a life-limiting illness, or who were caring for a child with a life-limiting condition (Branchett 2012; Contro 2002; DeJong-Berg 2006; James 1997; Laakso 2001; Laakso 2002; Meert 2007; Michelson 2013; Midson 2010; Price 2011; Richardson 2003; Rini 2007; Sullivan 2014; Yuen 2012; Wocial 2000);
- 2 studies interviewed both the parents and their child living with life-limiting conditions (Hsiao 2007; Hunt 2013); 1 study carried out a survey among social workers who had provided services to families with a child living with a life-limiting condition (Jones 2006);
- 2 studies involved both parents and service providers (Monterosso 2007; Kavanaugh 2010);
- 1 study focused on the perspectives of healthy siblings whose sister or brother had died of cancer (Nolbris 2005).

The majority of included studies collected data by semi-structured interviews or focus groups, 3 studies collected data by open-ended questions in survey questionnaires (Branchett 2012; dejong-Berg, 2006; Jones 2006). The most common data analysis method employed across studies was thematic analysis.

With regard to the setting of studies:

- 4 studies were conducted in the UK (Branchett 2012; Midson 2010; Price 2011; Hunt 2013);
- 8 in the USA (Control 2002; Hsiao 2007; Jones 2006; Kavanaugh 2010; Meert 2007; Michelson 2013; Rini 2007; Wocial 2000);
- 2 in Australia (Monterosso 2007; Sullivan 2012);
- 2 in Canada (DeJong-Berg 2006; James 1997);
- 2 in Finland (Laakso 2001; Laakso 2002);
- and 1 each in Sweden (Nolbris 2005), Ireland (Richardson 2003), and the Netherlands (Yuen 2012)

Except for information specifically relating to care of the body, evidence on all themes considered important by the Committee was identified. A number of further themes emerged from studies and were incorporated in the review.

To include the views of children and young people with life-limiting conditions and direct experience of the health service in the UK, a focus group was commissioned specifically for this guideline. A description of how this research contributed to the recommendations were added to the the ‘Linking evidence to recommendations’ section of this chapter (see section 4.8).

A brief description of the studies is provided in Table 12.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix G and in the GRADE profiles below. See also the TFSL focus group report in Appendix L. For presentation of findings, a theme map was generated according to the themes that emerged

from the studies (Figure 1). The mapping part of the review was drafted by 1 researcher from the guideline technical team but the final framework of themes was further shaped, and if necessary re-classified, through discussions with at least 1 other researcher. Due to the qualitative nature of these studies, evidence is summarised in adapted GRADE-CERQual tables within the evidence report. Therefore, no separate Appendix is provided for this.

## 4.4 Summary of included studies

A summary of the studies that were included in this review are presented in Table 12.

**Table 12: Summary of included studies**

| Study | Data collection methods | Participants /respondents | Aim of the study | Comments |
|---|---|---|---|---|
| Interviews/focus-groups | | | | |
| Contro 2002 | Interviews | N=68 parents/carers representing 44 families<br>USA | To obtain personal accounts of families' experiences to learn ways to improve care for paediatric patients and their families. | • data collection and analysis clearly reported<br>• researchers' role and potential influences in the analytical process not critically reviewed |
| Hsiao 2007 | Interviews | N= 20 parent and child pairs under the care of a paediatric oncology and cardiology department<br>USA | To identify the aspects of physician communication that children with life-limiting illnesses and their parents perceived to be facilitative or obstructive in paediatric palliative care. | • response rate for invited subjects was 57%;<br>• recruitment of patients was through healthcare providers who may have differing opinions on whether a patient fits the prognosis criteria;<br>• data collection and analysis clearly reported |
| Hunt 2013 | Interviews | N= 59 (41 parents plus 18 CYP); children diagnosed with a life-limiting condition and their families receiving palliative care<br>UK | To understand the met and unmet needs of children with life-limiting conditions and their families (Strand 2 of The Big Study for life-limited children and their families) | • data collection and analysis clearly reported<br>• researchers' role in the analytical process not critically reviewed |
| James 1997 | Interviews | N=12 parents (of children who had died of various types of cancer 1 to 3 years ago)<br>Canada | To identify parents' perceptions of their needs while their child was dying of cancer | • the method of sample selection may have created a biased sample. 46 families met the inclusion criteria, the physician eliminated 19 families for various reasons; 27 letters were sent out and 12 parents made up the final sample. Those families eliminated by the physicians may have been those with the |

| Study | Data collection methods | Participants /respondents | Aim of the study | Comments |
|---|---|---|---|---|
| | | | | highest levels of need during their child's palliative care |
| Kavanaugh 2010 | Interviews | N= 40 cases involved 54 parents/carers and 71 healthcare professionals who were in discussion about treatment decisions for an infant, due to threatened preterm delivery<br><br>USA | To describe nurse behaviours that assisted parents in making life support decisions for an extremely premature infant before and after the infant's death. | • a semi-structured interview guide was used<br>• data collection and analysis was guided by the Ottawa Decision Support Framework, clearly reported<br>• the evidence was indirect because the main focus of the study was not on information perceived or experienced as helpful/unhelpful |
| Laakso 2001 | Interviews | N= 50 mothers whose child died from illness under the age of 7<br>Finland | To analyse the mother's grief and coping with grief following the death of a child under the age of 7 years. | • low response rate: 174 mothers were contacted, only 50 interviewed |
| Laakso 2002 | Interviews | N=50 mothers whose child died between 1990 and 1994<br>Finland | To describe the grief and coping of mothers whose child had died under the age of 7 years. The paper describes the social support received as experienced by mothers. | • low response rate: 174 mothers were contacted, only 50 interviewed<br>• unclear whether saturation in data collection or analysis was achieved |
| Meert 2007 | Interviews | N= 56 parents whose child died 12 months earlier<br>USA | To investigate parents' perspectives on the desirability, content and conditions of a physician-parent conference after their child's death in the paediatric intensive care unit (PICU) | • interview guides were used during data collection<br>• data collection and analysis clearly reported<br>• a large number of eligible parents could not be contacted and there was a majority of mothers among participants |
| Michelson 2013 | Focus groups and interviews | N= 18 parents whose child died in the PICU between 2007 and 2009<br>USA | To describe the roles and respective responsibilities of PICU healthcare professionals (HCPs) in end of life care decisions faced by PICU parents. | • saturation of data collection was achieved; data analysis methods not clearly reported<br>• interviewers were physicians and social workers who were unknown to the parents |
| Midson 2010 | Interviews | N=55 parents | To explore the | • interviews were |

| Study | Data collection methods | Participants /respondents | Aim of the study | Comments |
|---|---|---|---|---|
| | | whose child died under the age of 17 years, between 12 and 18 months ago.<br>UK | experiences of parents within 1 tertiary centre, and the challenges that lay ahead in changing the barriers, attitudes, and culture that impede some aspects of end of life care. | conducted by phone, home visits or in a hospital room<br>• unclear about the relationship between the researchers and interviewees; researchers' roles and pre-knowledge and their influences on data collection and analysis not critically reviewed |
| Monterosso 2007 | Interviews | N= 38 parents plus 20 service providers<br>Australia | To obtain feedback from families of children receiving palliative and supportive care about their care needs in hospital and in community settings. | • data collection process clearly reported, however data analysis process was not described in detail<br>• researchers did not critically review their roles in the analytical process |
| Nolbris 2005 | Interviews | N=10 siblings whose brothers or sisters died of cancer 1.5 to 6 years ago<br>Sweden | To explore siblings' needs and issues when a brother or sister died of cancer, interviews were conducted with 10 surviving children and young adults. Of particular interest was their individual participation in and experience of the period of disease, dying and mourning. | • long interval between the siblings' deaths and interview time, so there may be recall bias<br>• researchers did not critically review their roles in the process |
| Price 2011 | Interviews | N=25 parents whose child had died from a life-limiting condition between 6 and 24 months earlier<br>UK | To redress the gaps in knowledge by exploring, retrospectively, parents' experiences of caring for children with both malignant and non-malignant conditions throughout the entire trajectory of their child's illness and subsequent death. | • the sample consisted of primarily of parents employed and of 'middle class'. The importance of social class in mediating experiences of illness should be noted;<br>• data collection and analysis process clearly reported |
| Redmond 2003 | Interviews | N= 17 mothers of children aged ≤ 4 years with severe intellectual disability and life-limiting condition | To explore the mothers' views of the usefulness of the financial, practical and emotional supports being offered to them and their suggestions for service | • data analysis process not clearly reported; the researchers' roles and potential influences in the analytical process not clearly reported |

| Study | Data collection methods | Participants /respondents | Aim of the study | Comments |
|---|---|---|---|---|
| | | Ireland | improvements. | |
| Rini 2007 | Interviews | N= 11 parents whose child died in the PICU<br>USA | To describe the presence (or the absence) and the role of anticipatory mourning in parents who recently experienced the death of a hospitalised child and to determine if there were consistent factors that they described as helpful or detrimental to them during this process. | • all parents who consented to the interviews were Caucasian<br>• data collection and analysis clearly reported<br>• saturation in data collection and analysis not clearly reported, researchers' roles in the analytical process not reported<br>• study results were verified by 2 parents interviewed |
| Sullivan 2014 | Interviews | N= 25 bereaved parents whose child died at the age between 3 months and 12 years<br>Australia | To examine parents' views and experiences of end of life decision-making. | • researchers' roles in the analytical process not critically reviewed<br>• unsure whether saturation in data collection or analysis achieved |
| Yuen 2012 | Interviews | N= 16 parents who had lost a child to lethal epidemolysis bullosa 1 year earlier<br>The Netherlands | To identify the needs of parents of parents who lost their child to lethal epidermolysis bullosa | • 25 parents were contacted for interview, 16 consented<br>• data analysis process not clearly reported<br>• the researchers' role and influences in the analytical process not critically reviewed |
| Wocial 2000 | Interviews | N=20 parents whose infants received (neonatal intensive care unit) NICU care<br>USA | To understand better parent perceptions of the decision-making process by making the following determinations including: what information was important to parents in reaching a decision about withholding and/or withdrawing treatment from their infants. | • informants of the study were a fairly homogeneous group<br>• study findings verified by a clinical expert in neonatal nursing<br>• researchers' role in and influences in the analytical process not critically reviewed |
| **Surveys** | | | | |
| Branchett, 2012 | Survey with open-ended | N=57 parents who had lost a child in the | To determine what parents had actually experienced relating | • Data were collected by a few simple open-ended questions initially posted |

| Study | Data collection methods | Participants /respondents | Aim of the study | Comments |
|---|---|---|---|---|
| | questions | neonatal period<br><br>UK | to neonatal palliative and end of life care and determine how this knowledge could be used to improve experiences for families in future. | on a parent's website. No guide was used to design and collect data. The study was undertaken by 1 researcher as part of scoping exercise within a bigger project therefore may lack some of the formal research rigour. |
| deJong-Berg 2006 | Survey with open-ended questions | N=29 parents/cares who had experienced the death of a child at the hospital or at home or were served by Children's Homecare<br>Canada | To evaluate a programme providing standard bereavement follow-up service after its 3 years' delivery | • Low response rate: 82 families were eligible, only 29 parents representing 21 families returned the survey<br>• Information perceived helpful/unhelpful was not the main focus of the study<br>• Data collection and analysis clearly reported implication of data collected by surveys not critically reviewed; |
| Jones 2006 | Survey with open-ended questions | N=131 social workers of a national voluntary membership organisation<br>USA | To identify the social workers' perspectives regarding the psychosocial needs of children with cancer at the end of life and their families | • 50% response rate to the study survey<br>• the survey used in the study was not previously validated through formal testing<br>• data collection and analysis process clearly reported |

Five categories/themes of information and information types that were found to be helpful before and after a child and young person died emerged, or were derived from included studies. The central theme was the need for timely, honest, accurate and consistent information which was a feature throughout all subthemes. People also reported that they found disease specific information; practical information; personalised information; and information that allowed active involvement of parents in the course of end of life care for their child (active involvement information) to be helpful.

## 4.5 Clinical evidence

### 4.5.1 Theme map

The theme map for Providing Information is presented in Figure 4.

### 4.5.2 Clinical evidence profile

At the centre of the map is the main theme which is overarching and was mentioned as part of most of the other themes and subtheme

**Figure 4: Theme map – barriers and facilitators for effective information provision**

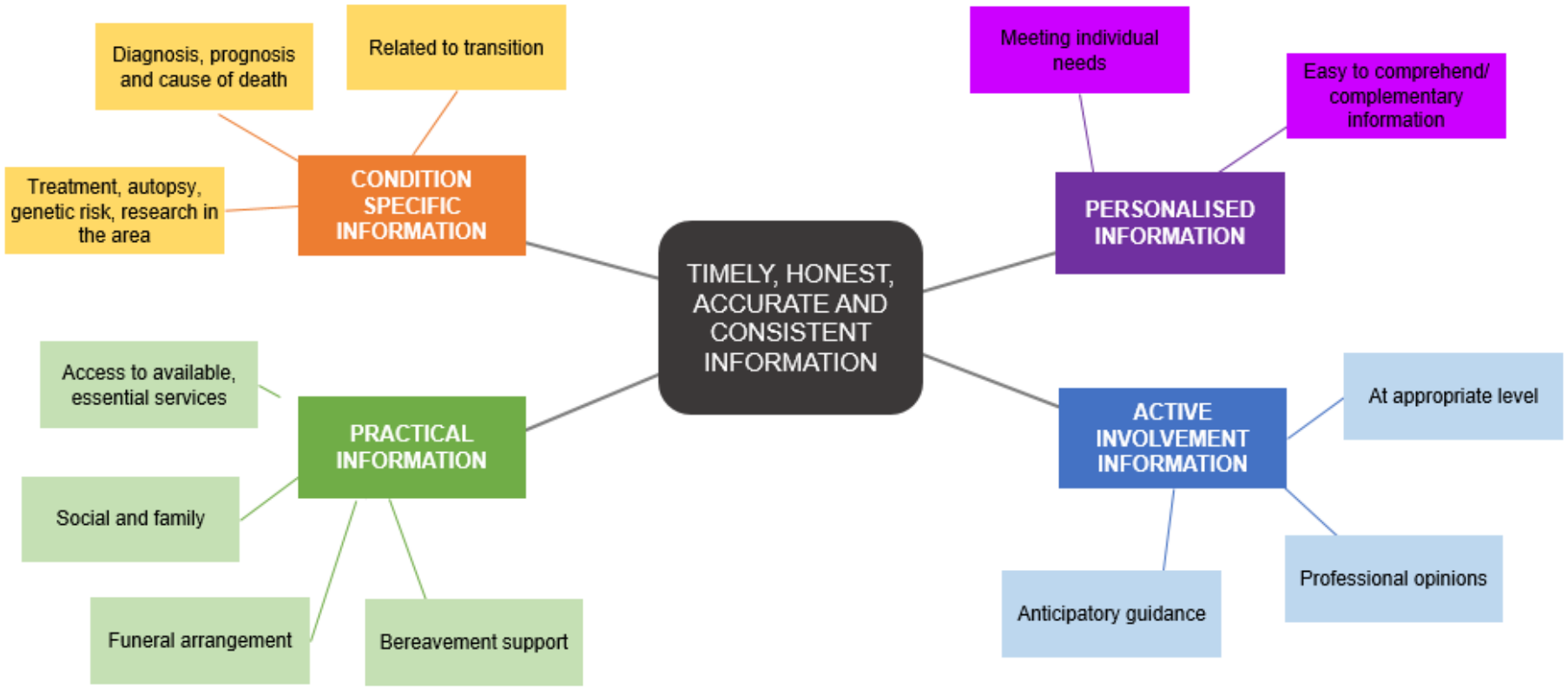



**Table 13: Summary of evidence (adapted GRADE-CER-Qual): Theme 1 – Timely, honest, accurate and consistent information type/content that was perceived helpful during the end of life care for children and young people**

| Study information | Description of theme or finding | Quality assessment |
|---|---|---|

| Number of studies | Design | | Criteria | Rating | Overall |
|---|---|---|---|---|---|
| **Timely, honest, accurate and consistent information** | | | | | |
| 10 studies (Branchett 2012; Contro 2002; Hsiao 2007; Hunt 2013; Jones 2006; Laakso 2001; Laakso 2002; Meert 2007; Monterosso 2007; Yuen 2012) | 9 studies used interviews and 1 study used surveys | 10 studies conducted in different settings (UK, Australia, Finland, USA, and the Netherlands) among parents and social workers reported that parents wanted timely, honest, accurate and consistent information in the end of life care for their infant/child, particularly at the points of diagnosis, transition and when a change of treatment occurred, the prognosis that the child has been recognised as being in the last days of life has to be communicated, and end of life issues/choices have to be discussed/made.<br><br>**Diagnosis:**<br>*"Be honest with parents and don't be scared of telling the truth. People cope – they don't have a choice"*<br>Transition:<br>*"Please keep parents informed. It seems a constant uphill struggle to obtain information...particularly in the hours immediately after delivery of transfer"*<br><br>**Being recognised as in the last days of life:**<br>Although parents thought it was hard to hear the news, they were glad they were informed honestly.<br>*"He could not make it better than it was. It was very hard to hear it, but on the other side, he couldn't have told it in a different way. I wouldn't want that" (parent)*<br>*"If you are not honest with people, then they keep hope...That will give problems, as you will give them more [treatment]. That should not happen" (parent)*<br><br>**Disease progression, and end of life issues/choices:**<br>*"Families need open discussion of disease...progression, symptom options and end of life issues/choices". (social worker)* | Limitation of evidence | Major limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | **Clear and consistent information:**<br>Social worker in 1 study also indicated that families need to have clear and consistent information to make the best decisions with and for their child.<br>The need for consistent information was also supported by parents interviewed in another 3 studies:<br>Parents and carers mentioned occasions when different professionals gave them conflicting advice and this was particularly disconcerting when parents were learning new complex medical procedures or when parents had to hand over the administering of medicines to their child.<br>*"The morning nurse said, 'He had a great day', then she leaned over and told the doctor, 'His "sats" went down.' I felt they weren't being honest with me. Just tell me! Sometimes I felt like they were telling me what they thought I wanted to hear."* | | | |

**Table 14: Summary of evidence (adapted GRADE-CER-Qual): Theme 2 – Condition specific information**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| **Sub-theme 1: information on the child's illness, diagnosis, prognosis, and cause of death** | | | | | |
| 9 studies<br>(Branchett 2012; Contro 2002; Hunt 2013; Jones 2006; Laakso 2001; Laakso 2002; Meert 2007; Monterosso 2007; Yuen 2012) | 8 studies used interviews and 1study used surveys | Many studies that mentioned the need for information on the child's condition mentioned them in relation to the aspects highlighted in the central theme.<br>In addition, 1 study that interviewed parents in the UK also highlighted that some explanation of the child's illness would be helpful for them:<br>*"Then the paediatrician phone one evening when my husband was out and said [the child] has got spinal muscular atrophy, if you want to look it up on the internet you can find* | Limitation of evidence | Major limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *out all about it. I remember thinking it was quite callous. It was shocking..."* | | | |
| **Sub-theme 2: information related to transition (information shared and correctly and consistently shared among involved healthcare organisations)** | | | | | |
| 2 studies (Branchett 2012; Hunt 2013) | 1 study used interviews and 1 study used surveys | In 2 studies from the UK, parents were interviewed and they reported that it would be helpful if information was correctly and consistently shared among the organisations involved in the transition of care:<br>Parents were particularly distressed about having to correct information or inform health professionals of previous events. They wanted to be able to rely on their care providers<br>*"Please record what happens in the delivery room and afterwards accurately. Having to correct notes or even worse, discover what they have been lost, causes untold misery and hurt..."*<br>*"Please inform all relevant people of what happened. One of the monitoring hospitals wasn't informed and we got chaser letters – very upsetting and totally unnecessary"* | Limitation of evidence | Major limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |
| **Sub-theme 3: information on treatment, autopsy, genetic risk, cause of death, research in the area** | | | | | |
| 2 studies (Hunt 2013; Meert 2007) | 2 studies used interviews | In 2 studies from the UK and USA where parents were interviewed, they reported that they would also find additional details helpful on the topics of this theme:<br>**Treatment**<br>*"I want to know about her medicines and the different beds they had her in and what role they played and what were they hoping to accomplish by putting her in those beds and with the machines that they used on her."*<br>Autopsy<br>*"We had issues about the autopsy which I would have liked to have explained a little bit more."* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | **Genetic risk**<br>*"Is it something genetic? Is it something to look for in my other children?"*<br><br>**Cause of death**<br>*"Nobody ever really told me what was wrong with him. It was some different things that they had said could be but nothing was a fact. I just want to know why he died."*<br><br>**The child's illness, research in the area**<br>*"The way we were given the diagnosis wasn't the best-it was in a normal clinical appointment. The doctor was looking at his watch at one point. I asked what sort of research was going on [to help] and the doctor said, 'don't worry about that, just love him' "* | | | |

**Table 15: Summary of evidence (adapted GRADE-CER-Qual): Theme 3 – Practical information**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: information about access to available services, or useful medical and paramedical services** | | | | | |
| 5 studies (Hunt 2013; James 1997; Monterosso 2007; Redmond 2003; Yuen 2012) | 4 studies used interviews and 1 study used surveys | In 5 studies from Ireland, the UK, USA, Australia and the Netherlands where parents and their children were interviewed, parents highlighted that having access to practical information that help them make use of available community resources or useful even essential medical/paramedical services available would be helpful, specifically: | Limitation of evidence | Major limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | **Voluntary services and support groups:**<br>In 1 study from the UK, parents commented that there appeared to be a lack of information about voluntary services and support groups.<br>Services families are officially entitled to:<br>In 1 study from the UK where parents were interviewed, it was highlighted that: the availability of information was very varied and easily accessible information was the exception rather than the rule. Parents commented that: *"It is a minefield finding out what you are entitled to. Most of the things we have found out by accident. There are all those services out there but they should make it more transparent – a directory or something"*<br><br>**Community resources:**<br>In 1 study from Canada where parents were interviewed, it was reported that parents found information they received about community resources very helpful.<br><br>**Central service point for children with severe intellectual disabilities:**<br>In 1 study conducted in Australia and included both parents and service providers, the following comments were made *"service providers consistently expressed concern with existing community-based disability services in terms of lack of palliative support for children and a central information point."*<br>The finding of need for a central information point was consistent with what was reported in another study from Ireland where parents of children born with severe intellectual disabilities were interviewed. Parents commented that: *"someone should be responsible for co-ordinating the services and calling to the person and saying A, B and C is available to you, and bringing the information."* | saturation | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *"a liaison officer or somewhere where all this information is gathered and when there is a child born with a disability or a particular syndrome, there is somebody responsible for passing on this information to the parents or family"*<br><br>**Information on practical medical and paramedical services that parents need for the care of their children born with severe intellectual disabilities (such as language therapists and physiotherapists):**<br>In the same study carried out in Ireland, many mothers also commented that the establishment of a central service, advocacy officer or even a telephone advice line whereby families can access the information which they need to avail of essential services would be helpful.<br><br>**Realistic options:**<br>One study conducted among parents in the UK commented that: *"there is not an equitable provision of community services across the UK. It is important that the options parents are offered are realistic. If, for example, a family wishes to take their child home to die the GP and Community Children's service would need to be able to offer support out of hours."*<br><br>**In different forms (such as oral, visual, and written forms)**<br>In 1 study conducted in the Netherlands, parents indicated that an important factor in the conversations was where the news was delivered and how, for example whether it involved the use of visual aids and written brochures. | | | |
| **Sub-theme 2: Social and family (information for other family members and friends)** | | | | | |
| 3 studies (deJong- | 2 studies used | In 2 studies from Canada and the USA, it was reported that | Limitation of | Major | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| Berg, 2006; Meert 2007; Nolbris 2005) | interviews and 1 study used surveys; | parents had information needs for their other family members. In another study from Sweden where healthy siblings of children who have died of cancer were interviewed it was reported that healthy siblings perceived that some relevant information or guidance would help them go through the process. It was highlighted that:<br><br>**Family members:**<br>Parents in 1 US study stated that they would like information in details they could give to other family members when asked:<br>*"After the fact, we had a lot of questions asked to us, by our own family. Everybody. We tried answering the best we could but when everything is going on it's really hard to communicate to the rest of the family all the details and everything."*<br>Parents in the same study also commented that they would like to know ways of how to help others who were experiencing the same:<br>*"My only thing now, is there anything I could do in terms of being there for other parents or helping them in that respect?"*<br><br>**Siblings:**<br>The need for information directly from the medical staff and information about the availability of social networks of other siblings who had the same experience.<br>Healthy siblings in 1 study from Sweden commented that they would have found it helpful if medical information about the life-limiting condition of their sisters/brothers could have been directly communicated to them by the medical staff. Furthermore they would have found it helpful to be informed about how to go through the process when their sisters/brothers were approaching the death, such as guidelines, literature, and contact with other siblings who had | evidence | limitations | |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | the same experiences.<br>In another study carried out in Canada where parents were interviewed they reported that they would like Information (in the form of stories) for younger children (siblings of the child with illness)<br>*"Providing stories for younger children (2-4 years)"* | | | |
| **Sub-theme 3: information on funeral arrangement** | | | | | |
| 3 studies<br>(DeJong-Berg 2006;<br>Laakso 2002;<br>Rini 2007) | 2 studies used interviews and 1 study used surveys; | **Information package for burial arrangement:**<br>In 2 studies, from Canada and the USA, parents who were interviewed said that information on burial and funeral arrangements would be helpful.<br>One parent suggested that the hospital have an information package available to parents, to help them with the process of burial for their child. Knowing what to expect, who to call for burial information and services, what costs to expect and how to make funeral plans was described as very important, and something that was not available.<br>This was also reported by parents in a study from Finland, where parents stated that they found information about purchasing a coffin, organising the funeral and buying funeral flowers helpful. | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |
| **Sub-theme 4: Information on bereavement support** | | | | | |
| 2 studies (deJong-Berg, 2006; Meert 2007) | 1 study used interviews and 1 study used surveys; | **Bereavement support: grief seminars and experts:**<br>In a study carried out in the USA where parents were interviewed, they stated that that they would have liked information on bereavement support:<br>*"Maybe talk to them about where you can get help ... I think it would be important if they think about telling you what you could do and where you could go."* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Sufficient | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | This was consistent with findings from another study carried out among parents in Canada, where they said that information on access to grief support and grief seminars and experts was helpful.<br>Parents found ….as well as grief seminars, to be useful aids in their grieving.<br>*"Try to channel people into [grief expert] seminars if this is possible"*<br><br>**Grief support in different forms:**<br>Parents in the same Canadian study also mentioned that they found books, music, poetry and websites useful aids in their grieving.<br><br>**Medical record of the child as grief support:**<br>In the same Canadian study, 1 parent said that access to the medical records of their deceased child would be a means of grief support:<br>*"I have felt the need to possess and someday read my daughter's medical records. While I cannot read them now, I know I will feel better knowing I have a copy of them when I am ready. I hope you will help me obtain them."* | | | |

**Table 16: Summary of evidence (adapted GRADE-CER-Qual): Theme 4 – Personalised information**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: information that meets individual needs** | | | | | |
| 5 studies | 4 studies used interviews and | **Developmentally appropriate information:** | Limitation of evidence | Minor limitations | MODERATE |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| (deJong-Berg 2006; Hsiao 2007; Hunt 2013; Jones 2006; Wocial 2000) | 1 study used surveys; | One study carried out among social workers in the USA commented that young people, even young children should be given developmentally appropriate information in the course of care so they could have autonomy, personal control over life and end of life decision-making issues.<br><br>**Psychologists:**<br>Parents in 1 study conducted in Canada stated that they found information from psychologists for bereavement support helpful.<br><br>**Readily available information when needed:**<br>Parents who were interviewed in a study from the USA stated that they wanted and appreciated information that was readily available to them. *"I want to be able to ask questions, because this was complicated, you know, this was hard...and several times, you know we had them call the specialist so we could ask them questions and stuff...They said, 'no, no problem, just give me a second and I will call them page them and have them come here and talk to you"*<br><br>**Spiritual perspective:**<br>Parents in 1 study, carried out in Canada, commented that:<br>*"Include more of a spiritual perspective/direct experiences should include more heart/soul rather than mind/intellectual anecdotal"*<br>**How to use equipment:**<br>Parents in 1 study from the UK commented that that they would like information on how to use equipment that a child or young person required. | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |
| **Sub-theme 2: Easy to comprehend information / complementary information** | | | | | |
| 2 studies | 2 studies used | One study from the USA reported that parents appreciated | Limitation of | Major | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| (Kavanaugh, 2010 Wocial, 2000) 2 Interview | interviews; | information that was easy to understand.<br>Parents mentioned how helpful it was to see x-rays or CAT scans of 'normal' babies next to their infant's test results.<br>*"Give that that knowledge you know, educate us so we can have some answers."*<br>*"We had to ask for his CAT scans...Obviously we are not medical students and a lot of the stuff may be you know a little tough to understand, but it can be broken down. We will comprehend it if you just lay it out there"*<br>Parents interviewed in another study from the USA also stated that they found it helpful when information was given in different forms/methods:<br>Several mothers reported that nurses gave them a tour of the NICU or booklets related to prematurity.<br><br>**Complementary information from multiple sources/ supporting staff such as nurses:**<br>One study from the USA reported that the majority of parents felt that nurses assisted them by explaining the care that the mother and infant were receiving or expected to receive, and providing information on the NICU or other resources. | evidence | limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

**Table 17: Summary of evidence (adapted GRADE-CER-Qual): Theme 5 – Active involvement information**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: information at appropriate level for informed and shared decision-making** | | | | | |
| 2 studies (Hsiao 2007; Hunt 2013) | 2 studies used interviews; | In 1 study from the USA where both parents and their children were interviewed, it was reported that parents thought there should be information provided to them and their child, as well as recognition and accommodation among | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | the HCPs about the desired level of child and parent involvement in communicating with physicians and participating in child's care.<br><br>**Non-patronising information and limited medical jargons:**<br>One study from the UK that interviewed parents commented that: there were numerous reports that hospital nurses and consultants addressed parents in either patronising ways, or conversely, they spoke in medical jargon which was difficult to parents to understand. | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |
| **Sub-theme 2: Information on professional opinions/guidance** | | | | | |
| 2 studies<br>(Jones 2006; Sullivan 2014) | 1 study used interviews and 1 study used surveys; | **Options/choices:**<br>In 1 study from the USA social workers commented that there should be information on symptom options and end of life issues/choices from the HCPs to families:<br>*"Families need open discussion of disease...progression, symptom options and end-of-life issues/choices..."*<br><br>**Recommendations and opinions from physicians:**<br>In 1 study from Australia parents commented that they found factual information in conjunction with the doctor's opinions or recommendation in relation to end of life issues and decision-making helpful:<br>*"[S]o we had a view and [name of the neurologist] gave us a view and were aware it was up to us..."* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |
| **Sub-theme 3: Information on anticipatory guidance** | | | | | |
| 3 studies (James 1997;<br>Hsiao 2007; | 2 studies used interviews and 1 study used | Parents who were interviewed in 3 studies from Canada, the UK and the USA commented that it was important for them to be kept informed about the child's prognosis, to prepare | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of | Coherent | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| Midson 2010) | surveys; | themselves and to know what to anticipate. In particular, parents highlighted the following:<br><br>**Change in treatment courses; physical changes of the child as their child approached death, early mention of death as a possibility, and adequate anticipatory guidance from the HCPs.**<br>*"I feel I needed more information about what to expect"*<br>*"There was lots of little things like that I found that weren't actually explained…a lot of trials and error of finding out things"* | findings | | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

## 4.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis.

## 4.7 Evidence statements

**A number of themes emerged from the interviews of parents and children, social workers, service providers, and healthy siblings. Although conceptualised as distinct and categorised individually, the central theme on timely, honest, and consistent information with subthemes of condition specific, practical, personalised, and active involvement information are interlinked and were perceived as important and helpful by those who had been involved in the end of life care as well as children, and young people and their parents. Timely, honest, accurate and consistent information (the central theme)**

Low quality evidence from 10 studies, carried out among parents and social workers using either interview or survey design, showed that parents would like to receive timely, honest, accurate and consistent information. This applied to throughout the course of end of life care ranging from diagnosis, transition and change of treatment, prognosis, and end of life issues/choices.

**Condition specific information**

Moderate to low quality evidence from 12 studies in which parents were interviewed, indicated that information sharing was very important. In addition to the condition specific information, when a transition of care occurred, parents would appreciate consistently and correctly shared information among involved healthcare organisations. They also described other information on aspects of care following death helpful, such as autopsy, genetic risk for family planning, and the cause of death.

**Practical information**

Moderate to low quality evidence from 12 studies taking account of the perspectives of parents, social workers and healthy siblings indicated that practical information on community services, voluntary groups, and medical and paramedical services that parents could make use of for the care of the child living with a life-limiting condition was important and helpful. Parents also expressed the view that information that could help them explain to other family members what happened, as well as information about how to help other people in similar situations, would be useful. Parents also expressed their information needs for funeral arrangements and about bereavement. For healthy siblings of the child with a life-limiting condition, it was highlighted that they would need information to support them to go through the process as well.

**Personalised information**

Moderate to very low quality evidence from 7 studies with populations of parents and social workers indicated that parents and their child benefitted from personalised information that met their individual needs. This included developmentally appropriate information for children and young people, as well as information from other sources such as psychologists, physicians, and spiritual perspective when needed.

**Active involvement information**

Moderate to very low quality evidence from 7 studies with populations of parents, children, and social workers indicated that parents found information that enabled them to be actively involved in informed decision-making for their child's end of life care issues and options helpful. It was also highlighted that parents found such information provided in conjunction with the physician's opinions and recommendations helpful, as well as anticipatory guidance that could help them to prepare for different aspects and phases of their child's condition.

## 4.8 Linking evidence to recommendations

### 4.8.1 Relative value placed on the themes considered

When developing the protocol for this review the Committee considered themes that were specific to end of life care as well as the general principles of good patient care that were developed for adults in the Patient Experience Guideline (CG138) they also considered other themes that emerged from this review. Some of the themes that the Committee considered to be important were about the mode of information provision (which would vary according to age) and the specific information that is needed by children and young people and their families during end of life care. The findings/themes that emerged or were derived from this review mirrored some of the general principles stated in CG138. However, the Committee considered that specific recommendations on information provision were particularly important and necessary in the context of end of life care for children and young people since this is often done inconsistently in current practice. Many of these were based on the central themes identified in the evidence review (timely, honest, accurate and consistent information) which were given particular weight in the discussion

### 4.8.2 Consideration of barriers and facilitators

Overall, the included evidence showed that families or carers found that timely, honest, accurate, and consistent information was perceived as the most important factor in information provision and influenced all other aspects of information provision. They also found the following helpful: personalised information; practical information; and information that facilitated active involvement in decision-making. The Committee thought the evidence was useful and relevant in terms of both general principles and details needed for information provision. Based on the main body of the evidence, the Committee made recommendations on both general principles for information provision, as well as specific guidance on helpful and supportive information provision during the end of life care for children and young people.

General principles of the recommendations were mainly informed by the central theme derived from the evidence. The Committee noted the importance of timely, honest, accurate and consistent information. The Committee acknowledged that there are many uncertainties in end of life care that impact on these concepts. There may be times when it is impossible to know what the 'most honest' information would be in light of the information that is available to clinicians (for example, related to the recognition of whether the child or young person is approaching the end of life). This also related to the other central concepts of accuracy, timeliness and consistency especially when the course of the condition may change. Reviewing information needs regularly was considered therefore particularly important. The Committee concluded that it was important not to make simple assumptions about what information would be needed or required by families. Instead, the information needs of the families or carers should be assessed and be individualised. As such, the information provided should be specific to the individual situation of the child or young person and their parents or carers (or those important to the child or young person), provided in a form that is easy to understand, consistent, and up-to-date.

Related to timely information, the Committee noted that it is important to consider the readiness of families to receive troubling information. When delivering information to families, their individual cultural, spiritual or religious background should always be considered, and taken into account when considering how to deliver information. The overarching principle that was discussed by the Committee was the tailoring of information to individual preferences and needs, for example with regard to how they would like to talk about end of life care, in what detail, and how they think their child should be told about their condition. Another important matter, noted by the Committee, might be the approach to addressing questions for the child or young person that they might not want to share or discuss with their parents. This would need consideration when establishing how they would like to discuss their life-limiting condition. The GC therefore made recommendations in this regard.

The Committee emphasised the importance of providing information in different modes according to the child and young person's preferences, which could include information via smartphones and other media, including social media. This was also highlighted in the research carried out for this guideline by children who have life-limiting conditions. Further, the Committee acknowledged that this information should be provided in a developmentally age appropriate way to the child or young person in end of life care. It was noted that information in different forms, mirrors guidance in the patients experience guideline (CG138) and applies both to adults and children. The Committee highlighted that these principles are particularly important in the context of end of life care for children and young people, where specific formats of information provision, (such as play and digital media), need to be tailored to the person's particular age and preferences. The Committee therefore thought that the information needs and preferences should also feature in the Advance Care Plan (a plan that provides information about the child or young person's care – see chapter 6).

To provide up-to-date information, the Committee thought that healthcare professionals should also identify any triggers that suggest the need for more information, or have discussions at the right time (such as when there are changes in the child or young person's condition), while at the same time being mindful about the emotional and psychological needs of the child and their parents or carers.

The Committee agreed that the information provided to the child or young person or their parents or carers should enable them to engage in the process. The information should therefore include their role and involvement in the care plan, who is on the multi-disciplinary team and their roles and responsibilities, the care options and choices available to the child and family (including options on specific treatments and place of care and death), and information on resources or support that could be provided to the family or carers.

The Committee discussed that although there was a lack of published evidence with regard to care of the body after death in this review, they thought it was important that healthcare professionals should provide families or carers with clear information about this, as well as the legal issues related to death. The group agreed that healthcare professionals should also provide information on registration of the death, funeral arrangements, post mortem examination (when needed) and bereavement support. The Committee particularly noted that such information should be locally relevant, based on what was available in the region.

#### 4.8.2.1 Barriers and facilitators highlighted in the TFSL report

The focus group work undertaken for this review highlighted many of the barriers and facilitators to effective information provision that have been discussed in the preceding section. The children and young people with life-limiting conditions emphasised that their needs for information varied, both between people and also for the same person over time and with changes in maturity, as they became more involved in decisions relating to their care, and as the course of their condition altered. Young people described a range of negative emotions when healthcare practitioners did not provide them with information in an appropriate way, or did not take the time to understand their needs. They recognised the

value of having someone who understood their situation to talk things through with, but felt that this was not always available. Access to reliable sources of information (particularly online) was felt to be helpful, although the variable quality of this, and the limited applicability to an individual's specific situation were acknowledged as barriers. Participants also described some information available on line as being scary or inaccurate, and made efforts to avoid these.

### 4.8.3 Economic considerations

A pre-requisite of good medical care is information provision and the recommendations made in this guideline reflect what the Committee considered to be good practice. Many of the recommendations address good principles for information provision (for example, being honest) and don't have a resource implication as such. The Committee considered that overall their recommendations regarding information would have a minimal resource impact and that they would promote the cost-effective use of NHS resources.

### 4.8.4 Quality of evidence

Moderate to very low quality evidence was found in this review. The main reasons leading to downgrading of the evidence shared by the majority of studies included:

- self-selection bias and recruitment bias: in many studies only about half or less than half of the respondents contacted consented to be interviewed. Subjects who chose to participate may be more open to communicating with unfamiliar people than those who refused to be contacted. On the other hand, in some studies participants were selected by the physicians who have provided care to the child, and those who were eliminated from participation may be the group that had different views and needs for information.
- lack of critical review of the researcher's role in sample recruitment, data collection or data analysis process: few studies clearly reported the relationship between researchers, interviewers and the respondents, whether the researchers had a pre-existing understanding about the topic or the possible influence of that in data collection and the analytical process.
- lack of verification of findings: few studies verified their findings with participants or external sources, nor reported the reason why verification was not necessary or applicable. Finally many studies did not report in detail how findings/themes were derived or emerged from the data in their research, although word limits in journal publications might be a reason for that.
- Saturation: Saturation either in terms of data collection or data analysis was not achieved or difficult to assess in many included studies, as well as the themes that emerged from those studies. This was because information provision was not the primary focus in many included studies. However, when considering the evidence as a whole, saturation was achieved on some meta-synthesised themes.
- Applicability: findings from the majority of included studies in this review were considered to be applicable to the UK setting because of the direct relevance of their participants, contexts, and the topics explored.

A variety of information and information types experienced or perceived as helpful/unhelpful were reported across studies, however due to the uncertainty in data saturation or sufficiency on many findings in this review, the Committee agreed that the evidence should be interpreted with caution.

### 4.8.5 Other considerations

The Committee noted the links between information and communication, and advance planning, but it was agreed that the recommendations informed by this review would focus on the content and form of information provision itself.

The Committee discussed information consistency, and agreed that a mechanism should be put in place where information could be correctly and consistently shared among involved healthcare organisations when transition of care occurred.

To keep families or carers informed about what is happening, the Committee considered that an interpreter should be engaged where needed, to facilitate the process and ensure information is delivered. However, the needs of the interpreter should also be noted and support provided if required.

The report of the research carried out specifically for this guideline highlighted further themes that directly addressed this topic. The views provided in this report were given particular weight by the Committee, and particularly related to how and what type of information children and young people wanted to receive.

The Committee discussed whether they wanted to prioritise this topic for a research recommendation, but they concluded that the combination of the evidence (including the focus group report), their experience and their expertise was sufficient to base the recommendations on.

#### 4.8.5.1 Other considerations related to the TFSL focus group findings

Children and young people who participated in the focus groups identified a number of sources of information that they found helpful.

Talking things through with someone who really understands was identified as important to many participants, and there were varied sources for this, including the experience of other young people with similar conditions or who had experienced the same treatments. Some participants in the focus groups accessed online forums to link up with other young people. Such contacts were felt to be helpful both in sharing experiences, but also in decision-making by the young person. Consideration should be given as to how this sort of interaction can be facilitated and supported.

The focus groups also identified clinicians, such as their consultant, as a key source of information, although there were many other commonly reported healthcare practitioners who provided a valued source of information. Parents were also an important source of information and advice, but the young people themselves also knew a lot as they had ‘the lived experience’ of their illness or condition. The internet was recognised as an important source of information, although the variable quality of this source was noted. Focus group participants described their efforts to avoid scary or inaccurate information online about their condition. For most participants, asking their consultant or other trusted professionals was preferred to using the internet for medical advice and information, although getting their advice at the weekend was sometimes difficult.

Preferences for how much information children and young people would like to receive varied. Some could be overwhelmed if they received too much information, while others required all the available information in order to reach a decision.

Having to repeatedly explain about their condition or care-needs to different people was also frustrating for young people, and asking for help or having to explain how care should be given sometimes made them feel embarrassed, scared and nervous.

Information around the time of transition between care settings or transition to adult services was identified as a problematic area in the focus groups. For young people who had already transitioned to adult services, lacking a single point of contact (for example, a consultant or specialist clinic) was described as a loss.

### 4.8.6 Key conclusions

The Committee noted that the evidence indicated that the 5 main themes identified were interrelated and linked throughout end of life care, and would also inform the general principles that can be translated into practice. The Committee agreed with the evidence that, currently, information around end-of-life care is often not provided in a timely and consistent manner. However, it was also discussed that there is a wide range of circumstances that makes the interpretation of what is considered 'timely' to be difficult. Condition specific and practical information are also important aspects of information provision. Cultural, spiritual, religious and ethnic backgrounds were highlighted as important factors that influenced the type of information that may be needed. The views of children who took part in the focus group research carried out for this guideline, provided important information which gave a strong rationale for the recommendations.

## 4.9 Recommendations

1. **Be aware that most children and young people with life-limiting conditions and their parents or carers want to be fully informed about the condition and its management, and they value information that is:**
    - specific to the child's or young person's individual circumstances
    - clearly explained and understandable
    - consistent
    - up-to-date
    - provided orally and in writing.

2. **Be aware that some children and young people and parents or carers may be anxious about receiving information about their condition.**

3. **Ask how children and young people and their parents or carers would like to discuss the life-limiting condition. For example:**
    - Ask which topics they feel are important and would particularly want information on
    - Ask whether there are topics they don't want detailed information on, and discuss their concerns
    - If appropriate ask parents or carers whether they think their child understands their condition and its management, and which professional their child would like to talk to about it.
    - If appropriate, ask parents or carers what they think their child should be told about their condition
    - Discuss with the child or young person and their parents or carers their right to confidentiality and how information about their condition will be shared
    - Review these issues with them regularly, because their feelings and need for information may change over time or if their circumstances change.

4. **When talking to children and young people and their parents or carers:**
    - be sensitive, honest and realistic
    - give reassurance when appropriate
    - discuss any uncertainties about the condition and treatment.

5. **Be alert for signs or situations that the child or young person or their parents or carers need more information or discussions, for example if:**
    - they are more anxious or concerned
    - the child or young person's condition deteriorates
    - a significant change to the treatment plan is needed.

6. **Provide children and young people and their parents and carers with the information they need on:**
    - their role and participation in Advance Care Planning (see 6.1)
    - the membership of their multidisciplinary team and the responsibilities of each professional (see 7.1)
    - the care options available to them, including specific treatments, preferred place of care and place of death (see 6.2)
    - any relevant resources or support available to them.

# 5 Communication

## 5.1 Review question

**What are the barriers and facilitators to effective communication between the child or young person, the family or carer and the healthcare professionals about the life-limiting condition and likelihood of imminent death?**

## 5.2 Introduction

Effective communication depends on sensitive and compassionate discussions between the child or young person, their parents or carers and the healthcare team. Although this is usually done in a supportive and empathetic way, many children and their families have been frustrated by ineffective communication about end of life care. Communication between healthcare professionals and parents about their critically ill children involves a number of challenges. For healthcare professionals these include for instance a reluctance to relay bad news, the uncertainty about prognosis and the wish to continue to provide hope. These issues may lead to delays in planning children and young people's end of life care until very late in the child's illness.

Parents often describe how stressful it is to receive contradictory information from different healthcare professionals, and stress the importance of feeling that they have received honest and complete information from the healthcare team.

Children also benefit from effective communication, as providing information and actively addressing their concerns can reduce anxiety, enhance the cooperation of the child, and lighten the burden of secrecy that may surround them. If information is withheld from children then this runs the risk of exacerbating their distress and fears.

## 5.3 Description of clinical evidence

The aim of this review was to identify themes in the experiences, opinions and attitudes of the child or young person with a life-limiting condition and their parents or carers, on the factors that encourage or prevent good communication. In particular we wanted to explore communication between children, their parents and carers and healthcare professionals when talking about the life-limiting condition or the likelihood that they are entering the last days of life.

Qualitative studies were selected for inclusion for this review. We looked for studies that collected data using qualitative methods (such as semi-structured interviews, focus groups, and surveys with open-ended questions and analysis of documented materials) and analysed data qualitatively (including thematic analysis, descriptive phenomenology, content analysis and so on). Survey studies restricted to reporting descriptive data that were analysed quantitatively were excluded.

Given the nature of qualitative reviews, findings/themes were summarised from the literature and were not restricted to those identified as likely themes by the Committee. Some of the anticipated themes were: healthcare professionals' experience and specialist training in communication skills; empathy and rapport; cultural and religious considerations; timing (when to initiate); resources (time spent with individuals and place of communication, that is, privacy in hospital); families acceptance of prognosis; translation services; different methods of communication (tools to facilitate, that is, written, online, play).

A total of 28 studies were identified for inclusion in this review. The majority (25 out of 28) of them focused on families/carers or/and healthcare professionals' perspectives, only 3 studies

involved children and young people living with life-limiting conditions, or their healthy siblings. Specifically:

- 16 studies (Branchett 2012; Caeymaex 2011; Contro 2002; Davies 2002; Davies 2003; Davies 2010; Gordon 2009; Hendricks-Ferguson 2007; Lundqvist 2002; Meert 2008; Meyer 2006; Midson 2010; Robert 2012; Weidner 2011; Wood 2010; Woolley 1989) focused on the perspectives of families/carers whose child had deceased due to a life-limiting condition or who were caring for a child with life-limiting condition
- 7 studies (Baverstock 2008; Contro 2012; de Sa Franca 2013; Forbes 2008; Pearson 2013; Price 2013; Stenekes 2014) focused on healthcare professionals (including consultants, physicians, and nurses) who were involved in end of life care and palliative care of children and young people living with life-limiting conditions
- 2 studies (Byrne 2011; Contro 2004) involved both parents and healthcare professionals
- 1 study (Steele 2013) interviewed parents and healthy siblings whose child or sister/brother had life-limiting conditions
- 1 study (Hsiao 2007) involved both the parent and their child living with life-limiting conditions in pairs
- 1 study (Gaab 2013) interviewed children and young people living with life-limiting conditions and their healthy siblings.

The majority of included studies collected data by semi-structured interviews or focus groups, 4 studies (Baverstock 2008; Branchett 2012; Forbes 2008; Meyer 2006) collected data by open-ended questions in survey questionnaires; a couple of studies collected data by reviewing archived materials (Byrne 2011) or diary writing and recording (Gaab 2013). The most common data analysis method employed across studies was thematic analysis and content analysis.

With regard to the setting of studies:

- 8 studies (Baverstock 2008; Branchett 2012; Davies 2003; Midson 2010; Pearson 2013; Price 2013; Wood 2010; Woolley 1989) were conducted in the UK
- 13 in the USA (Byrne 2011; Contro 2012; Contro 2002; Contro 2004; Davies 2002; Davies 2010; Gordon 2009; Hendricks-Ferguson 2007; Hsiao 2007; Meert 2008; Meyer 2006; Robert 2012; Weidner 2011);
- One in both the USA and Canada (Steele 2013 );
- 1 each from Australia (Forbes 2008), Brazil (de Sa Franca 2013), Canada (Stenekes 2014), France (Caeymaex 2011), New Zealand (Gaab 2013) and Sweden (Lundqvist 2002).

Evidence on all themes considered important by the Committee were identified. Because information provision and communication are topics that interweave, some top level (main) categories/themes identified for the communication provision review were also identified for the information review. However, a number of further themes or sub-themes that were particularly relevant to the aspects of communication and interpersonal interaction were identified. This included factors involved in the interpersonal interaction between families and healthcare professionals, such as empathy, sensitivity, trust; and emotional factors on the part of both parents and healthcare professionals.

Subsequently, a combined theme-map incorporating themes that are relevant to both information provision and communication was developed. In this map specific themes and sub-themes that featured specifically in the communication review were added to the overall structure.

To include the views of children and young people with life-limiting conditions and direct experience of the health service in the UK, a focus group was commissioned specifically for this guideline. A description of how this research contributed to the recommendations has

been added to the Linking Evidence to Recommendation section of this chapter (see section 5.8 and Appendix L)

A brief description of the studies is provided in Table 18. Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix G and in the GRADE profiles. The TFSL focus group report can be found in Appendix L. For presentation of findings, a theme map was generated according to the themes emerged from studies (Figure 5). The mapping part of the review was drafted by 1 researcher working on the guideline but the final framework of themes was further shaped and when necessary re-classified through discussions with at least 1 other researcher. Due to the qualitative nature of these studies evidence is summarised in adapted GRADE-CERQual tables within the evidence report. Therefore no separate Appendix is provided for this.

## 5.4 Summary of included studies

A summary of the studies that were included in this review are presented in Table 18

**Table 18: Summary of included studies**

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| Interviews/focus-groups | | | | |
| Caeymaex 2011 | Interviews | N=80 families with 86 individual parents<br>France | To explore parents' experience of the end of life decision-making process for their child in the neonatal intensive care unit | • limited response rate (37%) to participate<br>• whether data saturation in terms of collection or analysis was achieved was not clearly reported<br>• researchers' role in and influence on the analytic process was not critically reviewed |
| Contro 2012 | Interviews | N=60 Healthcare professional (HCP) staff members from multiple disciplines<br>USA | To examine the current state of bereavement care at a university-based children's hospital from the perspective of the interdisciplinary staff. | • whether data saturation in terms of collection or analysis was achieved was not clearly reported;<br>• researchers' role in and influences in the analytical process was not critically reviewed<br>• findings were verified with 1/3 of participants |
| Contro 2002 | Interviews | N= 68 parents representing 44 families<br>USA | To obtain personal accounts of families' experiences to learn ways to improve care for paediatric patients and their families. | • low response rate (44 out of 156 families contacted consented to participate);<br>• unclear whether data saturation in terms of collection or analysis was achieved<br>• researchers' role in and influences in the analytical process not critically reviewed |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| Davies 2002 | Interviews | Parents (participant number not reported)<br>USA | To provide insights into the meaning of optimal paediatric end of life care. | • No details on sample selection, data collection, data analysis methods reported<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Davies 2010 | Interviews | N=36 parents from 28 families<br>USA | To learn about experiences of Mexican American and Chinese American families who require paediatric palliative care. Parents' perceptions of information sharing by healthcare providers during their child's hospitalisations and at their child's death. | • unclear whether data saturation in terms of collection or analysis was achieved<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Davies 2003 | Interviews | N=23 married couples and 7 single parents<br>UK | To explore parents' experiences of care by paediatricians in the time leading up to and including diagnostic disclosure of a life-limiting condition in their child. | • participants were identified by professional colleagues of the authors and invited to take part by letter<br>• data collection process was not clearly reported (only reported that an in-depth interview was conducted)<br>• no discussion on whether saturation had been reached for any of the themes reported |
| de Sa Franca 2013 | Interviews | N= 10 nurses<br>Brazil | To investigate and analyse communication in palliative care in paediatric oncology from the viewpoint of nurses, based on Humanistic Nursing Theory. | • small sample size<br>• unclear whether data saturation in terms of collection or analysis was achieved<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Gordon 2009 | Interviews | N=51 parents<br>USA | To examine parents' perceptions of good and poor medical communication with the team who cared for their child prior to his or her death in the PICU. | • sample selection was not clearly reported<br>• unclear whether data saturation in terms of collection or analysis was achieved<br>• researchers' role in and influences in the analytical process was not critically reviewed |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| Hendricks-Ferguson 2007 | Interviews | N=28 parents<br>USA | To examine parents' perspectives of: 1) the timing and method used by healthcare professionals to introduce end of life options for their child, and 2) what their preference would have been regarding the selected time and method to introduce end of life options | • convenience sample, participants were selected by hospital staff<br>• no discussion on whether data saturation had been reached in terms of collection and analysis;<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Hsiao 2007 | Interviews | N= 20 parent and child pairs<br>USA | To identify the aspects of physician communication that children with life-limiting illnesses and their parents perceived to be facilitative or obstructive in paediatric palliative care. | • response rate for invited subjects for this study was 57%, participants recruited by HCPs<br>• non-English speakers excluded<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Lundqvist 2002 | Interviews | N=20 mothers<br>Sweden | To examine and illuminate mothers' experiences and perceptions of the care given to them at neonatal clinics while facing the threat and the reality of losing their baby. | • small sample size;<br>• data analysis was not clearly reported<br>• whether saturation was achieved in terms of collection or analysis was not reported<br>• findings were verified with mothers |
| Meert 2008 | Interviews | N=58 parents of 48 children who died in the PICU 3-12 months before the study<br>USA | To describe parents' perceptions of their conversations with physicians regarding their child's terminal illness and death in the paediatric intensive care unit (PICU). | • low response rate (30%);<br>• no discussion on whether data saturation had been reached in terms of collection and analysis;<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Midson 2006 | Interviews | N=55 parents who experienced the death of a child under age 17 between 12 and 18 months ago.<br>UK | To explore the experiences of parents within 1 tertiary centre, and the challenges that still lay ahead in changing the barriers, attitudes, and culture that impeded some aspects of end of life care. | • interviews were conducted by phone, home visits or in a hospital room<br>• unclear about the relationship between the researchers and interviewees; researchers' roles and pre-knowledge and their influences on data collection and analysis was not critically reviewed |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| Pearson 2013 | Interviews | N= 7 nurses out of 12 invited across 4 sites contacted with the assistance of ward managers<br>UK | To understand children's cancer nurses experiences of providing palliative care in the acute hospital setting | • data saturation during collection was achieved<br>• researchers critically reviewed their roles and influences in the process |
| Price 2013 | Focus groups | N= 35 healthcare professionals<br>UK | To investigate health and social care professionals' perspectives on developing services for children with life-limiting conditions at the end of life using issues identified by bereaved parents as priorities. | • the relationship between the researcher and the respondents was clearly reported (researcher had no managerial or other responsibility over participants)<br>• no discussion on whether saturation had been reached for any of the themes reported<br>• researchers did not critically review their roles and influences in the process |
| Robert 2012 | Focus groups | N=14 parents (whose children were age 10 years and older at the time of death)<br>USA | To describe and begin to understand the experience of bereaved parents whose deceased child had received paediatric oncology services at a tertiary comprehensive cancer centre. | • low response rate (9 families with 14 parents out of 47 families contacted consented to participate)<br>• focus group interview guide was developed based on a literature review<br>• researchers' roles and potential influences in the analytical process was not critically reviewed; |
| Steele 2013 | Interviews | N= 99 family members of CYP who died of cancer, including 36 mothers, 24 fathers, and 39 siblings from 40 families<br>USA and Canada | To determine how to improve care for families by obtaining their advice to healthcare providers and researchers after a child's death from cancer. | • data saturation was achieved for the analysis<br>• researchers' roles and potential influences in the analytical process was not critically reviewed |
| Stenekes 2014 | Focus groups and interviews | N= 29 HCPs<br>Canada | To examine the views of HCPs involved in perinatal palliative care in 3 tertiary care hospitals | • low response rate (29 HCPs out of 850 contacted consented to take part)<br>• Interviews were conducted over the phone |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| | | | in Canada. | • no discussion on whether saturation in terms of collection or analysis was achieved<br>• researchers' roles and potential influences in the analytical process was not critically reviewed |
| Weidner 2011 | Focus groups and interviews | N=29 parents<br>USA | To identify and define the dimensions of paediatric end of life care that are important to parents. | • low response rate (22%)<br>• how themes were derived was not clearly reported<br>• whether saturation in terms of data collection or analysis achieved was not clearly reported |
| Wood 2010 | Focus groups and interviews | N= 30 families<br>UK | To collect qualitative experiential data and use it to identify major themes and what events – in health, social and education domains – were considered to be 'milestones' by families and professionals caring for children with life-limiting conditions | • low response rate (40%)<br>• no discussion on whether saturation had been reached for the relevant themes reported<br>• researchers' roles and potential influences in the analytical process not critically reviewed |
| Woolley 1989 | Interviews | N=45 families<br>UK | To explore parents' experiences of the way in which they were told the diagnosis of life-limiting conditions of their child | • data analysis methods not reported<br>• no discussion on whether saturation had been reached for the relevant themes reported<br>• researchers' roles and potential influences in the analytical process was not critically reviewed |
| Surveys | | | | |
| Baverstock 2008 | Survey with open-ended questions | N=61 tertiary paediatric consultants<br>UK | To describe how paediatric consultants report dealing with child and neonatal deaths as part of their daily work. | • data analysis methods not clearly reported<br>• whether data saturation in terms of collection or analysis achieved was not reported<br>• findings not verified |
| Branchet 2012 | Survey with open-ended questions | N=57 parents who had lost a child in the neonatal period<br>UK | To determine what parents had actually experienced relating to neonatal palliative and end of life care and determine how this knowledge could be used to improve | • data were collected by a few simple open-ended questions initially posted on a parent's website. No guide was used to design and collect data. The study was undertaken by 1 researcher as part of scoping exercise |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| | | | experiences for families in future. | within a bigger project therefore may lack some of the formal research rigour. |
| Forbes 2008 | Survey with open-ended questions | N=162 respondents<br>Australia | To learn about doctor's current attitudes and practices relating to discussions concerning withdrawing or withholding life sustaining equipment (WWLSMT) in the paediatric setting | • low response rate (42%);<br>• data analysis not reported;<br>• whether data saturation in terms of collection or analysis achieved was not reported<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Meyer 2006 | Survey with open-ended questions | N=56 parents<br>USA | To present the parents' own words about what was most and least helpful at their child's end of life, ways to enhance communication, and advice about how to improve care. | • Limited response rate (58%)<br>• data analysis not reported<br>• whether data saturation in terms of collection or analysis achieved was not reported<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| **Survey and interviews** | | | | |
| Contro 2004 | HCP staff survey; and family interviews | N=446 HCPs + 68 family members<br>USA | To obtain personal accounts of HCPs and families' experiences to learn ways to improve care for paediatric patients and their families. | • data analysis methods not clearly reported, how themes were derived not clear<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| **Analysis of documented materials** | | | | |
| Byrne 2011 | Phenomenologic analysis of initial consults | N=43 initial consults led by 32 different physicians of paediatric advanced care<br>USA | To develop awareness of the consult reality from family, referring, and provider participant perspectives. | • a convenience sample was used<br>• whether data saturation in terms of collection or analysis was achieved was not clearly reported<br>• researchers' role in and influences in the analytical process not critically reviewed |
| **Diaries in writing or recorded** | | | | |
| Gaab 2013 | Diaries (in writing or recorded) | N= 16 young people (including 7 patients, 3 brothers, and 6 sisters from 8 families)<br>New Zealand | To describe self-identified factors that affect 9-to-18-year-old paediatric palliative care (PPC) patients and their siblings during the process of receiving paediatric palliative | • no discussion on whether saturation had been reached for any of the themes reported<br>• researchers' role in and influences in the analytical process was not critically reviewed |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| | | | care. | |

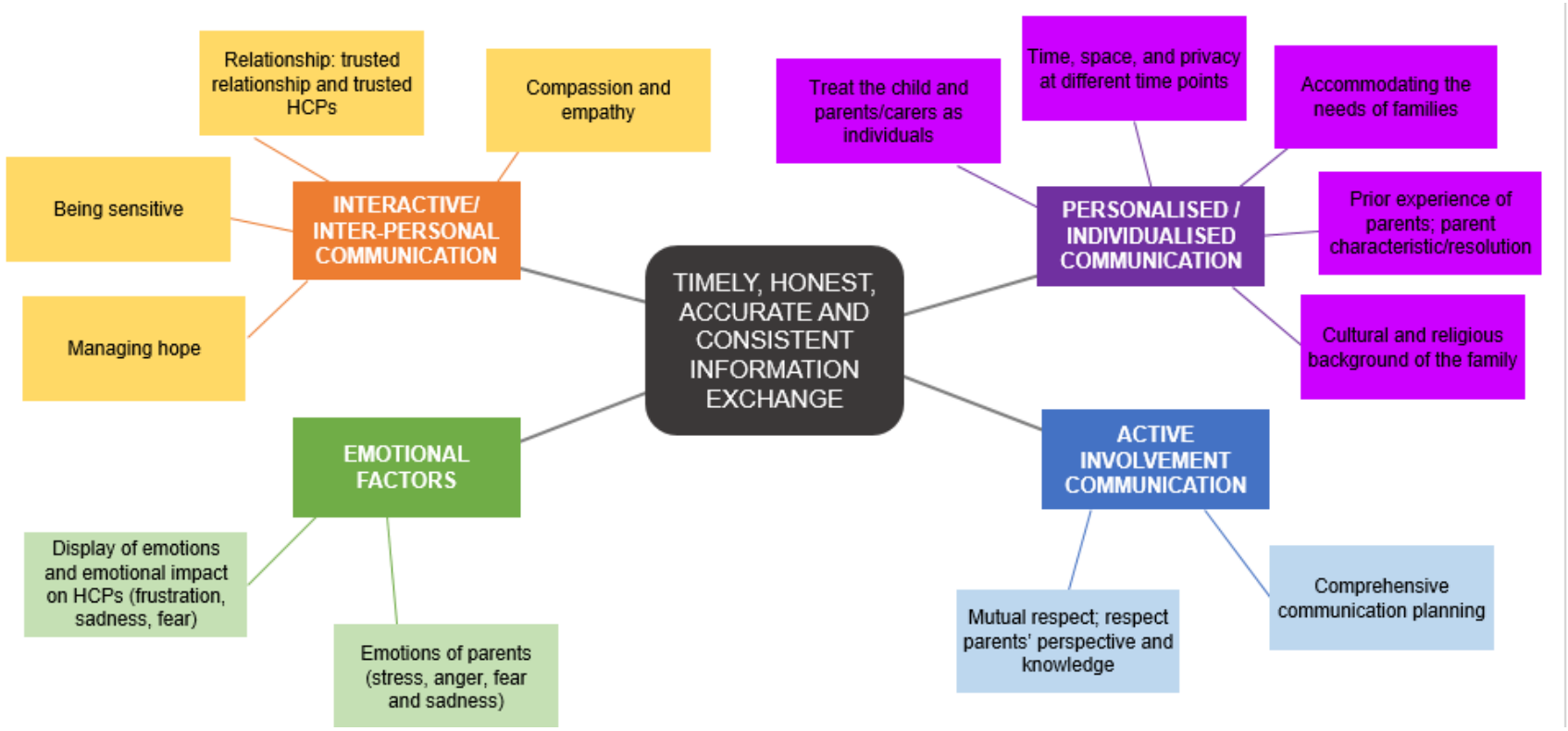
Five categories/themes related to the communication between families and Healthcare professionals that could act as either barriers or facilitators for effective communication emerged or were derived from included studies. The central theme that came from the review highlighted the need for timely, honest, accurate and consistent information exchange, which was a feature throughout all subthemes. Most themes were consistent with those that emerged from the information provision review. Additionally 4 further categories and themes emerged which highlighted specific features for effective communication:

- Personalised/individualised communication
- Inter-personal/interactive communication
- Emotional factors
- Active involvement in communication.

# 5.5 Clinical evidence

## 5.5.1 Theme map

**Figure 5: At the centre of the map is the overarching theme, which was mentioned as part of most of the other themes and subthemes, and relevant for both information provision and communication (details also reported in information provision evidence report)**



## 5.5.21 Evidence Summary

**Table 19: Summary of evidence (adapted GRADE-CERQual: Theme 1 – Personalised/individualised communication**

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| **Subtheme 1: Treat the child and parents/carers as individuals, incorporating their family context** | | | | | |
| 8 studies (Caeymaex 2011; Davies 2002; Davies 2003; Hsiao 2007; Hendricks-Ferguson 2007; Robert 2012; Steele 2003; Weidner 2001) | 8 studies used interviews; | 8 studies conducted in different settings and countries (including France, the UK, the USA) from a parental, and pairs of parents and children living with LLC, reported that parents and children appreciated doctors taking the time to get to know and treat the child as individuals, and unique individuals living with their families.<br><br>**View the child as an individual not as an illness:**<br>A father stated, *"You don't want to think that your child is just a patient at a hospital. Treat them more as an individual rather than just a patient on a clipboard."*<br><br>This is supported by findings in another 2 studies where parents stated that every child was unique, as was their diagnosis, and both required creative and personalised solutions and a dynamic work environment:<br>*"The feeling that you are there with your daughter and not just with somebody with an interesting malformation or some new science... but this is just this kid that you really love" (parent)*<br>*"They treated his body part or whatever it was at that time and he want a whole child"* (parent)<br>*"the less rules, the better. What was perfect for [one patient] was totally different for [our son]….Ask the kid."*<br><br>Communication based on the assessment of individual needs of families and the child:<br>In addition, 1 study conducted in the UK also highlighted the importance of communicating and assessing the child's and | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | families' desire on an individual basis, where researchers commented that:<br>*[It's appreciated that] staff gently ask as to the information parents might want; staff assess parents' desire on an individual basis to talk about sensitive topics, such as the child's impending death, funeral plans, and bereavement issues.*<br><br>**Interpersonal dialogue/communication incorporating individual family context:**<br>This was raised by parents in 2 studies conducted in France and the USA as an important positive facilitator to good communication (Caeymaex 2011, Davies 2003).<br>*"He explained that it was …I remember he said something: this isn't reasonable" (parent). The family context and the realities of life had to be taken into account. "The doctor left me the choice. He explained to me the risks of these choices. He told me, you already have a three-year-old daughter. He stayed in the context of our little family: for the child, for me, for my family. If something happens to you, who will take care of him? Very concrete questions"*<br><br>**Explaining the situation/consequences to children and young people according to their choices:**<br>In 1 study (Hendricks-Ferguson 2007), parents appreciated that HCPs spent time to explain the consequences of receiving end of life care at home to their teenagers. A mother shared her memory of the ICU when her 17-year-old daughter wanted to stop treatment and go home:<br>*"The healthcare team spent time to make sure my daughter understood the consequences of her decision to receive EOL care at home."* | | | |
| **Sub-theme 2: Personalised communication about diagnosis, death and around the time of death (time, space, and privacy at different time** | | | | | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| **points)** | | | | | |
| 4 studies (Baverstock 2008; Davies 2002; Meyer 2006; Woolley 1989) | 3 studies used interviews and 1 study used surveys; | In 4 studies where HCPs and parents were interviewed they reported that dedicated time, space, and pacing of delivering information were important at different time points, particularly when the diagnosis was delivered, and around the time of imminent death.<br><br>**Time, pacing and reaction of parents when the diagnosis is delivered:**<br>***Sufficient time to react:***<br>In 1 study (Woolley 1989) about imparting diagnosis of LLCs in children, interviewed parents stated that their immediate shocked reactions affected their ability to hear and take in what was being said. Many reported that it was essential to be given sufficient time. It was perceived to be especially helpful when doctors asked them what they had understood and then repeated and clarified points in different ways.<br>Parents in the same study also cited the doctors' ability to sit with them when they are upset or angry (not necessarily responding directly to this). Showing their feeling made them contributed to a perception of being understood and having a closer relationship with the person caring for their child.<br><br>**Privacy: in private, uninterrupted, unhurried, both parents being present**<br>Both parents and HCPs interviewed in the 4 studies stressed the importance of privacy, as well as dedicated space and time in communication with families and the child.<br>Parents in the previously mentioned study (Woolley 1989) commented that they appreciated being given time together in private to take the news in and to share their feelings.<br><br>***Time with the child and privacy at the time of death***: | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | In another study where parents whose children passed away (Davies 2002) were interviewed, they stated that at the time of death, they wanted staff to allow them as much time as they need with the child, without being rushed or criticised for *"taking so long"* and they appreciated *"privacy"*.<br>Privacy was highly valued during the final hours and days together. Some parents in another study (Meert 2008) described *"quiet time"* as moments of peacefulness when they could *"reach out and touch him"* or *"go and see him at all hours of the night."* For many, there was a wish to focus intensely on the time to *"say goodbye"*.<br>*"The nurse who took care of my infant was so kind and compassionate. She stayed in the room with us but also gave us our space, which was really good. They let us take as much time as we needed to say good-bye." "[Being able] to sleep with my son one final time." (parents)*<br><br>The same theme emerged from 3 other studies (Baverstock 2008; Meyer 2006; Stenekes 2014) where healthcare professionals were interviewed they commented the importance of *"right environment"* (time, privacy, separate room, tea, and so on) for communication with families around the time of the child's death.<br>*"And we often had a real lack of privacy. ... But then we would be sometimes in a room where in the next room you would hear a baby being born and the baby's crying, and this mother knows her baby is not going to cry. It was very hard and it was kind of like, you know what, we have KDPR there, the rooms are very private...it just makes so much sense." (HCPs)"* | | | |
| **Sub-theme 3: Accommodating needs of families and children/young people depending on their situations** | | | | | |
| 7 studies (Davies 2003; Gabb 2013; | 6 studies used interviews and | 7 studies reported on accommodating needs of families depending on their individual situations. This acted as | Limitation of evidence | Minor limitations | MODERATE |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| Hsiao 2007; Lundqvist 2002; Robert 2012; Stenekes 2014; Steele 2013) | 1 study used diaries in writing or recorded; | facilitator for effective communication. These studies incorporated the opinions of parents, CYP living with life-limiting conditions, and HCPs.<br><br>**Sensitive to parents' needs as a parent and a family:**<br>In 2 studies (Davies 2003, Lundqvist 2002) parents stated that they appreciated paediatricians who listened to them, took their concerns seriously and were able to respond with sensitivity, human sympathy and understanding.<br>*"Our baby wouldn't survive…. Often they [the babies] would fall asleep with the mother or father [the physician had said]. My first reaction was, I can't go through with this. But then, I thought he would recognize my heartbeats. Of course he will be in my arms…. We had to give him a name. We didn't want to baptize…. I had not wanted my baby to have a borrowed christening robe [crying]. The nurse had prepared a small bunch of flowers that we have dried and now keep in a book. She hadn't lit the candles, but we had candles. They had taken away almost all [the equipment from the baby's body]. My husband and I named him, and then we withdrew the ventilator. First the nurse put him beside his twin sister [to say good-bye] and then directly in my arms. There he quickly fell asleep. After a while we felt that we had said good-bye to him. Later on we heard that the reflective breathing had gone on for a long while, and the nurse had had him in her arms, which was so good to hear [crying]. Then, the day after they asked if we wanted to look at him again." (Mother)*<br><br>**Needs of the child living with life-limiting condition:**<br>*Personal and social concerns of the child:*<br>Children and young people interviewed in 1 study (Hsiao 2007) commented that they appreciated it if physicians taking time to inquire about their personal or social concerns in addition to treating physical symptoms. | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *Needs of the child/young people talking about death, concerns about mortality and feeling in control (based on individual needs):*<br>Parents interviewed in 1 study (Robert 2012) described the child's ambivalence to talk about death and the importance of child having control regarding end of life discussions:<br>*"Our daughter wanted to talk about [terminal cancer], then didn't...[A doctor asked her], 'What are you afraid of?...Dying?..Why?' That made it easier for her to talk to us,...to be in control...she could plan for her funeral."*<br>This was echoed by findings from another study (Gabb 2013) carried out among CYP receiving palliative care, where young people stated their concerns about mortality:<br>*"The thing I worry most is the, um, dying bit. That's what I don't like. The doctors tell you but...you want to know the truth, but in a way, you don't. Like stuff like that, you don't want to know that truth. Like, I don't. But in a way, you do...but yeah". (Young people)*<br><br>**Needs of the siblings:**<br>In 1 study (Steele 2013) where siblings were interviewed they provided advice about how medical teams could communicate more effectively with them and noted the need to be included in a developmentally appropriate manner.<br>One 17-year-old sibling stated, *"The doctors, they mostly just talked to my parents, but it might have been nice to have been included in stuff like that."*<br>Similarly a 14-year-old sibling added, *"They [doctors] talked to me, but they kinda talked down to me like I was stupid, 'cause I'm younger."*<br>*"Some people change depending on the situation they're around. Some people get more sophisticated than other kids. So they have more of an adult mind", added a 13-year-old* | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | *sibling.*<br><br>**Level of child and parent involvement:**<br>In 1 study (Hsiao 2007) where both parents and child were interviewed, it was noted that parents and their child did not always agree on the level of knowledge and involvement in the child's care.<br>*"You [the parent] need to talk to your child from the very beginning about what his or her condition is...Never underestimate something or oh this won't hurt...And don't deceive them, and I'll say the same for clinicians and physicians." (Parent)*<br><br>However, another parent stated that:<br>*"Do not talk in front of Marly, and any information that was gonna happen that day, like if any new things were going to change for Marly,...I want to know about it and I was going to tell her...of any change. Because the way I was going to tell would be a little different than perhaps someone else communicating that information". (Parent)*<br><br>**Needs for flexibility and formality:**<br>In 1 study (Stenekes 2014) conducted among healthcare professionals where several participants identified the needs for flexibility in the midst of unknown outcomes:<br>*"It is not always set out in stone. It can be very complicated at times. I know recently we had a situation where there was a plan that palliative care was involved, but there was confusion as to whether we would call neonatology or the resusc[itation] team...the team was not exactly sure why they should be present, if the baby would be palliative. So there was kind of like a flip-flop as to who would be caring for this child. So I think sometimes it's not always set in stone what's going to be done."* | | | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | | | | |
| **Sub-theme 4: Prior experiences of parents; parent resolution** | | | | | |
| 1 study (Byrne, 2011) | 1 study used archived materials | In 1 study where both parents' and HCPs' perspectives were taken into account (Byrne 2011), it was reported that prior experiences of parents, and parents' characteristics could act as either facilitators or barriers in communicating with families.<br>**Parents' prior experience and relationships:**<br>*"up against this dilemma, they (parents) felt no matter what they decide the net result would be an abandonment of the child they loved". "…with the marriage under enormous stress…" and the realization the treatment options were exhausted, the mother …"equates transfer to a palliative care program with "abandonment."* (HCPs)<br>**Parents' characteristics regarding resolution to diagnosis:**<br>The same study (Bryne 2011) reported that some parents had come to grips with the actuality of their child's diagnosis whereas the other remained essentially unresolved to this basic reality. Resolved parents still experienced sadness, doubt, and fear but were better able to listen during the consult and to utilise supports offered. Unresolved parents who questioned the diagnosis or were unrealistic about its implications remained ambivalent about any decisions to be made as well. | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |
| **Sub-theme 5: Cultural and religious background of the family** | | | | | |
| 4 studies (Contro 2002; Contro 2004; Davies 2010; Pearson 2013) | 3 studies used interviews, 1 study used both interviews | 4 studies reported on cultural and religious background of the family. This could act either as a barrier or facilitator in communication. These studies incorporated the opinions of both parents and HCPs. | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | and surveys; | **Attention to the cultural and religious background of the family:**<br>In 1 study (Davies 2010) where parents were interviewed it was reported that cultural and religious background of the family and the lack understanding of this could result in misunderstanding between families and healthcare professionals.<br><br>Parents interviewed in the study commented that some physicians incorporated the family's culture and religion when providing information and they appreciated that. One mother reflected,<br>*"the doctor would do everything he could, he didn't give us much hope." Knowing this family's strong religious belief, the physician said, "the one up above will have the last word. I will put myself in His hands, and I will do my best."*<br><br>In contrast, a Chinese mother was angry when a physician did not consider the cultural importance of family involvement. An intern *"impolitely"* asked the family to leave the room so that he could talk to the patient alone. The mother queried, *"how could the patient talk to him? The patient was very sick. He needed family to stay."* The mother described the intern as *"mean," stating, "He never considered our feelings."*<br><br>This theme was echoed by healthcare professionals interviewed in another study (Pearson 2013), where nurses stated:<br>*"They [the parents] all have different cultural and religious beliefs, so a lot of them led from their different cultural and religious beliefs"*<br>Also: | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | *"I personally was prepared for the family's reaction. They knew their child was going to die but when it happened, their response made me uneasy. They 'wailed' as part of their culture. I was unfamiliar with their culture so I was caught-off guard. I would like to know more about cultural differences with dying patients." (HCPs)*<br><br>**Language barrier and cultural differences:**<br>Another study conducted in the USA (Contro 2002) reported on this. It was noted that the lack of a common language compromised parents' ability to acquire complete information and to fully understand their child's medical condition, treatment, and prognosis. In addition, cultural differences could be detrimental to care. For example, if the Spanish-speaking parents' expectations that physicians show their child affectionate attention were not met, this became a barrier to trust and confidence in the medical team. These families reported feeling isolated, confused, and distrustful of the hospital system.<br>*"No one ever told me the baby could die. I never understood what was happening medically. The doctor came out during the operation and asked my wife if they should stop or continue the operation. I didn't understand that the baby would die either way at that point. No interpreter came during this conversation."* | | | |

1

2 **Table 20: Summary of evidence (adapted GRADE-CERQual): Theme 2 – Interpersonal/inter-active communication**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| **Sub-theme 1: Compassion and empathy** | | | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| 11 studies (Baverstock 2008; Branchett 2012; Contro 2002; Contro 2012; Davies 2002; Gaab 2013; Hsiao 2007; Meert 2008; Meyer 2006; Steele 2013; Weidner 2011) | 7 studies used interviews, 3 studies used surveys, and 1 study used diaries in writing or recorded; | Eleven studies conducted in New Zealand, the UK, and the USA interviewing HCPs, parents, and CYP receiving palliative care reported on the importance of compassion, empathy, affect and kindness in facilitating communication between HCPs and families. Parents appreciated that their grief and loss understood by HCPs and those who have provided care to their child shared their grief with them.<br><br>**Compassion:**<br>*Compassion and humanity:*<br>*"The need for compassion and humanity not to be just a technician (consultant)"*<br>"*If you do not have empathy, e.g. shed tears or reflect on these issues, it is time to retire" (consultant)*<br><br>***Compassion and care, allowing for hope when delivering the difficult news:***<br>In 1 study where parents were interviewed (Contro 2012) they emphasised that difficult news should be conveyed with compassion and care, using straightforward nontechnical language. Above all, they recommended giving difficult news directly and honestly while still allowing for hope. Parents also mentioned they would have appreciated better preparation that bad news was coming.<br><br>***Compassionate and sensitive in terms of timing of delivering the information of imminent death:***<br>Two studies (Weidner 2011; Contro 2012) reported on this and they incorporated the opinions of both parents and HCPs.<br>Parents commented that the timing of delivering the news of imminent death should be sensitive and compassionate. Health care providers should know what to tell parents and | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | ascertain when they are ready to accept information related to their child's death.<br>*"All of the doctors and nurses came over and started doing the drill of "it's very bad," which I wasn't prepared for...a little overwhelming. I would just say it's really important for folks to realise people handle this kind of stuff differently. " (parent)*<br><br>This was also recognised by HCPs in another study, where interviewed HCPs stated:<br>*"The timing of our interventions is usually too late…Sometimes we got called to work with a sibling right when the child is dying…that is way too late and way too awkward…" (child-life specialist)*<br>*"The problem is we still have trouble with addressing palliative issues in a timely manner…" (nurse)*<br><br>**Empathy:**<br>***Compassionate and HCPs showing emotions***:<br>This theme emerged from several studies (Gaab 2013; Meyer 2006; Steele 2013) where parents were interviewed:<br>*"Be compassionate and ask how parents are. Don't fall into that detached type of working. Parents need to feel that people really care, not that it's just a job. The people at the hospital who allowed themselves to have genuine feelings helped me the most." (parent)*<br>*"[The staff ] ...stood there with us and shared our grief. How can you improve on that? They communicated volumes with that simple act. (parent)*<br><br>One parent described the physicians' warm display of emotion at the time of her child's death:<br>*"I remember after we had our quiet time with S- after she passed, the doctors were all outside the door. And they were* | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *very kind and some of the young doctors were in tears. And it was very moving to see all these emotions because they had watched her fight for days." (Parent)*<br>A mother added: *"These kids are dying, and they know they are dying. Some of them [healthcare providers] need to be more compassionate."*<br><br>***Empathy and understanding:***<br>Parents longed for understanding, in 1 study (Branchett 2012) where parents were interviewed they reported that:<br>*"[My baby] had been in NICU for nearly 3 weeks and it helped that the nurses that had cared for him in that time came and said goodbye to him. It showed me that he was not just another statistic, he was my baby." (Parent)*<br><br>**Logistic barrier to honour parents' wishes around the child's death:**<br>However, in 1 study where HCPs were interviewed they noted that sometimes it is logistically difficult for them to honour parents' wishes around the child's death (Contro 2012).<br>*"I recently worked with a Jewish family who wanted to remain with the body overnight. I did everything I could to honour the family's important wish because I knew it was what they needed. However, finding space for this to happen took a miracle. I should have been doing others for the family but spent most of my time on this one issue." (Social worker)* | | | |
| **Sub-theme 2: Trusted relationship and trusted care providers near the time of the child's death** | | | | | |
| 7 studies (Baverstock 2008; Robert 2012; Hsiao 2007; Caeymaex | 6 studies used interviews; 1 study used surveys; | Several studies reported on the importance of developing trusted relationship in communication and having trusted and familiar HCPs around near the child's end of life. These studies took account of the perspectives of HCPs, parents, | Limitation of evidence<br>Coherence of findings | Minor limitations<br>Coherent | MODERATE |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| 2011; de Sa Franca 2013; Meert 2008; Davies 2003) | | and children living with a LLC. Trusted relationship:<br><br>**Trusting relationship with HCPs:**<br>In 1 study (Caeymaex 2011) where parents were interviewed they stated that they appreciated dealing with the same caregivers the whole time:<br>*"All 10 days, this paediatrician was there. She was really a person with whom we made decisions, choices, and she was there for us in the last seconds […] She shared everything with us". (Parent)*<br><br>This was supported by findings from another study where children and parent were interviewed (Hsiao 2007). They reported that they appreciated doctors who took the time to get to know the patients as individuals and develop a friendship with the patients.<br><br>**Trusted HCPs near the child's end of life:**<br>In 1 study (Robert 2012) where parents were interviewed it was noted that Intimacy was highly valued at the child's end of life. Trusted HCPs were increasingly relied upon, and some parents limited their child's interactions to persons well known to the family.<br>*"If somebody wasn't there throughout the whole ordeal, I wasn't interested in talking to them.....It's pretty hard to open to with somebody you don't know at that point in time in your life....I go back to the relationship and trust." (parent)*<br>Demonstration of effort and competence; determination to help and knowledge and capacity to do so:<br>In 1 study (Hsiao 2007) conducted in the USA where children living with LLCs were interviewed they cited that,<br>*"They really have a visible care for the patients...a determination and.. doggedness to help them in any way to* | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *go past the call (of duty)" (child)*<br><br>**Ties of trust established between child and HCPs:**<br>One study carried out among nurses (de Sa Franca 2013) reported on the authentic communication focusing on care to enable well-being and better-being, which built up the ties of trust between nurses and the child. One nurses cited that:<br>*"This communication issue, I always try to, like, reassure, especially in relation to pain. I try to talk to her, to address her [...] you look into that child's eyes, she is looking at you, she'll trust you. [...], it is a touch, a gaze; you have to show confidence (Nurse).*<br><br>This was supported by CYP interviewed in another study (Hsiao 2007), where a child stated:<br>*"It's not really a doctor-patient kind of thing...it's more just-I would say a friendship. ... It helped me deal with my pain, you know, when we talk to each other." (Child)*<br>On the other hand, behaviours that break trust acted as barrier to good and effective communication.<br><br>**Medical terms and pace:**<br>In 1 study carried out in the USA (Meert 2008), several parents commented on the complexity of language used by physicians when communicating about their child's condition. Parents wanted information provided in *"layman's terms"* or *"English terms"* rather than *"doctor talk"*. One parent described her inability to understand the treatment that was planned for her child<br>*"I kept asking, 'What is this? What are you telling me you are going to do for her?' They gave me answers in medical terminology. This is what I kept getting, and I'm like, 'Could you explain that?' No one really explained it to my satisfaction because I did not and still do not understand. And* | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *I would like to understand it in layman's terms. It was what you were gonna do for her". (parent)*<br><br>**Body language:**<br>Parents interviewed in several studies commented on physicians' nonverbal behaviours when giving bad news. Physicians' body language led some parents to suspect the physicians were *"guilty"* or had *"done something"*.<br>One parent described the physician's lack of eye contact, *"I wanted to ask the doctor, after he came out and talked to me after her procedure, why didn't he look me in my face, he kept his head down to the ground talking to me. Then when he lift his head up he turned the other way but he never looked me in my eyes. What went wrong?"*<br><br>**Insensitive, high-powered authoritarian attitude:**<br>In 1 study (Davies 2003) where parents were interviewed and they reflected that HCP's non-responsible attitude, delay in diagnosis and no apologies afterwards, persistence in treating parents in a dismissive and off-hand manner made them angry:<br>*"The first time we went to see him after she was diagnosed was the only time she was with him longer than two minutes and he had the cheek to say 'Yes, you could see she was classic MPS.' That made me so angry" (mother of daughter diagnosed with Sanfillipo Syndrome).* | | | |
| **Sub-theme 3: Being sensitive:** | | | | | |
| 9 studies (Contro 2004; Davies 2003; de Sa Franca 2013; Hendricks-Ferguson 2007; Gordon 2009; | 8 studies used interviews and 1 study used both interviews and surveys; | Nine studies reported on the theme of HCPs being sensitive to patients' situation and needs. They incorporated the opinions of both parents and HCPs.<br><br>**Being sensitive when breaking bad news:** | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of | Applicable | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| Hsiao 2007; Lundqvist 2002; Robert 2012) | | Parents commented that because careless and insensitive remarks caused families lasting pain and complicated their grief, they would appreciate it if HCPs were sensitive with regard to breaking the bad news, hospices care recommendation for child, or responding to parents and families' concerns. Many parents reported being devastated when physicians broke bad news in an insensitive manner.<br>*"I know we had to ask if we didn't want our son resuscitated. It's just the way he did it. It was very cold. He was saying 'if he has to be resuscitated, this is what's going to happen...' It was very negative talk about our son dying. (parent)*<br>*"They were sensitive when they told us but they told us outright" "there is a hospice programme here' 'He was very kind about it and matter of fact when he said, 'You will need help" (parent)*<br>*"Being congratulated by the nurse for having given birth to such a fine baby was painful under the circumstances. Still, the mothers were understanding about such behaviour. I don't think you can congratulate, even more, ask, "How are you?" or "Look here!"…It was almost as if it was thrown at me what is she saying? Don't congratulate me! He was lying there. Only by looking at him you would have understood that congratulations were not appropriate". (parent)*<br><br>**Unfamiliar staff near the time of the child's death:**<br>Parents in 1 study (Robert 2012) commented that:<br>*"Be sensitive. Trust comes from time and relationship. It was difficult when doctors that I have never seen come in at the end of. [They weren't going to] make his life more comfortable. They were researching, and were trying to participate, but once we cross that line, it was time for us, not them"*<br><br>**Request for organ donation at the wrong time:** | findings | | |
| | | | sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | Parents in 2 studies (Gordon 2009; Lundqvist 2002) told narratives of a request for organ donation and criticised the clinician's professionalism:<br>*"I remember when he was telling us my son was brain dead in the same sentence he was asking us to donate his organs. And I feel that was inappropriate at the time." (parent)*<br>*"Our last wishes were that we would be left alone when the ventilator was withdrawn. … But the physician came and asked, with a smile on his lips, about an organ donation. It was frustrating. … Our last moments together with the baby, and he could not wait. …" (parent)*<br><br>**Sensitive with the child's verbal and non-verbal communication:**<br>Nurses in 1 study (de Sa Franca 2013) noted the importance of being sensitive to child's ways of communication when providing care:<br>*"Communication is very important in palliative care. [...]. Children, sometimes, during the initial phase of the disease, do not communicate with words, but communicate with their gaze, with touch. You have to understand that! It is a call that the child is presenting to us. [...], Communication is not only with words: it's a gesture, it's eye contact, it's a way of waking up, it's a good day s/he gives you. It's a smile she transmits you; it is knowing how to recognize these signs" (Nurse)*<br>*"In communication with children, we have to pay attention to all the communication channels (verbal and non-verbal). So, we need to learn to read the children's sixth sense. [...]. In this sense, if she is in the terminal phase, she realizes it's changing, permits other things".(Nurse)* | | | |
| **Sub-theme 4: Managing hope (balance between hope and realism) and divergence:** | | | | | |
| 10 studies | 9 studies used | The theme of hope and managing hope, and managing | Limitation of | Minor | MODERATE |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| (Baverstock 2008; Contro 2002; Forbes 2008; Gordon 2009; Hsiao 2007; Meert 2008; Meyer 2006; Price 2013; Robert 2012; Wood 2010) | interviews and 1 study used surveys; | underlying tension caused by divergence between parents and HCPs emerged from 10 studies conducted in the UK and the USA. These studies incorporated the opinions of parents and HCPs.<br><br>**Managing parents' hope without creating false hope, balance between hope and realism:**<br>*"I mean when I asked questions, um, they were explaining things. But, you know, many times they came in during the day and, uh, there were things just – and then they walked out. And, kind of ignored us a little bit. And I realize now when I look back that – that the doctors realized certain things where we had still this glimmer of hope. And, um, but they had seen – have so much experience they do know and understands the signs. And, um, I don't know if they really wanted to tell us more about it. And, take this glimmer away" (parent)*<br><br>**False hope:**<br>In 1 study (Gordon 2009) some parents held clinicians directly responsible for creating or maintaining false hope as the death of their child approached:<br>*"Cause I would have much better they told me her chances were slim or her chances was nil or something. But she's not gonna be OK. And I got mad at them because they told me she was gonna be OK if she wasn't."*<br>*"Communicate honestly, false hope in this situation is unfair." (parent)*<br><br>**Allowing for hope:**<br>However, in another 3 studies (Contro 2002; Hsiao 2003; Wood 2010) where parents were interviewed, they hoped HCPs could provide hope during the end-of-care of their child. In 1 study (Contro 2002), parents stated that doctors | evidence | limitations | |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | need to relay medical facts honestly but always allow for a glimmer of hope, even if only for a miracle.<br>Mother: *"I mean what we've been through over the years with [daughter's] consultant in [local hospital], who I find is a very grey man with a very grey aura who gives you no hope, and I could, I just feel like screaming at him and saying, "Do you not understand, I have to deal with this every single day of my life why can't give me a glimmer of hope?"*<br><br>**Divergence, discord between parents and HCPs (regarding whether to deliver the truth to the child):**<br>Several studies (Baverstock 2008; Forbes 2008; Price 2013) conducted in the UK and USA reported that sometimes the divergence, disagreement, even discord posed challenge to the communication.<br><br>***On approaches regarding whether to deliver the "truth" to the child:***<br>HCPs explained that many parents sought to hide the *"truth"* of likely impending death in an effort to protect their child from further suffering, participants were unequivocal that the most appropriate strategy was to tell the child the *"truth"*. Disparity between professional and parental approaches was considered to create an underlying tension between the 2, resulting in additional stress felt by participants as they strove to uphold a partnership approach to care.<br><br>***Discord/disagreement relating to care in the process:***<br>HCPs in 1 study (Price 2013) also reflected that at least some degree of discord was associated with a wide range of issues, including: talking about death to children, whether or not to resuscitate, addressing sibling need, location of care, securing services, withdrawal of treatment/food/fluids, and parental denial. | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
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3 **Table 21: Summary of evidence (adapted GRADE-CERQual): Theme 3 – Emotional factors in communication**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| **Sub-theme 1: Display of emotions and emotional impact on HCPs** | | | | | |
| 7 studies (Byrne2011; Contro 2012; Forbes 2008; Meert 2008; Meyer 2006; Midson 2010; Price 2013) | 5 studies used interviews; 1 study used surveys; and 1 study used both interviews and surveys; | In 7 studies where parents and HCPs were interviewed, parents reported that warm display or expression of emotions from the HCPs was appreciated by them. On the other hand, HCPs interviewed reported on the personal emotional impact involved in end of life issues discussion with parents and children.<br><br>**HCPs displaying emotions:**<br>HCPs' warm display emotions at the time of child's death was appreciated by parents because they felt this showed the compassion and understanding from the HCPs.<br>*"I remember after we had our quiet time with S– after she passed, the doctors were all outside the door. And they were very kind and some of the young doctors were in tears. And it was very moving to see all these emotions because they had watched her fight for days."*<br><br>In another 2 studies (Contro 2012; Meyer 2006), parents endorsed staff's emotional expression both verbally and behaviourally. This was generally perceived as authentic and reflecting care beyond that embedded in the professional role. Some parents encouraged staff to *"be real people"* and to allow themselves to express real feelings. | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *"One of our fellows was so overcome that he sat in the corner of the room when the child died and cried. He felt bad that he wasn't more able to do something and sated, 'I was speechless.' The mother reported to me that this display of emotion meant more to her than any words ever could."*<br><br>**Personal emotional impact on HCPs:**<br>***Frustration, sadness:***<br>Although HCPs gained considerable fulfilment from their work, emotional impact was most frequently discussed in negative terms. This included strong feelings of inadequacy, frustration, and sadness arising from the complex, intense, and often protracted nature of professional engagement with dying children, their parents and wider family<br><br>***Fear of discussing difficult issues, transitions:***<br>Some HCPs reported *"fear"* of dealing with discussions such as withholding life sustaining equipment. Another study reported that HCPS could experience fear as well when transition was about to occur, especially when the goals of a medical team with an intense curative focus did not align an integrated palliative care focus, the consulting team needed to defer while also advocating for their view of the family's and child's best interests. This role exposed the medical team to its own frustrations, anger, and sadness, and the need to channel these appropriately to continue to work well with both the families and providers.<br><br>***Fear of death***<br><br>***Fear of reactions:***<br>HCPs in 1 study (Midson 2010) also reported that not knowing how a family, or child, might respond or how they | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | might feel if asked to discuss death and dying can lead to them avoiding the issue. This may lead to blocking the *"cues"* that children or families might use to try and ask about possible outcomes.<br><br>***Difficulties in acknowledging that the patient cannot recover*** | | | |
| **Sub-theme 2: Emotions of parents** | | | | | |
| 2 studies (Baverstock 2008; Price 2013) | 1 study used interviews; 1 study used surveys; | In 2 studies where HCPs were interviewed they reported that parents and children could experience stress, anger, and distress and this could act as barriers for communication.<br><br>**Anger, stress of the parents:**<br>HCPs in 1 study (Price 2013) spoke of open conflict and also of how they found themselves being the *"target"* of parents' anger and stress, particularly during the period immediately leading up to their child's death.<br>*"I have learnt to deal with anger and aggression as a symptom of distress" (consultant)*<br><br>***Parents' anger, fears and sadness:***<br>These emotions pervaded the presenting or underlying affect of parents as they participated in consults. They were expressed openly or kept covert and made apparent through silences, body language, and brief remarks. | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

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2 **Table 22: Summary of evidence (adapted GRADE-CERQual): Theme 4 – Active involvement communication**

| Study information | Description of theme or finding | Quality assessment |
|---|---|---|

| Number of studies | Design | | Criteria | Rating | Overall |
|---|---|---|---|---|---|
| **Sub-theme 1: Comprehensive planning; get parents prepared; and provide re-assurance** | | | | | |
| 7 studies (Baverstock, 2008; Forbes 2008; Hendricks-Ferguson 2007; Midson 2010; Robert 2012; Stenekes 2014; Weidner 2011) | 5 studies used interviews; 2 studies used surveys; ; | 7 studies reported on this theme. These studies incorporated the opinions of parents and HCPs. The importance of planning both among medical teams and with the parents to achieve good communication was noted.<br><br>**Comprehensive care plan with clear goals and roles of involved HCPs:**<br>HCPs frequently cited communication as the most crucial element in providing perinatal palliative care. When communication between teams was weak, the development of a comprehensive care plan was affected, which resulted in unclear goals.<br>*"When things go poorly, to me the first thing that goes wrong is communication...Another element that trends to fall apart is confusion about roles of the healthcare team. So we find on some occasions that it's not clear to the family or to the healthcare providers who is attending to what with regard to the baby's needs, and who is primarily responsible and accountable for the needs of the baby and the family" (HCPs)*<br><br>**Coordination of care and roles in the team:**<br>In 1 study (Midson 2010) where the opinion of HCPs were explored, it was reported that while junior staff are often at the bedside listening to children and families, it can be difficult for them to respond to the direct question of, *"Am I going to die?"* This is especially so if the consultant has not agreed a plan or discussions have not been held. (Researchers' comments)<br><br>***Good planning before discussion around the time of a child's death:***<br>Consultants interviewed in another study (Baverstock 2008) thought discussions tend to *"go well"* when there has been good planning and introductions, honesty and mutual respect | Limitation of evidence | Minorlimitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | and the *"right environment"*. Conversely consultants thought it more difficult when there was poor planning, lack of time, interruptions and when there was disagreement with parents.<br><br>***Communicate and documentation:***<br>Parents in another study (Robert 2012) reported that it could be difficult for them when communication, record keeping was lacking between departments. This was echoed by HCPs in another study (Forbes 2008) where they commented that poor documentation of previous discussions was not helpful when speaking to parents.<br><br>***Compassionate and caring when discussing EOL options:*** give options, give opinions, and focus on what's the best for the child<br>In 1 study (Hendricks-Ferguson 2007), a mother was grateful for how well the physician communicated the issue and helped the parents in making the best decision for their daughter and accepting her death.<br>*"He encouraged us to consider where our daughter would be most comfortable and where we would want her remaining time to be spent, in an out of the hospital or at home with us." (parent)*<br><br>***What to be expected in the dying process:***<br>As the child approached death, it was important to parents to be told what to expect so they could prepare themselves for physical changes they would see in their child. They depended on healthcare providers to explain what was going to happen next in the death process.<br>*"There are certain things that happen to a dying child that somebody who is not and an RN or somebody who is not medically qualified would not know about...more emphasis should be put on that. People should be prepared to know* | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *what's [going to] happen when, and what their child is going to look like. Things they can do. Just the overall picture"*<br><br>***Confirmation and reassurance from staff about the decision made:***<br>Parents talked about the conflict they felt over whether they had made the best decisions for their child; due to this, they appreciated the reassurance they received from healthcare providers.<br>*"That's probably the only thing I walked away from the hospital feeling conflicted about...Did they fully understand who she was and whether this was right? Should I really have taken her off the ventilation? Was it the right decision?...Knowing that I was dealing with people didn't necessarily know her, so they might not know the nuances, even though they know their crafty very well" (Mother)* | | | |
| **Sub-theme 2: Mutual respect; Respect the parent's perspective and knowledge** | | | | | |
| 5 studies (Caeymaex 2011; Davies 2003; Meyer 2006; Steele 2013; Weidner 2011) | 4 studies used interviews; 1 study used surveys; | Parents interviewed in 6 studies reported that they appreciated HCPs' respect and acknowledgement of their roles in the end of life care for their child, they valued being listened to, respected, and not judged.<br><br>**Mutual respect; respect parents' perspectives and knowledge:**<br>*"I want them [medical staff] to respect my point of view as much as I was respecting theirs. They were pressuring [me] to make decisions that I knew were not right at that time. We know that they've been taught. We are very grateful for what they are doing. They do their best, but there are those times that they have to listen to parents."* (parent)<br>*"Listen to what the parents have to say. Show more sincere compassion for the parents' and the child's needs. In the long* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | *run, the parents do know what is best for their child."*<br>*"When I would read my child's chart and see 'impaired coping' written, there was nothing more disrespectful. I'd like to see some of these people 'cope' with the same situation and have to read that someone thinks they're 'impaired.' I personally saw to it that one nurse who wrote that in the chart not take care of my son again." (parents)*<br><br>Respectful language toward the child and the parents left a memory of the doctor's positive intentions:<br>*"Doctor A always called the baby by her name: 'Lena has very serious sequelae'. She was a person, not an ordinary case". Inversely, a disagreeable, barely involved attitude encouraged subsequent questions about the decision taken: "This doctor, I don't ever want to see him again. When he told us that it was no longer legitimate to continue the resuscitation, he said it to us casually, without emotion, as if that happened to him every day. He was not warm. So, was he telling us the truth? That's a question"* | | | |

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## 5.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis.

## 5.7 Evidence statements

A number of themes emerged from the interviews with parents and children, multidisciplinary health care professionals, and healthy siblings. Around the central theme of timely, honest, and consistent information exchange that was still relevant for communication, subthemes of personalised/individualised communication, interpersonal/interactive communication, emotional factors, and active involvement communication were found to be interlinked. These were perceived as important for effective communications between families and healthcare professionals by those who had been involved in the end of life care for children and young people.

**Personalised/individualised communication**

Moderate quality evidence from 19 studies where parents, healthcare professionals, and children and young people were interviewed indicated that participants thought that communications tailored to the individual child, incorporating parents' and carers' needs, situations and family contexts were helpful. Specifically, these included treating the child and parents/carers as individuals; providing time, space and privacy at different time points; accommodating the needs of parents or carers and children/young people, prior experiences of parents/carers; and cultural, religious, and language differences.

**Interactive/interpersonal communication**

Moderate quality evidence from 23 studies where perspectives of parents/carers, healthcare professionals, children and young people were taken into account, showed that participants valued the important roles played by compassion and empathy; trusted relationships and trusted healthcare professionals; healthcare professionals being sensitive; and managing hope in facilitating the interactions between families and healthcare professionals. During the time leading up to and around their child's death, parents appreciated it if their situations, grief, and loss could be understood and empathised in a compassionate and caring way. On the other hand, in echoing parents' needs for compassionate care, some healthcare professionals recognised there could be logistic barriers when trying to honour the families' wishes such as dedicated space for parents and child around the time of death.

**Emotional factors**

Moderate quality evidence from 8 studies where healthcare professionals and parents were interviewed showed that emotional factors could act as either facilitators or barriers in the communication between families and healthcare professionals. Parents reported that they appreciated healthcare professionals displaying their real emotions as they took that as an authentic gesture of caring and understanding. However, it was noted by healthcare professionals that the personal emotional factors involved in end of life care for children and young people such as fear, sadness, and frustration could act as barriers for them in initiating different discussions with parents. Further, some healthcare professionals also reported that sometimes they found themselves being the targets of parents' anger and stress especially around the time of child or young person's death.

**Active involvement communication**

Moderate quality evidence from 11 studies in which parents and healthcare professionals were involved, showed that healthcare professionals found it helpful if there was comprehensive planning for communications among medical teams and with the families. This included the discussion around the time of the child's death. On the other hand, parents reported that they appreciated being informed of what to expect, especially during the child's dying process, and confirmation and reassurance from healthcare professionals after a decision was made. Furthermore, parents also reported that they appreciated it if their perspectives and knowledge of the child and the child's care needs could be respected as well, when important decisions were made.

# 5.8 Linking evidence to recommendations

## 5.8.1 Relative value placed on the themes considered

Evidence on most of the expected themes considered important during the protocol development was identified. The Committee also considered other themes that emerged from the literature. Because of the close link between information provision and communication, the main themes that emerged from this review shared some similarity with the information provision review. In addition to the themes identified in the information provision review, further, such as individualised communication, interactive/interpersonal communication, and communication that facilitates active involvement in the child or young person's care were also considered. . Themes that promote an individualised approach to communication were considered particularly important (such as emotional factors that may help or hinder good communication or personalised communication).

## 5.8.2 Consideration of barriers and facilitators

Overall, the Committee thought that the themes and their sub-level themes emerged or derived from the evidence were useful and relevant in terms of both general principles and details needed for communication with children and young people living with life-limiting conditions and their families. They agreed that the evidence fit their clinical observations in terms of what delivered good communication in this context.

Based on the evidence and the discussion the Committee noted that communication in end of life care for children and young people should be undertaken as a continuous process. Healthcare professionals should think about the best ways of communicating during the whole course of the child or young person's condition, which could range from the time of diagnosis, regular communication at intervals as part of the Advance Care Plan, at the time when the child or young person is more unwell, to when the child or young person may be approaching the end of life. One thing they thought healthcare professionals should always be aware of in this process is that they should not make assumptions about what would be needed by the families when communicating with them, and they should instead ask families what forms and timings of communication work best for them.

As suggested by the evidence, the Committee agreed that healthcare professionals need to tailor communications to individual/family situations, taking account of the families' cultural, spiritual or religious background. Some families may have special needs such as a translation service from an interpreter. Healthcare professionals also need to make sure that all people with parental responsibility are communicated with and kept informed at every stage of care.

The Committee also noted that not all healthcare professionals would have the cultural understanding and competences to manage clear and sensitive communications with the parents or carers in all circumstances during end of life care. Some healthcare professionals may lack the skills needed when delivering difficult and distressing information. However, the

need to have urgent discussions can arise unexpectedly, for example if a child or young person experiences a sudden deterioration in their condition, or if it appears that they are likely to die soon. The Committee discussed that the decision regarding which healthcare professional is best able to lead the discussion with the child or young person or their parents or carers at any particular time, there are a number of factors to consider. These factors include: the healthcare professionals' expertise and ability to discuss the matter in question; their availability at a time when frequent discussion is appropriate (for example, in the context of a serious deterioration in the condition); and the views of the child or young person and that of the parents or carers, because sometimes they may have established a relationship with particular members of the teams with whom they feel comfortable communicating. It should be recognised that this will not necessarily be the function of a specific job role.

Many of the themes emerging from the review highlighted the importance of an individualised approach to communication. The Committee therefore discussed the importance of accommodating the needs and preferences of the child or young person and their parents' or carers', when talking about death. They stressed that the children and young people's needs should not be neglected in this process. Some children and young people may not be willing to discuss difficult issues such as death, while others may not have the ability to do so. However, the Committee thought it was important to explore this with the child or young person. Healthcare professionals should be aware that the way they approach this may influence the child so that they do not report their true feelings. The Committee also thought it was important to consider whether the child or young person and their family members would need support in talking to one another, for example about death. They noted that in addition to the parents' and carers' responsibilities, the child's and young person's right and entitlement to know about their diagnosis and prognosis should be respected and considered, taking into account their ability to understand the issues, and whether this discussion would be in their best interest.

The Committee recognised the importance of accommodating the needs of families around the time of the child's death in terms of allowing them time and space to stay with their child. They recommended that dedicated space and time should be arranged while providing necessary support. This family support should be planned in advance and any costs or other difficulties anticipated and allowed for.

The Committee thought it was important for healthcare professionals to realise that the children and young people living with life-limiting conditions and their parents or carers are in a vulnerable situation, and so sensitive and compassionate care is needed. However, the Committee also agreed that it was essential for healthcare professionals to be open and honest in their discussions. The Committee recognised the importance of discussing clinical uncertainty, and this was reflected in the evidence. They also highlighted the importance of providing reassurance when this was appropriate, while avoiding unrealistic statements in any discussions.

The Committee agreed that the child or young person and their parents/carers should be involved in decision-making with regard to difficult issues such as withdrawal of life sustaining treatment. Moreover, as suggested by the evidence, the Committee noted that there would be emotional burdens such as frustration, fear, and anger among parents or carers when the child or young person is approaching the end of life. The Committee agreed that healthcare professionals should provide support through empathy, and through attentive and compassionate listening.

The Committee recognised the importance of communication with parents or carers around the time of the child's death. They recognised the importance, if healthcare professionals think a child or young person is likely to be approaching the end of life, of explaining to the parents or carers why they think this, and if this is uncertain to discuss their reasoning and any matters around it. The healthcare professionals should also help families (including parents and siblings) prepare for what may be expected in terms of symptoms and signs

developing in a child or young person who is dying, and how these symptoms and signs might be managed. This information may need to be provided on more than 1 occasion.

The Committee noted that details about the content of training of healthcare professionals was outside of the remit of NICE. However, they considered that it was appropriate to recommend that all healthcare professionals who provide end of life care to children and young people, should be equipped with the appropriate skills for communicating with the child or young person and their families during their end of life care.

#### 5.8.2.18 Barriers and facilitators highlighted in the TFSL report

Many of the children and young people identified their consultant as the key source of medical information about their conditions because they trusted their expertise. It was highlighted that asking their consultant or other trusted professionals was preferred to using the internet for medical advice and information, although they reported that getting advice out-of-hours was sometimes difficult.

Participants highlighted that it was important for them to have time and opportunities to ask questions. Speaking to other young people was perceived as helpful and important as well. Participants varied in how confident they felt about asking questions during consultations, with some being actively involved at the time and others preferring to ask later, or to listen and then check other information sources for additional material. Some participants also reported that they knew as much as or more than their parents about their condition, while for others, parents continued to be an important source of information and advice.

### 5.8.3 Economic considerations

While there are aspects of communication which have opportunity costs, such as the staff time and some of the different communication formats that may be useful in this population, these are typically relatively small and would ordinarily be considered within the provision of standard services and care. Good communication is recognised as important within all healthcare provision, and that patient care can suffer as a result of poor or ineffective communication. Therefore the Committee were of the view that their recommendations on communication would promote a cost-effective use of NHS resources.

### 5.8.4 Quality of evidence

Moderate quality evidence was found in the review. The main reasons leading to downgrading of the evidence that was shared by the majority of studies included:

Low response rate from participants and self-selection bias: in many studies only about half or less than half of the respondents contacted consented to be interviewed. Subjects who chose to participate may have been different from those who refused to be interviewed. On the other hand, in some studies participants were selected by the physicians who had provided care to the child, and those who were excluded from participation may have been the group that had different views and needs for communication.

Uncertainty in terms of saturation in data analysis and data collection: the majority of studies did not report whether saturation was achieved in terms of data collection or data analysis. It was difficult to ascertain from the information reported in the studies. However, when considering the evidence as a whole, saturation was achieved on some meta-synthesised themes.

Lack of the critical review of the researcher's role in sample recruitment, data collection or data analysis process: few studies clearly reported the relationship between researchers, interviewers and the respondents, whether the researchers had a pre-understanding about the topic or the possible influence of that in data collection and the analytical process.

Lack of verification of findings: only a few studies verified their findings with participants or external sources.

Finally, many studies did not report in detail how findings/themes were derived or emerged from the data in their research, although word limits in journal publications might be a reason for that.

Applicability: findings from the majority of included studies were considered to be applicable to the UK setting because of the direct relevance of their populations, contexts, and the topics explored.

None of the studies included in the review of the literature presented views from children affected by life-limiting conditions within the UK setting. For this purpose a focus group study was carried out specifically for this topic to bridge this gap.

### 5.8.5 Other considerations

The Committee noted that the quality of evidence was generally moderate, so the resulting recommendations were made based on the evidence which corresponded with their observations.

The Committee discussed the need for communication skills training for healthcare professionals, although they noted this was outside of NICE's remit. They considered it important to place healthcare professionals with the appropriate skills in the right setting, to communicate difficult and sensitive issues with families at the most appropriate time. This was reflected in the recommendation.

The Committee discussed whether they wanted to prioritise this topic for a research recommendation, but they concluded that the combination of the evidence (including the focus group report), their experience and their expertise was sufficient to base the recommendations on.

#### 5.8.5.1 Other considerations related to the TFSL focus group findings

The Committee considered that the findings reported in the TFSL's report, reinforced the evidence review which showed the importance of communication based on individual needs and situations, what should be considered when planning to communicate at different time points, and who should be providing the communication as already discussed. As described in the report, children and young people choosing how much they want to know and opportunity for the child or young person and their families or carers to ask questions were perceived important by those interviewed and emerged as important themes.

Children and young people with life-limiting conditions varied in how much information they wanted to know about their condition and about possible treatments or procedures. This varied by individual, and over time as some young people became more involved in decisions about their care; and also varied from decision to decision. Sometimes too much information was seen as intimidating and caused participants to worry about what might happen; for others, not receiving all the information could make them distrust the person providing it.

The Committee noted that children and young people with life-limiting conditions agreed that it was important to have time and opportunity to ask questions, which helped them to learn and understand more if they wished. Some children or young people were well connected to their care team and had a contact to arrange this for them. Some asked parents to facilitate communication, or would wait for their next planned consultation to ask questions. The Committee discussed the importance of healthcare professionals checking with children or young people with whom, when and about what they wanted to communicate about their condition, and about care planning and the importance of ensuring channels of

communication were available to children and young people when they felt the need to talk and ask questions.

#### 5.8.5.23 Key conclusions

The qualitative evidence provided insights into what people involved in end of life care of children with life-limiting conditions perceived as important and helpful. A report specifically conducted for this guideline provided more direct evidence in what children or young people in the UK setting would perceive as effective communication strategies or styles. This was deemed as particularly helpful in the recommendations drafting process.

## 5.99 Recommendations

7. **When difficult decisions must be made about end of life care, give children and young people and their parents or carers enough time and opportunities for discussions.**

8. **Think about how to provide information for children and young people with life-limiting conditions, taking into account their age and level of understanding. When appropriate, use formats such as:**
    - one-to-one discussion
    - play, art and music activities
    - written materials and pictures
    - digital media, for example social media.

9. **When deciding how best to communicate with the individual child or young person and their parents or carers, focus on their views and take account of:**
    - their personal and family situation
    - their religious, spiritual and cultural beliefs and values
    - any special needs, such as communication aids or the need for interpreters.

10. **Ask children and young people with life-limiting conditions and their parents or carers:**
    - if there are other people important to them (such as friends, boyfriends or girlfriends, teachers, or foster parents) who they would like to be involved, and if so
    - how they would like those people to provide a supporting role.

11. **Think about how best to communicate with each child or young person and their parents or carers:**
    - when the life-limiting condition is first recognised
    - when reviewing the Advance Care Plan
    - if their condition worsens
    - when they are approaching the end of life.

12. **Ensure that all parents or carers are given the information and opportunities for discussion that they need.**

13. **When deciding which healthcare professional should lead on communication at a particular stage in a child or young person's illness, take account of:**
    - their expertise and ability to discuss the topics that are important at that time
    - their availability, for example if frequent discussions are needed during an acute illness or near the end of life
    - the views of the child or young person and their parents or carers.

14. **When a life-limiting condition is first diagnosed, tell the child or young person (if appropriate) and their parents or carers about the condition and what it may mean for them.**

15. **Be aware of the importance of talking about dying, and if appropriate discuss with children and young people:**
    - whether they want and are able to talk about dying
    - whether they or their parents or carers would like support in talking to each other about this.

16. **When a child or young person is likely to die within hours or days, support them and their parents or carers by:**
    - listening to any fears or anxieties they have and
    - showing empathy and compassion.

17. **If a child or young person is likely to die within hours or days, explain to them and their parents or carers:**
    - why you think this is likely, and any uncertainties
    - what clinical changes can be expected
    - whether you think the treatment plan should be changed.

18. **Be aware that children and young people may have difficulty asking directly if they are going to die or are dying. Explore and discuss their concerns if you think they want to talk about this.**

19. **Be aware that parents or carers may have difficulty asking directly if a child or young person is dying. Explore and discuss their concerns if you think they want to talk about this.**

# 6 Shared decision-making and Advance Care Planning

## 6.1 Advance Care Planning

### 6.1.1 Review question

**What are the barriers and facilitators to the child or young person, the family or carer of the infant, child or young person and the multidisciplinary team in being involved in decision-making to inform the development, assessment and reviews of personalised, parallel and Advance Care Planning (including if appropriate decisions about continuing or stopping life-sustaining treatment and attempting cardiopulmonary resuscitation)?**

### 6.1.2 Introduction

Personalised, parallel and Advance Care Planning are processes that involve considering, discussing and documenting the wishes of a child or young person as appropriate, and their parents or carers, for their future care. Where a child or young person lacks capacity their parents' wishes should drive this process, taking account of the best interests of their child. Parallel planning refers to the development of plans that allow for unpredictability in the course of the condition. Therefore thinking about a care plan should take place in anticipation of a change in the progression of the condition in the future.

The process of Advance Care Planning involves discussions with children and young people and their parents or carers about the goals and desired direction of their care, particularly with regard to end of life care. This comprises personalised as well as parallel planning for important stages when changes may occur. For the purpose of this guideline, we will refer throughout to Advance Care Planning. It typically covers their concerns and wishes about their care, including what should be done, where, how, when and by whom. Importantly, Advance Care Plans also consider what should not be done. An effective care plan allows care to be delivered according to the wishes of the child or young person and their parents or carers, allowing them to retain autonomy, to influence how they are looked after and what is done to them. The discussion around an Advance Care Plan provides a forum for honest and direct communication between members of the multidisciplinary team, the child or young person and their parents or carers. People can talk about their fears and uncertainties, ask questions and regain some control over what happens to them.

Currently, however, too often discussions about Advance Care Plans happen late in a person's illness, and may focus principally on medical issues, such as the withdrawing of limiting of life sustaining therapies, rather than taking a more individualised view of their care. This review seeks to explore the barriers and facilitators to the development of personalised care plans.

### 6.1.3 Description of clinical evidence

The aim of this review was to explore the positive and/or negative experiences and opinions of the child or young person with a life-limiting condition, and of their parents, families, carers and multidisciplinary teams. This was done so that personalised care plans (including parallel and advance) could be formulated for the last days of life, including planning the care of infants with life-limiting conditions. The resulting personalised care plans can then be used to improve current practice.

A search was carried out for studies that collected and analysed data qualitatively (with collection methods such as semi-structured interviews, focus groups and surveys with open-ended questions, and analysis which included thematic analysis, framework thematic analysis and content analysis. Survey studies which reported only descriptive data that were analysed quantitatively were excluded.

Given the nature of qualitative reviews, findings and themes are summarised from the literature and were not restricted to those identified as likely themes by the Committee. Themes identified by the Committee were: reluctance to include the child or parents or carers in decision-making; timing of planning; need for regular reviews; assessment of needs, professional roles; cultural, religious and ethical differences; dealing with uncertainty; and emotional burden.

While a search was carried out for general as well as advanced and parallel care planning (as set by the review protocol), the majority of evidence identified related to Advance Care Planning.

A total of 11 studies were identified for inclusion in this review. Of them:

- 5 studies focused on the perspective of parents caring for a child with a life-limiting condition or whose child had died due to life-limiting condition (Erby 2006, Hammes 2005, Hinds 2001, McHaffie 2001, Parker 1999)
- 2 studies focused on the perspective of healthcare professionals (El-Sayed 2013, Lotz 2015)
- 1 study involved children or young people living with a life-limiting condition (Dunsmore 1996)
- 1 study involved both the parents and the child or young person living with a life-limiting condition (Zwaanswijk 2007)
- 1 study involved both the parents and the child or young person living with a life-limiting condition, as well as the physicians involved in their care (Hinds 2005).

With regard to the countries in which studies were conducted:

- 2 studies were conducted in the UK (Mitchell 2005, McHaffie 2001)
- 2 in Australia (Dunsmore 1996, Parker 1999)
- 2 in the USA (Erby 2006, Hammes 2005)
- 1 in Canada (El-Sayed 2013)
- 1 in Germany (Lotz 2015)
- 1 in the Netherlands (Zwaanswijk 2007)
- 1 in both the USA and Australia (Hinds 2005)
- 1 in Australia, the USA and China (Hinds 2000)

Regarding the methodology of the studies, the majority collected data by interviewing the participants, although 1 used online focus groups (Zwaanswijk 2007). The most common data analysis method employed across studies was thematic analysis.

Evidence on all of the themes considered important by the Committee was identified, and a number of additional themes that emerged were also incorporated into the review. A summary of the included studies is provided in Table 23.

To include the views of children and young people with life-limiting conditions and direct experience of the health service in the UK, a focus group was commissioned specifically for this guideline. A description of how this research contributed to the recommendations has been added to ‘Linking evidence to recommendations’ in this chapter (see sections 6.2.8.2.1 and 6.2.8.5).

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix G.

To help present the findings, a theme map was generated that highlights the themes that emerged from studies (Figure 6). The theme map was drafted by 1 researcher from the guideline technical team, and the resulting framework themes was further shaped and, when necessary, re-classified through discussion with at least 1 other researcher. Due to the qualitative nature of these studies, evidence is summarised in adapted GRADE-CERQual tables and, therefore, there is no separate appendix provided for this.

### 6.1.41 Summary of included studies

A summary of the studies that were included in this review are presented in Table 23.

**Table 23: Summary of included studies**

| Study | Data collection methods | Population | Aim of the study | Comments |
|---|---|---|---|---|
| Dunsmore 1996 (Australia) | Self-administered questionnaire with closed- and open-ended items | N = 51 young people with cancer<br>• Young people's age (mean, range): 18 (15 to 24) years | To identify information support and decision-making needs and preferences of young people with cancer. | • This study includes indirect population, as participants' age ranged from 15 to 24.<br>• Sample selection was limited to young people who attended a summer camp.<br>• The relationship between the researcher and respondents was not indicated.<br>• Researchers did not discuss saturation of data.<br>• The results were presented in a descriptive manner; thematic analysis would have been more appropriate. |
| Erby 2006 (USA) | Interviews | N = 19 parents of children and young people with Duchenne muscular dystrophy<br>• Children and young people age range: 8 to 27 years | To explore the attitudes and experiences of parents of children and adolescents with Duchenne muscular dystrophy regarding clinical management options and Advance Care Planning (ACP) | • Children's age range outside of scope.<br>• The aims were too broad and did not only focus on planning.<br>• The relationship between the researcher and respondents was not indicated.<br>• Data was presented to support the findings, but it was unclear if saturation has been achieved.<br>• Hypothesis-generating model. |
| El Sayed 2013 (Canada) | Interviews | N=12<br>Postgraduate physician trainees in | To explore the challenges for trainees when end of life | • The study included international physician trainees. |

| Study | Data collection methods | Population | Aim of the study | Comments |
|---|---|---|---|---|
| | | neonatology<br>• Neonates | decisions are undertaken, and to encourage them to reflect on how they might influence such decision-making. | • The response rate was quite low (12 out of 25), and the relationship between the researcher and the respondents was unclear.<br>• Thematic analysis was described, but researchers did not discuss saturation of data. |
| Hammes 2005 (USA) | Interviews | N=12 families of children with neuro-degenerative conditions (13 interviews in total, because 1 father and 1 mother were interviewed separately)<br>• Children's age at signing of AD (median/ range): 4.89 years (2 days to 12 years old) | To describe the process and population involved in paediatric ACP and to discuss the parents' perceptions of the planning process. | • The study included a small sample size, with children with neuro-degenerative conditions only.<br>• Participants were identified for inclusion by the existence of an AD, and it is not known how many parents may have declined to consider an AD. Five families (1/3) refused to participate.<br>• Thematic analysis was described, but researchers did not discuss saturation of data. |
| Hinds 2005 (USA and Australia) | Interviews | N children and young people= 20<br>N parents =19<br>N physicians = 16<br>• Children and young people age range: 10 to 20 years | To identify the preferences of children and adolescents with advanced cancer about their end of life care and the factors that influenced their decisions. | • Children's age range outside of scope.<br>• Included CYP, parents and physicians from 2 different countries.<br>• Cancer patients only.<br>• It was unclear whether the responders differed to those who were contacted but did not participate.<br>• The relationship between the researcher and the respondents was not indicated.<br>• Thematic analysis was described, but researchers did not discuss saturation of data. |
| Hinds 2000 (Australia, China and the USA) | Interviews | N = 43 parents of children and young people with cancer<br>• Children's age range: 1 year and 8 | To describe parental decision-making about treatment options for children with cancer, and determine the | • Children's age range outside of scope.<br>• Included patients from 3 different countries.<br>• Cancer patients only.<br>• The study included 4 different groups of parents, |

| Study | Data collection methods | Population | Aim of the study | Comments |
|---|---|---|---|---|
| | | months to 19 years and 11 months | feasibility of a similar but larger international study. | depending on the stage of the disease, so some of the evidence was considered indirect.<br>• Although the sample selection was described, it was unclear whether the patients who responded differed to those who were contacted but did not participate.<br>• The relationship between the researcher and the respondents was not indicated.<br>• Thematic analysis was described, but researchers did not discuss saturation of data. Themes were similar across groups, although it was expected that the parents would raise different issues depending on the stage of the disease. |
| Lotz 2015 (Germany) | Interviews | N=17 health care professionals (HCPs)<br>• Children's age: not indicated. | To investigate the attitudes, barriers and benefits as well as requirements for paediatric Advance Care Planning (ACP) from the view of HCPs, and to generate hypotheses on paediatric ACP that could be tested in a larger cohort. | • Paediatric population, age not indicated.<br>• Sample decisions were made a priori based on reasonable criteria rather than theoretical saturation (selective sampling).<br>• Response rate was 100%, but participants with no interest in the topic were excluded which may have biased the results.<br>• Data analysis was reported and thematic analysis was also described. |
| McHaffie 2001 (UK) | Interviews | N=108 parents/ 59 families of 62 babies<br>• Babies | To explore parent's perceptions of treatment withdrawal/ withholding, and their experience and opinions about this. | • Study with large, Scotland-based population, but results were mostly descriptive.<br>• Sample selection procedures were vaguely reported; it was unclear if all parents who lost a child were contacted.<br>• Data collection process was vaguely reported.<br>• Researchers did not discuss saturation of data.<br>• Unclear why interview data |

| Study | Data collection methods | Population | Aim of the study | Comments |
|---|---|---|---|---|
| | | | | was only analysed as frequency numbers or rates when a thematic analysis would have been more appropriate. |
| Mitchell 2005 (UK) | Interviews | N = 14 healthcare professionals<br>• Paediatric population in NICU and PICU; age not reported | To explore the experiences of senior medical and nursing staff regarding the challenges associated with ACP in relation to children and young people with life-limiting illnesses in the NICU/PICU environment and opportunities for improvement. | • The age was not indicated in the paediatric population.<br>• UK-based study.<br>• Conducted in NICU/PICU setting, limiting the generalisability of results to other settings.<br>• Thematic analysis was described. |
| Parker 1999 (Australia) | Interviews | N = 13 families (9 bereaved and 4 current families)<br>• Age of bereave children and young people range: 8 to 31 years | To examine the potential role for palliative care services in the care of individuals with muscular dystrophy and spinal muscular atrophy. | • Children age range out of the scope<br>• The aim is broadly described, but the study is not specifically aimed at looking at planning (indirect evidence).<br>• The authors used a convenience sample due to the low prevalence of the conditions, but they do not report the response rate.<br>• The data collection process is vaguely reported and saturation of data is not discussed. It is unclear why interview data was only analysed in a narrative manner, when a thematic analysis would have been more appropriate. |
| Zwaanswijk 2007 (Netherlands) | Focus group | N patients = 7<br>N parents = 11<br>• Children and young people age (mean, range): 11.6 (8–16) | To gain insight into the interpersonal, informational, and decisional preferences of participants involved in paediatric oncology. | • The study mainly focuses on communication as a way to enable their active participation in decision-making.<br>• It included only oncology patients, who were either survivors or in active treatment.<br>• The response rate was low (< 25%), although there were no differences regarding demographic characteristics. |

| Study | Data collection methods | Population | Aim of the study | Comments |
|---|---|---|---|---|
| | | | | • Authors used an online focus groups, using a recommended approach by a previous research group.<br>• The data analysis was reported; thematic analysis was also described. However, although the results were presented using relevant quotes from participants, a higher number of quotes would have been more useful to reflect the views of the participants in the different groups. |

### 6.1.5 Clinical evidence

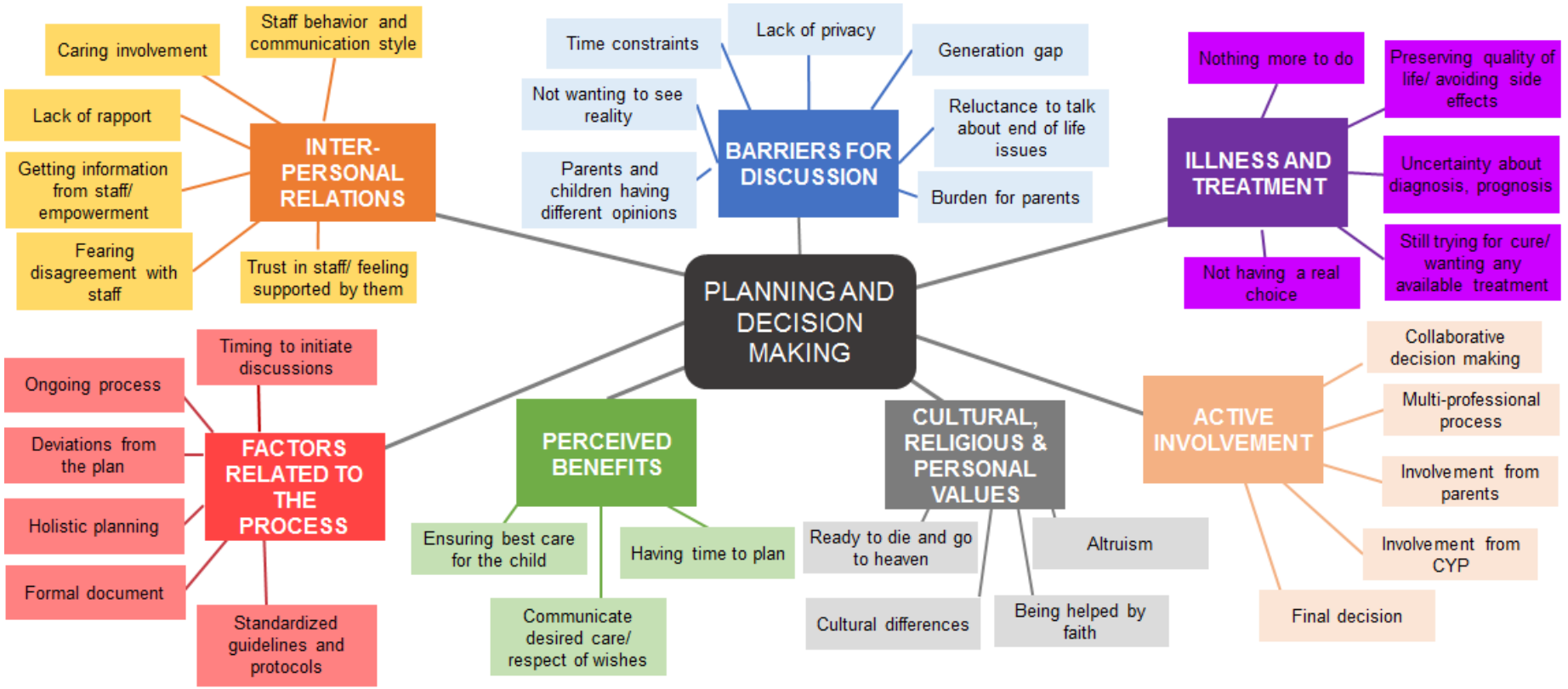
#### 6.1.5.1 Theme map

The theme map for Advance Care Planning is presented in Figure 6

#### 6.1.5.2 Clinical evidence profile

The clinical evidence (adapted GRADE-CERQual) for care planning is presented in Table 24, Table 25, Table 26, Table 27, Table 28, Table 29 and Table 30

**Figure 6: At the centre of the map is the overarching theme, which was mentioned as part of most of the other themes and subthemes, and relevant for Advance Care Planning**



1 **Table 24: Summary of clinical evidence (adapted GRADE-CERQual): Theme 1 – Factors related to the illness and the treatment**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: nothing more to do** | | | | | |
| 4 studies (Hinds 2000, Hinds 2005, McHaffie 2001, Mitchell 2015) | 4 interviews | 2 studies, conducted in Australia, China, UK and the USA, which included CYP with cancer, their parents and the physicians looking after them. 1 study was conducted in the UK with parents of neonates and 1 in the UK with healthcare professionals working in paediatric intensive care. They reported that the decision to stop treatment was related to the point when all people realised or understood that there was, most likely, nothing else that could be done:<br>• *"If anything was going to work, then would have done it before now." (16-year-old boy with a solid tumour)*<br>• *"We've been through the main steps…if it was going to work, probably it already would have worked. Stuff like the radiation, the chemo stuff, and then the experimental chemo, and we have been through several chemos and stuff, so I figured if it was going to slow it down or stop it, you know, it would have done it by now. And, if it hadn't done it by now, it's just going to grow, so I may as well be at home having fun." (17-year-old boy with a solid tumour).*<br>• *"We decided not to go with chemo because I don't want to be sick the rest of my days, and it's not like it is going to cure me, so I just said, 'we'll go home and take it from there." (15-year-old girl with acute lymphoblastic leukaemia)*<br>• *"often the nurses are way ahead of us, often the nurses are the people who suggest it … And sometimes it's us who realise. Sometimes it's the specialty consultants who realise enough is enough. It's rare for the families to suggest it, but I have had families suggest it to me." (doctor)*<br>• *"We had exhausted all of the conventionally useful drugs and experimental drugs." (physician)*<br>• *"parents who have seen their children having very umm, frightening events, life threatening events, cardiac arrest, the* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *parents that have witnessed a number of cardiopulmonary resuscitations, they'll get to a point where they can't watch it anymore." (doctor).* | | | |
| **Sub-theme 2: preserving quality of life/ avoiding adverse events from treatment** | | | | | |
| 3 studies (Hinds 2000, Hinds 2005, McHaffie 2001) | 3 interviews studies | 2 studies, conducted in Australia, China, UK and the USA, included CYP with cancer, their parents and the physicians looking after them. 1 UK study UK included parents of neonates. It was reported that children and parents contemplated the potential negative impact of certain drugs or therapies on the child:<br>• *"This would have meant extra days in the hospital…injections at home…probably less time off between treatments. He might not get the time to recuperate in between." (mother of a 14-year-old male with a solid tumour).*<br>• *"I knew it would make me a little bit sick and that I would be in the hospital for a few days each time. I could also have tried vincristine, but I had that before and I didn't think my body could get through that." (18-year-old male with a solid tumour).*<br>• *"It was explained to me that every new patient would get a stronger dose, every time. Mine would be the highest dose, and I could get all the symptoms the first day that others got on the 10th or 11th day." (18-year-old female with a solid tumour).*<br>• *"She would have an easier death than if we had done a lot of manipulation with machines." (physician).* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |
| **Sub-theme 3: still trying for cure/ wanting any available treatment** | | | | | |
| 3 studies (Hinds 2000, Hinds 2005, Parker 1999) | 3 interviews | 2 studies, conducted in Australia, China, UK and the USA, included CYP with cancer, their parents and the physicians looking after them. 1 Australian study included bereaved | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of | Coherent | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | parents of children with muscular dystrophy and spinal muscular atrophy. All reported that both parents and children wanted whatever treatment was available to them:<br>• *"We were kind of really happy that they had chemotherapy, something else that we could try." (15-year-old girl with a solid tumour).*<br>• *"I am…prolonging the inevitable until a cure comes along…I want her to be healed. I keep telling her to hold on…". (mother of a 14-year-old girl with a brain tumour).*<br>• *"In terms of what was available, this would be the one that could give him some potential help in controlling his tumour and pain relief…." (physician).* | findings | | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |
| **Sub-theme 4: not having a real choice** | | | | | |
| 3 studies (Dunsmore 1996, Hinds 2000, Zwaanswijk 2007) | 3 interviews | 3 studies conducted in Australia, China, the Netherlands and the USA, including CYP with cancer and parents reported that although more than 1 treatment option was available, only 1 option was seen as viable. It was either *"treatment or death".* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 5: uncertainty about diagnosis, prognosis** | | | | | |
| 4 studies (Lotz 2015, McHaffie 2001, Mitchel 2015, Zwaanswijk 2007) | 3 interviews, 1 focus group | 4 studies, conducted in the UK, the Netherlands and Germany, with healthcare professionals working in paediatrics, parents of neonates, and CYP with cancer and their parents. It was reported that the lack of diagnostic precision was an obstacle to undertaking ACP. In paediatrics, clear diagnoses frequently could not be made:<br>• *"On the other side, it is the experience that one can also mis-assess the situation, also in the negative sense. So, the* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *situations where one would have thought, based on experience, that this cannot turn out well, they have stabilised once again […] Therefore, one is very cautious. You first have to come to the point for yourself when you say: o.k., I really don't see, to the very best of my knowledge and belief, any chances left." (Intensive care physician)*<br>In 1 study conducted in the UK with parents of neonates, it was reported that parents are able to tolerate a degree of uncertainty and they demonstrate trust in the expertise of senior clinicians. Some parents also showed doubts (after child passing away) due to the lack of concrete evidence of a bleak outcome. If parents can be shown abnormal scan results... the accuracy of medical assessment is reinforced. | saturation | | |

**Table 25: Summary of clinical evidence (adapted GRADE-CERQual): Theme 2 – Active involvement in decision-making**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: collaborative decision-making** | | | | | |
| 2 studies (Dunsmore 1996, Zwaanswijk 2007) | 1 interview, 1 focus group | 1 study, conducted in the Netherlands, reported that both the CYP with cancer and their parents preferred decisions about treatment to be made in collaboration between patients, parents, and healthcare providers.<br>Likewise, 1 Australian study with young people with cancer reported that, in general, young people preferred to have discussions with professionals with parents present. Variability was highlighted, because some wanted to limit the discussion to physicians and themselves, others wanted to make decisions independently, and a few indicated that they did not | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | want to be involved. | | | |
| **Sub-theme 2: multi-professional process** | | | | | |
| 3 studies (El-Sayed 2013, Lotz 2015, Mitchel 2015) | 3 interviews | 2 studies, conducted in the UK and Germany with healthcare professionals working in paediatrics, reported that ACP was seen as a multi-professional process that should include all relevant HCPs in the community.<br>Studies frequently raised some aspects that affect staff involvement.<br>In 2 studies, 1 conducted in the UK and 1 in Canada participants described experiences where gaining consensus among the healthcare professionals involved had been a significant barrier to the advance care planning process:<br>• *"[W]e can be a lot more proactive given the opportunity, but often we're just trying to, er, persuade our colleagues who are providing care at the time, long before I see admission [to PICU], to raise the point." (Doctor)*<br>• *"[B]efore you can convince any parents, you have to convince the other specialties. You have to bring them on board. If they're not on board, you have no chance, or your chances with the family are much, much less." (Doctor)*<br>• *"In the end, no one should feel like he/she made the decision. It is a shared decision". When there is divergence of opinion, it leads to trainee anxiety and they often had trouble going forward with the proposed plan as this trainee shared. It is only when I'm able to establish consensus from my whole team that I will go ahead. Then…I know that I am not the only one, the whole team has decided. I am a representative of the team and…I establish some balance of dealing with this issue, so I do not get into that kind of distress which I used to get." (Trainee in neonatology).*<br>In 2 studies conducted in Canada and Germany, healthcare professionals in neonatology and paediatrics felt that the lack | Limitation of evidence | No limitations | MODERATE |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | of coordination was also an issue. They mentioned insufficient information-sharing between HCPs, lack of round tables and lack of a continuous contact person.<br>In 2 studies conducted in Canada and Germany, healthcare professionals in neonatology and paediatrics believed it was very important to receive formal training in end of life care, to reduce the many uncertainties of ACP. They particularly stressed their need for education about the legal situation and for training in communication skills:<br>• *"There should be more training, more mock cases, more sessions on how to manage end of life, which is not easy and we encounter every single day." (trainee in neonatology).*<br><br>In 1 study conducted in Canada, trainees in neonatology raised their need to manage personal internal conflict and separate their personal beliefs when decision-making with parents:<br>• *"It is something I have to deal with. I've learned to actually withdraw my own personal religion from whatever decision that is made. I've had to."*<br>• *"I put it in the back burner. I say: 'This is the way I am going to deal with it and hopefully I'll be forgiven in whatever decision it will have to be."*<br>The emotional impact on staff of frequently witnessing death was described, but was more widely recognised and managed by nursing staff compared with their medical colleagues:<br>• *"death is difficult and it is emotive and upsetting, but at the same time it is unavoidable, we have to deal with it." (D8)*<br>• *"when I was a registrar it was easier for me because I had to just sit and have a debrief with my … consultant, as I love to cry. But now, I have to be this brave person and it's very … very difficult." (doctor).*<br>• *"It's not real life what's going on in there, it's just so horrendous what is happening every single day … so 'No,* | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *you're not doing it today. You've done it a couple of times recently and that is enough'. Because if you do it too often you have to leave. You have to protect yourself." (nurse).* | | | |
| **Sub-theme 3: involvement from parents** | | | | | |
| 2 studies (El-Sayed 2013, McHaffie 2001) | 2 interviews | 1 UK study with parents of neonates reported that most parents would want to be involved, whereas some others would prefer not to take part. In practice, many parents felt that they took responsibility for decision-making, either jointly with doctors or on their own. Those who felt they were not involved, subsequently wished that they had taken responsibility for the decision at least in part.<br>In 1 study conducted in Canada with trainees in neonatology, they suggested that a degree of provider recommendation and parental guidance would be helpful without necessarily shielding parents from any unpleasant information or taking over their decisions.<br>• *"I think sometimes we can be a little bit more definite in our guidance because that is a big decision for parents to actually make and to feel like they have to make. I don't think that is something I could ever decide to do. I don't even have kids and I can't imagine being told "Go home and think about it. Come and tell us what your decision is." (trainee in neonatology).* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: involvement from children and young people** | | | | | |
| 6 studies (Dunsmore 1996, Hinds 2000, Hinds 2005, Lotz 2015, Parker 1999, Zwaanswijk | 5 interviews, 1 focus group | In 1 study conducted in Australia with young people with cancer, it was noted that there was some variability about their preference related to active involvement. In general young people believed that they should not make the decisions on their own, but a few believed that they should make the decisions themselves. A few indicated that they did not want to | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| 2007) | | be involved at all.<br>Similarly, in 1 study conducted in the Netherlands with CYP with cancer and their parents, some young children (aged 10 years) expressed a preference for a passive role in making major decisions on treatment whereas other children did want to take part in decisions.<br><br>In 2 studies conducted in the USA, China and Australia, parents of CYP with cancer **wanted to plan everything in accordance to the child's expressed preferences**:<br>• *"I talked with my child about what to do if we ever faced that decision, and I knew ahead of time what she wanted me to do, and that helped. I know I was doing what she would have wanted." (mother of a 12-year-old girl with a brain tumour).*<br><br>However, 1 study conducted in Germany with healthcare professionals working in paediatrics reported that it was difficult to know what the child's wishes are.<br><br>In 1 study conducted in Australia, bereaved parents of CYP with muscular dystrophy and spinal muscular atrophy found it difficult to initiate discussions concerning emergency care and treatment decisions with their sons, and this was interpreted as an implicit rejection of the use of advance directives:<br>• *"we never talk much about the future, especially with him, he never asks for it. Sometimes I probe a little but he really doesn't really want to, so it is an unwritten law: we never talk about it. I am quite sure he knows what is happening, but we never talk about it and I believe in that". (bereaved parent of a child with muscular dystrophy).*<br>In 1 study conducted in Australia and the USA, CYP with cancer and their physicians described that their decisions were influenced by family preferences. | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | • *"…the father identified that it was important that they try everything that was a potential benefit. That was important for both the son and the father, but especially the father." (physician).*<br>• *"If I don't take it, my family would support me, but they don't want me to quit. Grandpa said he would worry himself to death if I don't try it. My boyfriend wants me to take it for him. I don't want to do it but for my family." (female with a solid tumour).*<br><br>Also, in 1 study conducted in Australia with young people with cancer, some said they had considered giving up treatment, but some said they had no say, either through physicians or their parents, and that they had simply been told that their treatment should continue.<br><br>The age of the child was a recurrent theme in many studies.<br>In 1 study conducted in Germany, healthcare professionals working in paediatrics stressed that all children should be informed in an age-appropriate way about the decisions made (for example, using children's stories). It was reported that their treatment preferences should be considered regardless of age:<br>• *"If the patient himself says he wants this and this and that, no matter how old the child or adolescent is, when he can express it I think it has to be considered." (Outpatient nurse).*<br><br>In another study conducted in the Netherlands with CYP with cancer and their parents, some children referred to the importance of patient age in determining the appropriate level of CYP's participation in the decision-making process:<br>• *"[...] if you're older than fifteen, you're allowed to have a say in the decision and to decide for yourself sometimes. If you're younger than fifteen, you should decide together. I think* | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *children younger than fifteen don't really know what's good and bad for them" (Survivor, aged 17).*<br>In 1 study conducted in the Netherlands with CYP with cancer and their parents, it was reported that sometimes the child or young person was too ill or depressed to decide. | | | |
| **Sub-them 5: final decision/ sign the document** | | | | | |
| 2 studies (Lotz 2015, Zwaanswijk 2007) | 1 interview, 1 focus group | In 1 study conducted in the Netherlands with CYP with cancer and their parents, it was reported that even though parents could be of assistance in reaching a decision, young people emphasised that they should be the ones to have the final say.<br>In another study conducted in Germany, some healthcare professionals working in paediatric care said that everyone who has attended the discussions and are relevant to the individual case should confirm their consent to the decisions with their signature. Most healthcare professionals stressed the importance of a physician signing the advanced directives to validate it medically, while some considered it sufficient to certify that informed consent had been given (that is, that the CYP/parents were fully informed and given enough time to reflect their values and preferences in order to come to an informed decision). The interviewees also disagreed on whether the parents should always sign an advanced directive or whether only the CYP should be given the option to do so (given the high burden of responsibility). | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |

**Table 26: Summary of clinical evidence (adapted GRADE-CERQual): Theme 3 – Interpersonal relations**

| Study information | Description of theme or finding | Quality assessment |
|---|---|---|

| Number of studies | Design | | Criteria | Rating | Overall |
|---|---|---|---|---|---|
| **Sub-theme 1: trust in staff/ feeling supported by them** | | | | | |
| 4 studies (Dunsmore 1996, Hinds 2000, Hinds 2005, Zwaanswijk 2007) | 3 interviews, 1 focus group | 2 studies, conducted in Australia, USA and China, reported parents of children with cancer their feelings being sustained by the healthcare team's obvious concerns for their child, and by the team's continuous efforts to cure their child's disease, and allowed the HCPs to guide them in decision-making.<br>They also reported that staff listened to the CYP or parents' concerns and responded to them, explained situations or conditions in a compassionate and easy-to-understand way, or made efforts to secure the needed information:<br>• *"Nobody on the staff there is going to think that I made the wrong decision. They always made me feel like I did the right thing for my child." (father of a 13-year-old boy with leukaemia).*<br><br>Similarly, 1 study conducted in Australia with young people with cancer reported that the knowledge and professional expertise is the basis for confidence in health professional's skills and ability to make the 'right' decision on the patient's behalf.<br>On the other hand, in 1 study conducted in the Netherlands CYP with cancer and their parents raised that lack of trust in the physician's expertise was an important barrier. | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 2: staff behaviour and communication style** | | | | | |
| 3 studies (Dunsmore 1996, Hinds 2000, Parker 1999) | 3 interviews | Some styles of staff behaviour and communication were seen as facilitators whereas others were seen as barriers for decision-making.<br><br>In 1 study conducted in Australia with bereaved parents of CYP with muscular dystrophy and spinal muscular atrophy, how sensitive issues were approached was important. One parent commented that when their son had been seen by a | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | respiratory specialist regarding breathing difficulties, and options of care had been discussed, the specialists had been very blunt. Later, seeing another specialist who was *"much more gentle and less confronting"*, she felt her son was able to understand and make informed decisions regarding his future management.<br><br>In 1 study conducted in Australia with young people with cancer, interactional communication and the ability to allow and encourage feedback and questions, or professional friendship were reported as positive, as were expressions of genuine concern for the patient as an individual (not just as a disease), a sense of humour and a certain level of personal disclosure. On the other hand, an impersonal, detached or overly professional manner were viewed as uncaring and intimidating, as was the use of jargon and high-powered authoritarian behaviour, particularly the use of medical terminology, which respondents viewed as an attempt to keep them powerless.<br>.<br>In 1 study conducted in the USA, Australia and China with parents of children with cancer, parents described **feeling that they were being forced by staff** and of being made to choose a treatment option when they did not want to make the decision.<br>They also reported reacting negatively to the way in which options were offered or the abbreviated time frame in which the decision needed to be made (sense of urgency). | | | |
| **Sub-theme 3: getting information from staff/empowerment** | | | | | |
| 3 studies (Dunsmore 1996, El-Sayes 2013, | 3 interviews | 3 studies, 1 conducted in Australia, China and the USA with parents of children with cancer, 1 conducted in Australia with young people with cancer and 1 conducted in Canada with | Limitation of evidence<br>Coherence of | Major limitations<br>Unclear | VERY LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| Hinds 2000) | | trainees in neonatology, reported that honesty and a straight-forward approach was appreciated, as well as the provision of all cancer information including sensitive topics (for example, not being able to have children). They also reported that explanations from doctors and other professionals about certain symptoms or behaviours or updates and progress reports were quite useful in understanding their child's changing situation. | findings | | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: fearing disagreement with staff** | | | | | |
| 1 study (Hinds 2000) | 1 interview | In 1 study conducted in the USA, Australia and China, the parents of children with cancer wanted to avoid displeasing the healthcare team and by doing this they believed they would lose the team's support. | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 5: caring involvement** | | | | | |
| 1 study (Hinds 2000) | 1 interview | In 1 study conducted in the USA, Australia and China, the parents of children with cancer described being hampered in decision-making by the staff members with the strongest affection to their child and concern about how their child's death will affect staff. | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 6: lack of rapport with the family** | | | | | |
| 1 study (Mitchell 2015) | 1 interview | In 1 study conducted in the UK, healthcare professionals working in a PICO setting reported that not having an | Limitation of evidence | Minor limitations | LOW |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | | Criteria | Rating | Overall |
| | | established rapport with the family prior to raising the issue of end of life care for the first time during acute situations, including resuscitation, was a difficulty:<br>• *"it's very difficult for us, because it is usually in the acute settings, unusual that we even get an opportunity to speak to them before the breathing tube goes down." (doctor).*<br>• *"It [ACP] should have happened before they came to ICU, for a lot of children. And I know the challenge is that we never know when that end is going to be, but if the families have had no preparation that this is likely to take place, it's even harder." (nurse).*<br>• *"worst case scenario … would be [discussing end-of-life] right in the arrest situation … then you try to give parents the heads up about that, actually this is not really going to be a successful resuscitation, and to try to prepare them … in a very short space of time. Umm, depending on the parents that might be with them watching at the bedside." (doctor).* | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

**Table 27: Summary of clinical evidence (adapted GRADE-CERQual): Theme 4 – Cultural, religious and personal values**

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | | Criteria | Rating | Overall |
| **Sub-theme 1: deal with different cultures** | | | | | |
| 2 studies (El-Sayed 2013, Lotz 2015) | 2 interviews | 2 studies conducted in Canada and Germany with trainees in neonatology and healthcare professionals working in paediatrics reported on the need to learn about cultural expectations at the time of end of life discussions, as well as how to best support the cultural and religious needs of various | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | families. Recognising the importance of religion could be seen either as a barrier or as helpful:<br>• *"You will find people from every part of the world in Toronto so that makes it enriching for us as physicians, but sometimes difficult because you have to individualize each case according to the understanding which you grasp from the first meeting with parents. Difference would be the culture." (trainee in neonatology).*<br>• *"I try to avoid the babies whose parents have very strong religious beliefs because I don't know how to properly talk to them." (trainee in neonatology).*<br>• *"Many people think that if you involve God in this decision, then you might find it difficult… but if these parents have a strong belief in God or whatever that is then I think important to appreciate and to understand it." (trainee in neonatology).* | evidence | | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 2: being helped by my faith** | | | | | |
| 2 studies (Hinds 2000, Hinds 2005) | 2 interviews | 2 studies conducted in Australia, China and the USA with CYP with cancer, parents and healthcare professionals reported that for some parents, strength comes from spiritual beliefs and practices:<br>• *"I don't care what you want to call it, my belief had a lot to do with believing that there is something better out there for her." (mother of a 15-year-old girl with leukaemia).* | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: altruism/ helping others** | | | | | |
| 2 studies (Hinds 2000, Hinds 2005) | 2 interviews | 2 studies conducted in Australia, China and the USA with CYP with cancer, parents and healthcare professionals reported that in some cases parents' and young people's decisions were influenced by the possibility of helping others:<br>• *"What my daughter goes through would be very important to* | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of | Unclear | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | *another child. It's not just to save her but children in the near future that could possibly come down with this particular type of cancer…" (mother of a 17-year-old girl with a solid tumour).*<br>• *"Hopefully and in some way, we will be able to get through all this and go on with our life, but if it does not work out, well I want someone else to benefit." (mother of a 17-year-old girl with a brain tumour).*<br>• *"If I can help someone else, that's wonderful, I think." (14-year-old girl with a brain tumour).* | evidence | | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: ready to die and to go to heaven** | | | | | |
| 1 study (Hinds 2005) | 1 interview | In 1 study conducted in Australia and the USA with CYP with cancer reported on the certainty of living an afterlife that would be better than their current life circumstances:<br>• *"When the Lord is ready for you, you are going to leave. It doesn't matter if you are on a machine or not, you are going to leave." (20-year-old male with a solid tumour).* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | NA | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Unclear | |

**Table 28: Summary of clinical evidence (adapted GRADE-CERQual): Theme 5 – Factors related to the planning process**

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: timing to initiate discussion** | | | | | |
| 5 studies (Erby | 5 interviews | In 1 study conducted in the UK with healthcare professionals | Limitation of | Minor | VERY LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| 2005, Hinds 2005, Lotz 2015, Mitchell 2015, Parker 1999) | | working in paediatrics, most participants called for **early initiation of advance care planning** shortly after diagnosing an incurable condition. However, some professionals recognised that early initiation is unrealistic in many cases, because the parents often need considerable time to process the bad news. Therefore, they gave priority to the family's readiness for advance care planning **discussions**.<br><br>Similarly, another study conducted in Australia and the USA with physicians working in paediatrics, recognised that it is important that both the parents' and ill child's grasp of the **seriousness of the clinical situation** facilitates efforts to assist them with end of life decision-making:<br>• *"He has been very realistic about his situation, and that has helped me with this." (physician).*<br>In 3 studies conducted in UK, Germany and the USA with healthcare professionals working in PICU and paediatrics and parents of children with Duchenne Muscular Dystrophy, it was reported that conversations can be started as **specific life events**, such as deterioration of the child, home discharge, before admission to PICU, *"transitioning to a wheelchair*", *"getting a feeding tube"*:<br>• *"we get called in as intensive care doctors to help, er, the people who are managing the case long before a critical episode to talk through what a resuscitation would involve and what the treatment we provide involves. And that, um, parents will often agree in that situation that what we're contemplating doing is abhorrent in some way; you know it's just a step too far." (doctor).*<br>• "*In our community, people always ask, 'is he still walking?' I mean that is the BIG question ... because a lot of your issues medically that come up occur after the walking stops. I remember when he was really young, I would think to myself,* | evidence | limitations | |
| | | | Coherence of findings | Not coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *'well, let's see, one down, so we probably have about another four years before he stops walking." (mother of a 14 year old).*<br>• *"He was only 8 when this 13 year old boy died ... he wanted assurance that when he got to 13 that wasn't going to happen. So I think as he's going past 13 ... he's realized that it is very different for different people ... I mean this particular boy stopped walking at 9. And he knows that that is a big thing, a big benchmark for him ... as long as he is walking, he doesn't worry too much." (mother of 14 year old).*<br>• *"He is aware because a lot of the people that we know who have Duchenne's that are in our age group are getting tracheotomies, have night time breathing machines. So we do know that this is possibly in our future ... when he is at that point, I'm sure we will have discussions on those topics and give him time to make a decision on how he wants it ... I want him to be more involved. I think the awareness is there. The involvement is really not." (mother of an 18 year old).*<br>In 3 studies conducted in Australia and the USA with parents and children, it was raised that seeing **somebody else going through the same treatment or die** could open up an opportunity for discussion.<br>• *"Why would I want a tube in my throat? I saw two other patients like that – I don't want that. I wouldn't be able to talk with my family or hold my Mom's hand. That is not living." (15-year-old girl with acute lymphoblastic leukaemia).*<br>• *"Seeing other members of my family on tubes. You just lay there. I don't like it. I wouldn't want it for me. I don't want to be kept alive like that. If someone is ready to die, I say 'let them die,' you know?" (15-year-old with acute myeloid leukaemia).* | | | |
| **Sub-theme 2: ongoing process** | | | | | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| 1 study (Lotz 2015) | 1 interview | 1 study conducted in Germany with healthcare professionals working in paediatrics reported that advance care planning should be conceived as an ongoing process, adapted to the individual family. Decisions should be regularly reviewed. | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: deviation from the plan** | | | | | |
| 1 study (Mitchell 2015) | 1 interview | 1 study conducted in the UK with healthcare professionals working in PICU reported that acute clinical deteriorations often cause a crisis where even the most detailed advance care planning does not prevent a last-minute deviation from the plan:<br>• *"if they make an ACP they may still change their mind right at the very end, um … But at least they will have had the opportunity to sit down and seriously think about what they want for their child." (nurse).* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: holistic planning** | | | | | |
| 2 studies (Erby 2005, Lotz 2015) | 2 interviews | 1 study conducted in in Germany with healthcare professionals working in paediatrics reported that besides concrete emergency planning, there is a need to discuss daily life issues and plan for the end of life. This included planning of future support options in everyday life, dying, and bereavement.<br>Another study conducted in the USA with parents of children and young people with Duchenne Muscular Dystrophy also reported that family members did talk a great deal about their sons' futures in terms of education, career, and need for independence in the face of increasing disability.<br>• *"Well mainly, when he is 18 of course, we will have college* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *and if he goes to college, will he stay at home or will he live there. There are some schools now that offer residential service for kids like him that need nursing care, which he may or may not need at that point." (mother of a 14 year old)*<br>• *"We have talked about, 'You will go to college and grow up while you are at college' ... this is a rite of passage ... We are focusing now on things that he is good at and how could he make a living that will not be affected by his muscular dystrophy." (father of a 16 year old)* | | | |
| **Sub-theme 5: need for a formal document** | | | | | |
| 2 studies (Lotz 2015, Mitchell 2015) | 2 interviews | 1 study conducted in the UK with healthcare professionals working in PICU reported that the use of a formal document in advance care planning was generally regarded positively by participants, with perceived benefits including the provision of a framework for discussions, empowering both healthcare professionals and parents to agree a care plan which reflects the family's wishes:<br>• *"I'm going to see somebody on the ward that's collapsed and you're considering whether they need ICU, you know, to look in their medical records, you see the [ACP], and you can quickly identify rather than going through tons of medical notes to find out what's happening." (nurse)*<br>Also, another study conducted in Germany with healthcare professionals working in paediatrics pointed out that written documents should be distributed to emergency services and local hospitals to prepare them for potential emergency situations. | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 6: standardised guidelines/ protocols** | | | | | |
| 1 study (El-Sayed 2013) | 1 interview | 1 study conducted in Canada with trainees in neonatology recognised their wish to have standardised | Limitation of evidence | Minor limitations | LOW |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | | Criteria | Rating | Overall |
| | | guidelines/protocols for end of life care across NICUs. | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |

**Table 29: Summary of clinical evidence (adapted GRADE-CERQual): Theme 6 – Perceived benefits of having an Advance Directive (AD)**

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | | Criteria | Rating | Overall |
| **Sub-theme 1: ensuring best care for the child** | | | | | |
| 5 studies (El-Sayed 2013, Hammes 2005, Hinds 2005, Lotz 2015, Mitchell 2015) | 5 interviews | 2 studies conducted in Australia and the USA with parents of children with neurodegenerative conditions and cancer, and 3 studies conducted with healthcare professionals working in PICU, neonatology and paediatrics reported that planning helps to **preserve the quality of life** of the child and to avoid unnecessary treatments:<br>• *"I have very strong convictions about quantity versus quality. Deciding to go home – I'm just tickled…She is a whole different person." (Mother of a 15-year-old girl with leukaemia).*<br>• *"I have seen however many mothers here as well, who have never even held their baby, and the baby's stuck here with their chest open for three weeks, and then we finally withdraw care and they still haven't even held their baby." (doctor).*<br>• *"I don't think the meaning of life sustaining treatment is* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *always explained very well. How invasive it is and how uncomfortable, and how it takes you away from your normal environment, it takes you away from family interaction … lots of things we do carry significant risk of complications, and you should only really do them if, at the end of it, it is going to improve someone's quality of life."* (doctor).<br>• *Healthcare professionals were also able to recall instances where advancer care planning discussions had resulted in achieving a peaceful terminal phase of illness and death in a preferred place of care. Positive feedback had been given by parents at subsequent bereavement meetings:*<br>• *"I do believe it's helping. Well I know it is because I've seen parents coming back to us and talking about it, and saying how they feel it's, it's helped them." (nurse).*<br>• *"When he died I think it was all as sort of planned and predicted and … Yeah, the family were grateful, which is usually a good sign." (doctor).*<br><br>In 3 studies, healthcare professionals working in neonatology and paediatrics described the moral and emotional distress associated with the provision of care and interventions that were not felt to be in the best interests of the patient or their family: (when **ACP is inadequate**)<br>• *"we get faced with decisions that are out of our control, someone else has decided actually, either between the family and the team, the medical team, the nursing team, they have decided that this child needs to come to ICU, and it is taken out of our hands." (doctor).*<br>• *"I rather see the realistic situation in a way that you have a patient in the critical care unit where you have to painfully realize: this was somehow wrong, this won't work, ok? And THEN you say: Ok, now he is already here but we tie our own hands and say this and that we WILL NOT DO* | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *anymore." (Intensive care physician)*<br>• *"What I might interpret as bad or poor quality of life may not be the family's opinion…When they make the decision to continue treatment that personally this is a baby that I would withdraw on, I do feel bad about the situation thinking that this baby is going to continue really suffering, having pain. The family can't see it the way I'm seeing it." (trainee in neonatology).* | | | |
| **Sub-theme 2: having time to make decisions and plan** | | | | | |
| 2 studies (Hammes 2005, Mitchell 2015) | 2 interviews | In 1 study conducted in the UK with healthcare professionals working in PICU and 1 study conducted in the USA with parents of children with neurodegenerative conditions, ADs were seen as a tool that allows to made plans in anticipation for different scenarios:<br>• *"Sometimes they have quite specific needs that they, or specific wants, they want to, and you can't always facilitate them if you don't … know in advance." (nurse)*<br>• *"if we want to get this child home, you know, we bring the community teams in, meet the teams. If you want to take your child afterwards to a hospice, let's go … let's go to the hospice, let's go and see the bedroom, let's go and … it's just all about preparing them and getting the, to … just so that they're not frightened by – you know, new faces or different people." (nurse)* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: helps to communicate desired care/ respect of children's and parent's wishes/ sense of control** | | | | | |
| 2 studies (Hammes 2005, Lotz 2015) | 2 interviews | 1 study conducted in the USA with parents of children with neurodegenerative conditions and 1 study conducted in Germany with healthcare professionals working in paediatrics, it was reported that having ADs ensures respect of children's and parent's wishes and avoids confusion and conflicts | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of | Applicable | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | between physicians and carers:<br>• *"I think it can take the burden off the parents to a certain degree, and this having-to-be present all the time as well. This family for example would really love to go on vacation for a week. But then they say, in fact they don't dare to, because surely he will be hospitalized then […] So there is this fear: the moment I turn my back on the nurses, they do what in fact we don't want." (Primary care physician).*<br>• *"So, that they then know exactly what has been discussed, what was decided. To have it in black and white […] it also conveys, I believe, additional security, so you know: It is all right if I do NOT dial the emergency/critical care number now so somebody gets here because it's getting critical. It's all right the way it is." (Nurse in a special nursing facility).*<br>• • *"It's important to establish at least a little bit of clarity for the staff, for the parents, just what common goal is pursued and also which measures ARE taken and which are simply omitted. Insofar, I just think it is really IMPORTANT and makes a whole lot of sense for everyone involved with the child. Therapists included, doctors, nurses, parents. Just to always provide clarity and to just fix one GUIDELINE. Otherwise everyone is always very INSECURE in their doing and acting, and this just provides clarity and thus security." (nurse in a special nursing facility)* | evidence<br>Sufficiency or saturation | <br>Not saturated | |

**Table 30: Summary of clinical evidence (GRADE-CERQual): Theme 7. Barriers for discussing Advance Directives (ADs)**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: time constraints** | | | | | |
| 2 studies (Dunsmore 1996, Mitchell 2015) | 2 interviews | 1 study conducted in the UK with healthcare professionals working in PICU and 1 conducted in Australia with young people with cancer, raised the lack of time for discussion. | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 2: lack of privacy** | | | | | |
| 1 study (Dunsmore 1996) | 1 interview | In 1 study conducted in Australia with young people with cancer, it was reported that they would like this discussions with their physician were conducted in private, rather than during public ward rounds. Many respondents commented about this large public ward rounds, which made them feel stupid and invaded their privacy. | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: not wanting to see reality** | | | | | |
| 2 studies (Erby 2005, Lotz 2015) | 2 interviews | In 1 study conducted in Germany physicians and nurses expressed their own reluctance to address end of life issues with the patient/parents because they tend *to "close their eyes to the facts and don't want to picture the worst case scenario"* (Intensive care physician).<br><br>Similarly, in 1 study conducted in the USA with parents of children with Duchenne Muscular Dystrophy, parents reported that children themselves often dictated when they needed to separate themselves from the muscular dystrophy community, and this distancing may diminish opportunities for families to | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | discuss issues relevant to planning for future quality of life.<br>• *"Now, he got to a point where he said, 'I don't want to go to camp anymore.' So I said, 'Well, can you tell me why.' So he just said, 'I just don't want to be with other children that have the same thing as me." (mother of an 8 year old)*<br>• *"I mean he is very low key about his role as it related to the MDA and I think he had the opportunity to [play a prominent role], but he said, 'No, let somebody else do it. I want to get back to being with my buddies and my family again.' So... he is very cognizant of how others feel about him... perceive him; and, I think in two ways, the disease and then the notoriety that goes along with him having already played a prominent role."* (father of a 16 year old) | | | |
| **Sub-theme 4: burden for parents** | | | | | |
| 1 study (Lotz 2015) | 1 interview | In 1 study conducted in Germany, physicians raised they are afraid of taking away hope, forcing and overburdening both the parents and the patient as well as destroying the trusting relationship with the family. Also it is a responsibility for parents when they sign the AD for their child. | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 5: reluctance to talk about end of life issues** | | | | | |
| 3 studies (El-Sayed 2013, Erby 2005, Parker 1999) | 3 interviews | In 1 study conducted in the USA with parents of children with Duchenne Muscular Dystrophy, it was reported that parents wanted to delay having discussion about end of life care issues:<br>• *"... and I guess, in this household, it is always we will cross that bridge when we come to it. Yeah, the disease is progressing and he is not as strong as he once was, but he is still okay ... . so if I don't have to deal with it, then why deal* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *with it." (mother of a 16 year old)*<br>• *"I am very vague on what an advance directive is ... I think it has to do with like a DNR? I have not discussed that with him because we're not there yet. We are not even close to being there." (mother of an 18 year old)*<br><br>In 1 study conducted in Canada with trainees in neonatology, they commented that withdrawal of nutrition and hydration was the hardest for them to participate in:<br>• *"The nutrition thing I'm not comfortable with at all. I have been here for two years and I've heard a lot about it. Now I can hear…but I'm still not comfortable doing it and I don't think I'll be doing it. I'm not at that stage yet." (trainee).*<br><br>Also in 1 study conducted in the Australia that included bereaved parents of children and young people with muscular dystrophy and spinal muscular atrophy, it was reported that children are reluctant to talk about end of life issues and they just want to *"live for the moment"*. | saturation | | |
| **Sub-theme 6: parents and children having different opinions** | | | | | |
| 1 study (Mitchell 2015) | 1 interview | 1 study conducted in the UK with healthcare professionals who worked at PICU reported situations where the patient was a young person with capacity who wished to be involved in his or her own care planning, including difficult scenarios where the opinion of the child differed from that of his or her parents:<br>• *"She herself had her own end-of-life care programme for her in another hospital. Unfortunately when she deteriorated, the parents diverted to us … we didn't know about the end-of-life care plan. At the very best we knew from the way she looked that she had a life-limiting illness, but she was intubated. She woke up and she was extremely angry with the parents.* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | *Extremely angry." (doctor)* | | | |
| **Sub-theme 7: generation gap** | | | | | |
| 1 study (Dunsmore 1996) | 1 interview | 1 study conducted in Australia reported that young people with cancer perceive a generation gap: perceived discomfort of some health professionals when dealing with young people, especially regarding sensitive topics for example: *"too clinical", "too text-bookie", "humorless", "ancient", "stuffy"*. | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 8: lack of support in the community** | | | | | |
| 1 study (Hammes 2005) | 1 interview | In 1 study conducted in the USA with parents of children with neurodegenerative conditions, parents said that community members and other relatives did not support the idea of the child having an advanced directive.<br>• *"community though we were choosing whether the child lives or dies" (mother).* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Unclear | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |

### 6.1.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis.

### 6.1.7 Evidence statements

#### Illness and treatment

Very low to low evidence from 6 qualitative studies conducted with children and young people with life-limiting conditions, parents of children and young people with life-limiting conditions, and healthcare professionals looking after them, showed that the factors related to treatment and the illness, are important when making decisions related to end of life care. This was applicable to decisions related to stopping treatment, when they feel that there is nothing else to be done, and avoiding side effects from treatment and preserving quality of life is prioritised; or decisions regarding continuing treatment, when there is still a realistic prospect for longer survival. An uncertainty regarding prognosis was perceived by both parents and professionals as a possible barrier to effective decision-making about the care of the child or young person.

#### Active involvement

Very low to moderate quality evidence from 7 qualitative studies conducted with children and young people with life-limiting conditions, parents of children and young people with life-limiting conditions and healthcare professionals looking after them, looked at the importance of collaborative decision-making. Professionals felt that decision-making should be a multi-professional process and that consensus among professionals was needed. They showed some disagreement regarding the level of involvement of the child or young person, raising issues regarding age, and difficulty in knowing what their wishes were. They identified their own internal conflict as an important barrier, because they had to separate their beliefs when making decisions with the parents. Parents as well as the children or young people felt that they should be involved in the process, although some noted that they did not want to take part, or that they felt too responsible. In particular, children and young people described that they wanted to take a more active role, but they also highlighted that some children may be too young or too ill to make decisions about their care.

#### Inter-personal relations

Very low to low quality evidence from 7 qualitative studies taking into account the perspectives of children and young people with life-limiting conditions, parents of children and young people with life-limiting conditions and healthcare professionals looking after them, indicated that the relationship with professionals is very important. Trusting staff, feeling supported by them and getting information that is honest and straight-forward was described as helping parents and young people make decisions regarding treatment. Parents and children also pointed out that the way they were approached by staff could act as either a facilitator or a barrier. They disliked a sense of urgency or feeling forced to make a decision. An impersonal and detached professional manner was also viewed as intimidating.

#### Cultural, religious and personal values

Very low quality evidence from 6 qualitative studies conducted with children and young people with life-limiting conditions, parents of children and young people with life-limiting conditions and the healthcare professionals looking after them, reflected on the importance of cultural and religious aspects. Dealing with different cultures and religious backgrounds was seen as an issue by some healthcare professionals, whereas others found it helpful.

Some parents and children referred to faith being a source of strength in difficult situations. Altruism and the idea of helping others also influenced decision-making for both parents and children.

**Factors related to the decision making process**

Very low to low quality evidence from 6 qualitative studies conducted with parents of children and young people with life-limiting conditions and healthcare professionals looking after them, indicated that there were factors regarding the decision-making process that could facilitate or hinder discussions. The timing of initiating discussions was an important aspect for both parents and healthcare professionals. While some healthcare professionals supported early initiation of discussion shortly after diagnosis, others gave priority to parents or carers readiness. Specific events were seen as a prompt for discussion, such as a deterioration in the condition, getting a feeding tube, or seeing someone else going through the same treatment. Other important aspects mentioned by healthcare professionals were that planning should be an ongoing process and that deviations from the plan should be allowed.

**Perceived benefits of having an Advance Directive**

Low to moderate quality evidence from 5 qualitative studies conducted with parents of children and young people with life-limiting conditions and healthcare professionals looking after them, reported that perceiving benefits of having an Advance Directive (AD) facilitated discussions about end of life care. According to both parents and professionals, having an AD ensured the best care for the child, avoiding unnecessary treatments, and helped parents to communicate desired care. It also allowed for their wishes to be respected, giving them sense of control, and allowed for time to plan and think ahead, in anticipation of different scenarios.

**Barriers for discussing Advance Directives**

Very low to low quality evidence from 6 qualitative studies, conducted with children and young people with life-limiting conditions, parents of children and young people with life-limiting conditions and the healthcare professionals looking after them, reported on a number of barriers to the discussion of Advance Care Planning. The most significant barrier according to parents and professionals, was the reluctance of parents and children to talk about end of life issues and accepting the professionals' view that now is an appropriate time to talk about it, or accepting that there may be benefits in talking about it. Professionals reported situations in which parents and children had different opinions from each other, and they also identified time constraints as an issue. On the other hand, young people noted their discomfort due to the lack of privacy when discussing decisions regarding their treatment, as well as a perceived generation gap.

## 6.1.8 Linking evidence to recommendations

### 6.1.8.1 Relative value placed on the themes considered

Although themes were mainly identified from the literature, the Committee identified some expected themes that they thought would be important during the protocol stage. They agreed that the following themes would provide useful perspectives: the involvement of the child and/or the parents or carers in all decisions in the development of plans; timing of planning; the need for regular reviews; the assessment of needs; professional roles; cultural, religious and spiritual differences; dealing with uncertainty; and the emotional burden associated with making end of life decisions. One of the main themes identified described

particular barriers to effective shared decision making and the Committee considered this theme and its subthemes to be particularly important (e.g. ‘differences in opinion’, ‘time constraints’, ‘lack of privacy’).

#### 6.1.8.2 Consideration of barriers and facilitators

An important aspect to note about this review was that all the evidence identified was related to the concept of Advance Care Planning, and not the day-to-day clinical management.

The Committee discussed the importance and the benefits of having an Advance Care Plan, as well as the drawbacks and the considerations for implementation. In light of uncertainties with regard to future treatments or prognosis, the group agreed that the Advance Care Plan needed to include sufficient flexibility to provide options if changes occurred (parallel planning) and to allow regular reviews of the plan as necessary.

As pointed out in the literature, the Committee agreed that developing an Advance Care Plan provides all involved with the opportunity to talk about the future, and to consider all aspects of management. The Advance Care Plan should be discussed and developed in partnership with the child or young person and their parents. The Committee made specific recommendations regarding this collaborative approach. Sharing the Advance Care Plan among all relevant healthcare professionals and settings was clinically important.

Although it was not specifically addressed in the literature, the Committee recognised the importance of assessing the needs of the child or young person and their parents or carers. Regular reviews should be carried out. The importance of revisiting the plan was therefore discussed at length. It was also important that the Advance Care Plan is not ‘set in stone’, and that it can be changed whenever necessary. The preferences of the child or young person and their parents or carers need to be weighed up in light of what may be in the best interest of the child or young person, particular when their condition or other circumstances change.

Based on the available evidence, the Committee emphasised the need to consider in the Advance Care Plan the information and approach to communication with the child or young person and the parent or carer as the end of life. They stressed the importance of informing parents or carers of the care and support they could access or receive at that time. This should be initiated as early as possible, taking account of the family members’ personal needs and feelings.

Healthcare professionals should provide honest information regarding the prognosis and the treatments available to the child or young person and their families or carers to facilitate decision-making. In case of uncertainty about prognosis, this should also be discussed.

##### *6.1.8.2.1 Barriers and facilitators highlighted in the TFSL report*

There were 5 main themes that emerged from the focus group interviews on the topic of care planning. All of them were considered in the discussion on the recommendations on the Advance Care Plan and influenced what was drafted. The main themes were ‘ambiguity and variation’ which referred to children expressing different levels of understanding about the care plan process. Another theme was ‘sharing information about me’ where children explained that they had to repeat themselves and that plans were not shared between professionals. Other children were reluctant to make plans and wanted to live in the present (‘getting on with it’). Only some children and young people told the researchers that they had an Advance Care Plan that was updated or reviewed. This was captured in a theme called ‘managing disruption and change’. There were other children that did not know whether they had an Advance Care Plan or whether there was a care plan or they did not mention it (this was a theme referred to as ‘other’).Advance Care Plan

#### 6.1.8.3 Economic considerations

The Advance Care Plan is intended to be an evolving document that allows shared decision-making and helps healthcare professionals fulfil the wishes of children, young people and their families. Aspects of the Advance Care Plan may suggest when and what interventions should be used and thus do carry a cost component. However, the use of Advance Care Planning is already a recognised component of current practice in the NHS and the recommendations in this guideline largely relate to the principles that should inform the Advance Care Plan and the content it should include. Therefore, the recommendations themselves do not have important resource implications and, to the extent that the Advance Care Plan improves the experience and outcomes of care for the child or young person, then the Committee agreed that their recommendations would promote efficient resource use within the NHS.

#### 6.1.8.4 Quality of evidence

Moderate to very low quality evidence was presented in the review. The main reasons leading to downgrading the evidence included limitations in how the data were collected, a low response rate from participants, self-selection bias, and an awareness that people who chose to participate may differ from those who refused to be interviewed. On the other hand, in some studies participants were selected by the physicians who provided care to the child, and those who were not selected may have provided a different perspective.

Another reason was the lack of the critical review of the researcher's role in sample recruitment, data collection or the data analysis process. None of the studies clearly reported the relationship between researchers, interviewers and the respondents, whether the researchers had a pre-understanding about the topic or the possible influence of that in data collection and the analytical process. Lack of verification of findings was not reported either in any of the studies.

Some of the studies reported data in a descriptive fashion only, when thematic analysis would have been more appropriate and informative. Among those studies where thematic analysis was done, the authors did not always report in detail how findings/themes were derived or emerged from the data in their research.

The findings were on the whole coherent, and any differences in opinions were well explained. However, sometimes the evidence was not directly applicable to the guideline population. There were 2 reasons why this was a problem. Some of the studies included children and young people and parents of children and young people with a life-threatening condition, but not approaching the end of life (for example children with cancer, but receiving treatment aimed at cure). There were also 2 studies that included young people over the age of 18. Efforts were made to only include quotes from people up to 18 years of age, but it was not always indicated in the text.

It was noted by the Committee that some of the evidence related to advanced directives, and these are not relevant to the UK setting, as they are not directly applicable to children. However, the evidence was not downgraded further as the Committee agreed that the evidence from these studies could be extrapolated to Advance Care Planning in general.

Furthermore, it was unclear whether the data was sufficiently saturated which means that not enough detail was provided and the views were not explored in detail. The majority of the studies did not report whether saturation was achieved in terms of data collection or data analysis, and it was difficult to ascertain from the information reported. When considering the evidence as a whole, it did not appear to be very saturated, as many themes were just raised in 1 study and there were few quotes to support them.

The Committee had more confidence in the findings from the focus groups that were carried out for this guideline, both in terms of methodological robustness and applicability.

#### 6.1.8.5 Other considerations

Based on their experience, the Committee considered that an Advance Care Plan should include the following sections:

- Demographic information about the child or young person as well as their family or carers, and contact details for the child or young person, the family or carers and the healthcare professionals looking after the child
- Information about the child's condition
- Details regarding the child's wishes, as well as the wishes of their parents or carers
- Records of the discussions between with the child and/ or their parents or carers in relation to treatment plans, place of care, withdrawal of treatment, parallel planning or funeral arrangements.

The Committee discussed that Advance Care Plans need to be flexible and therefore it is important that they are regularly reviewed, for example if the child moves to a different setting or when there are significant changes in the child's condition.

Special emphasis was placed on the importance of sharing the Advance Care Plan with relevant healthcare professionals, such as GPs, consultants, community nursing teams, or hospice staff, as well as with others relevant to the care of the child. It was highlighted that it is important that the plan should be transferable to other settings and regions. This was also supported by the literature.

In relation to the people that should be involved in developing the Advance Care Plan, the Committee agreed, based on the evidence from a number of themes and subthemes, that it is important that this needs to be a collaborative process in which all relevant professionals involved in the care of the child or young person take part, as well as child or young person and their parents or carers. Discussion about Advance Care Plan issues can be burdensome for the families, so it should be dealt with in a sensitive manner and at the appropriate time. In this sense, it was discussed that the level of involvement may vary from 1 family to another, or even during the course of the illness, and that healthcare professionals should respect the parents or carers' wishes on this regard. However, the Committee agreed that involvement should be strongly encouraged so that their preferences are known and documented. The family's values and their cultural and/ or religious background should also be taken into account when discussing aspects related to treatment, in particular regarding issues such as withdrawing treatment.

It was also agreed that it is important to involve the child or the young person, using appropriate language for their age and their condition. Professionals should also be aware of the possibility of disagreements between the child or young person and their parents about decisions regarding their care. This would need careful consideration by the healthcare professional to support them to reach an agreement.

Finally it was also mentioned that it is important to follow the Advance Care Plan unless for some reason it transpired that it was in some respect no longer in the child or young person's best interest.

The Committee agreed it was vitally important to make sure that healthcare professionals did not mistakenly believe that an Advance Care Plan was a statement of intent 'not to treat'. They therefore made specific explanatory recommendations accordingly.

The Committee discussed whether they wanted to prioritise this topic for a research recommendation, but they concluded that the combination of the evidence (including the focus group report), their experience and their expertise was sufficient to base the recommendations on.

#### *6.1.8.5.1 Other considerations related to the TFSL focus group findings*

Children and young people in the focus group felt that they were knowledgeable about their condition, even more so than their parents. The young people who were interviewed were keen to take part in decision-making and planning but their preference of the level of involvement varied. It is also important not to make assumptions about the child or young person's preferences when developing their Advance Care Plan. The interviews highlighted that children and young people can be realistic, for instance, about times when they need to be in hospital, and therefore appropriate discussions should take place to involve them. The Committee noted that the young people were frustrated by having to repeatedly tell their story and by not being provided with individualised care that met their specific needs. These points resonated with the Committee members' experience and they therefore agreed it was important that recommendations were drafted to promote good practice.

### 6.1.8.6 Key conclusions

Based on the available qualitative evidence and findings from the focus group, the Committee concluded that planning, assessment and reviews go hand in hand. The development of the Advance Care Plan is individual and should take place in partnership with all relevant people (healthcare or other professionals, the child or young person and their parents or carers). Honest information regarding the prognosis and treatments available should be provided to the child or young person and their families or carers to facilitate decision-making. In case of uncertainty about prognosis, this should also be discussed.

## 6.1.9 Recommendations

**20. Recognise that children and young people with life-limiting conditions and their parents or carers have a central role in decision-making and care planning.**

**21. Regularly ask children and young people and their parents or carers how they want to be involved in making decisions about their care, because this varies between individuals, at different times, and depending on what decisions are being made.**

**22. Explain to children and young people and to their parents or carers that their contribution to decisions about their care is very important, but that they do not have to make decisions alone and the multidisciplinary team will be involved as well.**

**23. Manage transition from children's to adult's services in line with the NICE guideline on transition from children's to adult's services.**

**24. Develop and record an Advance Care Plan for the current and future care of each child or young person with a life-limiting condition. The Advance Care Plan should include:**

- demographic information about the child or young person and their family
- up-to-date contact information for:
  - the child or young person's parents or carers and
  - the key professionals involved in care
- a statement about who has responsibility for giving consent
- a summary of the life-limiting condition

- an agreed approach to communicating with and providing information to the child or young person and their parents or carers
- a statement covering what information about the child or young person and their parents or carers will be shared, and with whom
- an outline of the child or young person's life ambitions and wishes, for example on:
  - family and other relationships
  - social activities and participation
  - education
  - how to incorporate their religious, spiritual, and cultural beliefs and values into their care
- a record of significant discussions with the child or young person and their parents or carers
- agreed treatment plans and objectives
- education plans, if relevant
- a record of any discussions and decisions on
  - parallel planning of end of life care and medical care that is specifically for the underlying condition
  - the preferred place of care or place of death
  - organ and tissue donation (see 1.1)
  - management of life-threatening events, including plans for resuscitation or life support
  - specific wishes, for example on funeral arrangements and care of the body
- a distribution list for the Advance Care Plan.

**25. Begin discussing an Advance Care Plan with parents during the pregnancy if there is an antenatal diagnosis of a life-limiting condition.**

**26. Develop and regularly review Advance Care Plans:**

- with relevant members of the multidisciplinary team and
- in discussion with the child or young person and their parents or carers.

**27. Advance Care Plans should take account of the child's or young person's life as a whole.**

**28. When developing the Advance Care Plan, take account of the beliefs and values of the child or young person and their parents or carers.**

**29. Explain to children and young people and their parents or carers that Advance Care Planning should:**

- help them be involved in planning their care and give them time to think about their views carefully
- help them to understand the life-limiting condition and its management
- ensure that relevant professionals can plan, develop and implement a management plan for now and the future
- help to prepare for possible future difficulties or complications

- support continuity of care, for example if there are changes in the professionals involved or in the care setting (such as a hospital admission or discharge).

**30. Share the Advance Care Plan with the child or young person and their parents or carers, and with relevant professionals and services involved in their care, such as:**

- GPs
- hospital consultants
- hospices
- respite centres
- community nursing services
- their school and other education services
- ambulance services.

**31. Update the advance care plan when needed, for example if:**

- new professionals become involved
- the care setting changes (for example, hospital admission or discharge)
- the child or young person and their parents or carers move home.

**Discuss the changes with the child or young person (if appropriate) and their parents or carers.**

**32. Share the Advance Care Plan with everyone involved each time it is updated.**

**33. When making an Advance Care Plan, discuss with the child or young person and their parents or carers:**

- the nature of their life-limiting condition, its likely consequences and its prognosis
- the expected benefits and possible harms of the management options.

**34. Be aware that all children and young people with life-limiting conditions should have an Advance Care Plan in their medical record, and that this should not be confused with a do-not-resuscitate plan.**

**35. Be aware that any existing resuscitation plan for a child or young person may need to be changed in some circumstances, for example if they are undergoing general anaesthesia.**

**36. Never assume that there is a do-not-resuscitate plan in place for a child or young person unless this is explicitly stated in their record.**

**37. Be aware that discussing the Advance Care Plan can be distressing for children and young people who are approaching the end of life and their parents or carers, and they may:**

- be reluctant to think about end of life care
- have difficulties discussing end of life care with the professionals or with one another
- have differences of opinion about the care plan.

38. **When making or reviewing the Advance Care Plan for a child or young person approaching the end of life, talk to the parents or carers about the care and support they can expect when the child or young person dies. Discuss their personal needs and feelings about this.**

39. **When a child or young person is approaching the end of life, think about and discuss with them and their parents or carers their specific support needs. Review these needs regularly.**

## 6.2 Preferred place of care and place of death

### 6.2.1 Review question

**What preferences do children and young people with a life-limiting condition and their family members or carers have for place of care and for place of death, and what determines those preferences?**

### 6.2.2 Introduction

In the past, children and young people with life-limiting conditions had little choice in terms of place of care and more specifically place of death as this was invariably within the hospital setting. However, with the advent of children's hospices, increasing technology that can be used within the community and increasing levels of skill in palliative care within the community both medically and in terms of nursing, the choices of place of care and deaths have increased.

The recognition of the high cost of care within the hospital environment and the parallel recognition of its often unsatisfactory environment for families, has led to a preference among the healthcare community to try to care for the child and young person within the community. With the children's hospice movement providing medical and nursing support at little or no cost to the healthcare community (although increasingly NHS commissioners are funding these services in part) there has been a significant increase in children being cared for through charitable organisations.

In the midst of all these changes, the question arises about what the preferences are for the child or young person or their parents' in terms of care and place of death and what determines these preferences.

### 6.2.3 Description of clinical evidence

The aim of this review was firstly to ascertain the preferences that children and their parents or carers have for their place of care and place of death, and secondly to examine correlates of those preferences.

For the first part of the review question we looked for information that would indicate the preference for place of death provided by parents or carers. For the second part of the questions. To explore what would determine the choice of the place of death we also included qualitative findings from studies to understand the reasoning behind those choices. We identified 1 systematic review (Bluebond-Langner 2013). This included children but also young people up to the age of 25 years. It was therefore not updated because it did not fully fit our criteria, but included studies were ordered and cross-checked to identify those that would be appropriate according to our review protocol (see Appendix D). Two of the included studies were applicable, and 1 further study that was published since was identified (Hechler 2008; Kassam 2014; Vickers 2007). The main characteristics of these were as follows:

- Qualitative in design (including studies that utilised either interviews or surveys to collect information), specifically:
- 1 study used semi-structured interviews to collect information however only the frequencies of preferences were reported.
- 1 study asked parents to rank their preference based on hypothetical scenarios and only the ranking of preferences was reported.
- Another study used close-ended questionnaires to collect information from children with terminal cancer and their families on preferred place for death.
- The population in the first 2 studies was parents of children who had died of progressive cancer (1 conducted in Germany the other in Canada).
- The third study prospectively recruited children (and their families) with progressive cancer who were assessed as being terminal despite maximal therapy. This study was conducted in the UK.

Evidence that was relevant to the topic of this review was found, they were: preferred place of death, preferred place of care, change of preference over time, congruence between actual and preferred place of death, congruence between actual and preferred place of care, factors associated with congruence between actual and preferred location of death, factors associated with congruence between actual and preferred place of care, and information provided to parents about preferred place of care.

To include the views of children and young people with life-limiting conditions and direct experience of the health service in the UK, a focus group was commissioned specifically for this guideline. A description of how this research contributed to the recommendations is added to the Linking Evidence to Recommendation section of this chapter (see section 6.2.8)

A brief description of the studies is provided in Table 31**.** Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix G exclusion list in Appendix H and Focus group report in Appendix L. Due to the qualitative nature of these studies, evidence is summarised in adapted GRADE-CERQual tables within the evidence report. Therefore no separate Appendix is provided for this. Full details of the Focus Group can be found in Appendix L.

### 6.2.4 Summary of included studies

A summary of the studies that were included in this review are presented in Table 31.

**Table 31: Summary of included studies**

| Study | Data collection methods | Population of respondents | Aim of the study | Comments |
|---|---|---|---|---|
| Hechler 2008 | Interviews | Parents of 48 children who died from cancer (11 fathers and 45 mothers)<br>Germany | To investigate bereaved parents' perspective on 5 areas. One of these was 'characteristics of death' which included information on the place of death. | • For the purpose of this review only rates of preference were extracted ('in hindsight which locale of death would you regard now as most appropriate for your child').<br>• Unclear how long after the death of the child the interviews took place (possible recollection |

| Study | Data collection methods | Population of respondents | Aim of the study | Comments |
|---|---|---|---|---|
| | | | | bias).<br>• More than half of the children died in hospital (some in the intensive care unit).<br>• Even though described as an interview design only descriptive results were reported (rates of question responses rather than reasons for preferences) |
| Kassam 2014 | Descriptive (close ended) survey | Parents of children who died from cancer at least 6 months before enrolment (N=75)<br>Canada | To determine bereaved parent and clinician preferences for location to end of life care and death. | • Parental response rate was only about half of the participants (54%)<br>• The results of this study were based on a clinical vignette rather than on the parents' own experience. Parents were asked to rank order each setting, home, hospice and hospice separately as first, second or third choice.<br>• This study also included the views of clinicians but for the purpose of this review the results are not reported here. |
| Vickers 2007 | Descriptive (close ended) survey | All children (and their families) registered over a 7 month period through the United Kingdom Children's Cancer Study Group (UKCCSG) for whom in the view of the treating oncologist a cure was no longer possible because of recurrence /progression despite maximal therapy.<br>UK | To describe effectiveness of an outreach team model of palliative care in allowing home death for children with incurable cancer. | • The focus of this study was to investigate whether a type of service enables children to die in their preferred place of death rather than investigating the preference and reasons for the preference as such.<br>(N=185 of which data could be analysed from N=164 children and their families) |

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### 6.2.5 Clinical evidence

The clinical evidence profile for preferred place of death is presented in Table 32.

**Table 32: Summary of clinical evidence (adapted GRADE-CERQual)**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Preferred place of death** | | | | | |
| 3 studies (Hechler 2008; Kassam 2014; Vickers 2007) | 1 study used interviews and 2 studies used surveys | In 1 study from Germany, 88% of bereaved parents of children who had died from cancer reported that in hindsight chose 'home' as the most appropriate for their child to have died.<br>In 1 study from Canada bereaved parents of children who had died from cancer were asked about their preference place of death (based on a clinical vignette of descriptions of settings) 70.8% of parents ranked home as their first choice. Hospital was ranked as the first choice by 23.9% and hospice by 5.7% of parents.<br><br>In 1 study from the UK of children with terminal cancer and their families 68% recorded a preference for home deaths. | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Preferred place of care** | | | | | |
| 1 study (Kassam 2014) | 1 study used surveys | In 1 study from Canada bereaved parents of children who had died from cancer were asked about their preferred place of care (based on a clinical vignette of descriptions of settings) 57/72 (79.1%) of parents ranked home as their first choice, 11/72 (15.2%) ranked hospital as their first choice of care and 5/72 (6.9%) hospice. | Limitation of evidence | Major limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Change of preference over time** | | | | | |
| 1 study (Vickers | 1 study used | In 1 study from the UK of children with terminal cancer and | Limitation of | Minor | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| 2007) | surveys | their family reported 68% of home preference at the beginning of the study and 80% by the last month of life. | evidence | limitations | |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Congruence between actual and preferred place of death** | | | | | |
| 3 studies (Hechler 2008; Kassam 2014; Vickers 2007)<br>3 studies (Hechler 2008; Kassam 2014; Vickers 2007)<br>3 studies (Hechler 2008; Kassam 2014; Vickers 2007)<br>3 studies (Hechler 2008; Kassam 2014; Vickers 2007) | 1 study used interviews and 2 studies used surveys<br>1 study used interviews and 2 studies used surveys<br>1 study used interviews and 2 studies used surveys<br>1 study used interviews and 2 studies used surveys | In 1 study from German in which bereaved parents of children who had died of cancer were interviewed, 48% of children died at home even though 88% of the parents chose 'at home' as the most appropriate locale of death in hindsight.<br>In 1 study from Canada bereaved parents of children who had died from cancer were asked about their preference of place of death (based on a clinical vignette) and the actual place of their child's was also recorded. Of those who chose home as the preferred location of death 39/51 (76.1%) of their children had died at home; 16/17 (94.1%) who had ranked hospital as their first choice reported that their child had died in hospital; but none of the children whose parents had ranked hospice as their first choice had died in a hospice (0/4).<br>In 1 study from the UK of children with terminal cancer and their families, 120 of 140 for whom a preference for home death was recorded at any point actually died at home (86%).<br>In 1 study from German in which bereaved parents of children who had died of cancer were interviewed, 48% of children died at home even though 88% of the parents chose 'at home' as the most appropriate locale of death in hindsight.<br>In 1 study from Canada bereaved parents of children who had died from cancer were asked about their preference of place of | Limitation of evidence | Major limitations | VERY LOW<br>VERY LOW |
| | | | Coherence of findings | Not coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | death (based on a clinical vignette) and the actual place of their child's was also recorded. Of those who chose home as the preferred location of death 39/51 (76.1%) of their children had died at home; 16/17 (94.1%) who had ranked hospital as their first choice reported that their child had died in hospital; but none of the children whose parents had ranked hospice as their first choice had died in a hospice (0/4).<br>In 1 study from the UK of children with terminal cancer and their families, 120 of 140 for whom a preference for home death was recorded at any point actually died at home (86%).<br>In 1 study from German in which bereaved parents of children who had died of cancer were interviewed, 48% of children died at home even though 88% of the parents chose 'at home' as the most appropriate locale of death in hindsight.<br><br>In 1 study from Canada bereaved parents of children who had died from cancer were asked about their preference of place of death (based on a clinical vignette) and the actual place of their child's was also recorded. Of those who chose home as the preferred location of death 39/51 (76.1%) of their children had died at home; 16/17 (94.1%) who had ranked hospital as their first choice reported that their child had died in hospital; but none of the children whose parents had ranked hospice as their first choice had died in a hospice (0/4).<br>In 1 study from the UK of children with terminal cancer and their families, 120 of 140 for whom a preference for home death was recorded at any point actually died at home (86%).<br>In 1 study from German in which bereaved parents of children who had died of cancer were interviewed, 48% of children died at home even though 88% of the parents chose 'at home' as the most appropriate locale of death in hindsight.<br><br>In 1 study from Canada bereaved parents of children who had | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | died from cancer were asked about their preference of place of death (based on a clinical vignette) and the actual place of their child's was also recorded. Of those who chose home as the preferred location of death 39/51 (76.1%) of their children had died at home; 16/17 (94.1%) who had ranked hospital as their first choice reported that their child had died in hospital; but none of the children whose parents had ranked hospice as their first choice had died in a hospice (0/4).<br>In 1 study from the UK of children with terminal cancer and their families, 120 of 140 for whom a preference for home death was recorded at any point actually died at home (86%). | | | |
| **Congruence between actual and preferred place of care** | | | | | |
| 1 study (Kassam 2014) | 1 study used surveys | In 1 study from Canada bereaved parents of children who had died from cancer were asked about their preference of place of (based on a clinical vignette) and the actual place of their child's was also recorded. Of those who chose home as the preferred location death 48/51 (84.2%) of their children had been cared for at home; 7/11 (63.6%) who had ranked hospital as their first choice reported that their child had been cared for in hospital; but none of the children whose parents had ranked hospice as their first choice had been cared for in a hospice (0/5). | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Factors associated with congruence between actual and preferred location of death** | | | | | |
| 1 study (Kassam 2014) | 1 study used surveys | In 1 study from Canada bereaved parents of children who had died from cancer were asked about their preferred place of death and the actual place was also recorded. There were 2 characteristics independently associated with the likelihood of dying in the preferred location. The child having a hematologic malignancy decreased the likelihood whereas the involvement of a palliative care team increased the likelihood of dying in the preferred place. | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Not coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | | saturation | Not saturated | |
| **Factors associated with congruence between actual and preferred location of death** | | | | | |
| 1 study (Kassam 2014) | 1 study used surveys | In 1 study from Canada bereaved parents of children who had died from cancer were asked about their preference for location of care and the actual place was also recorded. There was only 1 variable that seemed to have some independent association with the likelihood of being care for in the preferred location. The involvement of a palliative care team increased the child's likelihood of being cared for in the preferred place. | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Not coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Information provided to parents about preferred place of care** | | | | | |
| 1 study (Hechler 2008) | 1 study used interviews | In a German study in which bereaved parents of children who had died of cancer were interviewed, almost half of the parents reported to have been informed of the possibility of palliative home care for their child. | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Not coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

## 6.2.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis.

## 6.2.7 Evidence statements

### 6.2.7.1 Preferred place of care and preferred place of death and change of preference over time

Very low quality evidence from 3 studies using interview or survey designs indicated that the majority of bereaved parents of children who had died of cancer, or families with children who have terminal cancer, prefer home to be the place for their child to die (68%–88%). Very low quality evidence from 1 of these studies indicated that home was also the highest ranked place for the child to be cared for (79.1%). Low quality evidence of 1 of the studies suggested that there was a 12% rise in home to be the preferred place of the child's death from entrance into the study to the last month before the child died (an increase from 68% to 80%).

### 6.2.7.2 Congruence between actual and preferred place of care and preferred place of death and factors associated with congruence

Very low quality evidence from 3 studies using interview or survey designs showed an inconsistent pattern for the congruence between actual and preferred place of death. In 1 study congruence was low (48% died at home even though 88% chose home in hindsight), whereas in the other 2 studies congruence was higher (76.1% and 86% of those whose parents or carers had indicated a preference for the child's death to be at home had died there). Congruence for a first choice of a death at hospital and the actual death having occurred there was high (94.1%), but none of the children whose parents had ranked hospice as their first choice had died there. Very low quality evidence from 1 of the studies indicated a high congruence between actual and preferred place of care for home, but this was lower for the hospital setting (84.2% and 63.6%, respectively). Whereas none of the children whose parents indicated hospice as their first choice of place of care had been cared for in this setting. Very low quality evidence also identified independent factors associated with whether or not children would die or were cared for at the preferred place. Having hematologic malignancy decreased the likelihood of dying at the preferred place, whereas the involvement of a palliative care team increased this likelihood. Only the involvement of a palliative care team increased the likelihood of the child having been cared for at their parents' preferred place of care.

### 6.2.7.3 Information provided to parents about preferred place of care

Very low quality evidence from 1 study using interviews with bereaved parents of children who had died of cancer, indicated that almost half of the parents had been informed of the possibility of palliative home care for their child.

## 6.2.8 Linking evidence to recommendations

### 6.2.8.1 Relative value placed on the themes considered

Determinants and correlates of preferred places for end of life care or death were considered critical outcomes for this review question. This was because of their high relevance in understanding reasons behind preferences and what could influence those preferences.

### 6.2.8.2 Consideration of barriers and facilitators

The Committee discussed the available evidence and the information related to benefits and harms from the available study data. They noted that congruence between the actual place of death and the stated preference for place of death was often low. The reasons for this were not described in detail in the studies, and in any event these studies were from countries where clinical services may have differed significantly from those in England. In the evidence the most frequently reported preference of children and young people as well of families was to be cared for and to die in their own home. The Committee discussed the fact that preferences may change during the course of a child or young person's illness. This was noted also in the evidence, the wish to be at home during the final phase of the illness being more often stated towards the end of life. For each individual family and child or young person a range of factors might influence the preferred place or care or death, including their own views and feelings, the support available to them, unsuccessful symptom control, often inadequate pain control, religious / social issues, and the nature of their clinical condition.

#### *6.2.8.2.1 Barriers and facilitators highlighted in the TFSL report*

Regarding place of care, evidence identified from TFSL's report (which was NICE commissioned and conducted among children and young people living with life-limiting conditions in the UK) showed that the majority of children and young people living with life-limiting conditions expressed that they preferred to stay at home when possible. Home was where participants felt the most relaxed, and where equipment and facilities were adapted to their specific needs. However they also pointed out that when being cared for at home they hoped to have better access to appropriate medical care.

Regarding hospices, participants in this study who had regularly stayed at hospices thought that although hospices resembled some best aspects of home, it wasn't home. It was highlighted by children and young people interviewed that having home comforts and technologies around them and things to do, were important to them, and this impacted on their experience of staying in hospital or at a hospice.

Regarding being cared for in hospital, the children and young people interviewed stressed the importance of feeling safe and looked after, and they felt they did not always experience this in hospital. Young people thought hospitals could be more aware of their needs, including their technological needs to help them stay connected to their friends during frequent or prolonged stays and, in some situations, their need for parents or carers to be present. On the other hand, a few participants reported that going into hospital eased the pressure on their parents to look after them when they were unwell.

It is important that this study found that children and young people, although not wanting to spend time in hospital, acknowledged that sometimes hospital was the preferred place of care because of the specialist medical expertise, tests, treatments, and medicines available. This environment could then have a reassuring effect on young people when they were unwell.

### 6.2.8.3 Economic considerations

There is a clear benefit to children and young people in providing services that facilitate their preferred place of care and place of death. Sometimes it may be necessary to provide rapid transfer from one setting to another to achieve this, which incurs costs associated with transfer, as discussed in Section 7.3.8.3. Where home is the preferred setting then there will be costs associated with providing day and night nursing support and day and night specialist advice (see Section 7.3.8.3). The recommendations do note that in order to facilitate home care, home adaptations, changes to living arrangements and equipment may be necessary. There is a cost to this but there is also likely to be some off-setting reduction in hospital costs as a consequence of reduced hospital admission.

#### 6.2.8.4 Quality of evidence

Low to very low quality of evidence was identified in this review. The quality of evidence was lower because only survey data on frequencies of preferences were reported in studies, and there was a lack of qualitative data analysis. The UK-based study was relatively large, however its data were not informative because the objective of the study was to assess the effectiveness of an outreach team model of palliative care to enable children to die at the preferred place of death, which was not directly applicable to our context. The Committee concluded that the evidence was not very useful in informing their recommendations due to its generally low quality. The Committee concluded that the evidence was not very useful in informing their recommendations due to its generally low quality as well. It was also noted that the included studies focused on children and young people with cancer, however cancer accounts for only about a quarter of those needing end of life care in the UK. Importantly the Committee also agreed that the preference for both place of care and death could be informed by the specific illness or condition and could change quickly with symptoms evolving. The Committee recognised that evidence indicating the main determinants for preferred place care or death, and especially the reasons underlying the stated preferences, were likely to be difficult to find in clinical studies. A wide range of factors would likely influence such preferences, and these were not readily identifiable in close-ended survey studies. It was important to understand whether the preferences expressed were determined by an awareness that resources were lacking to support the true preference for the child's place of death. It would be important to know what a child, young person or their parents preferred in ideal circumstances, and to understand what specific factors then influenced their choice. They might prefer home, but choose hospital as the preferred place of death if they believed they would not be adequately cared for at home, as indicated in the evidence.

A directly applicable study was conducted for this guideline to directly address the views of children and young people with life-limiting conditions in the UK setting. Regarding place for care, findings from TFSL's report indicated that children and young people preferred to be cared at home because they felt they had home comforts and technology (for example, Wi-Fi connection) around them and things to do. However, they also explained that sometimes they preferred hospital stay because of the availability of specialised medical care there when they were unwell. Due to ethical and practical reasons, it had been decided that this commissioned study would be focused on place for care rather than preferred place of death when interviewing children and young people living with life-limiting conditions.

#### 6.2.8.5 Other considerations

Due to the lack of directly relevant qualitative evidence and evidence quality, the Committee based the recommendations mainly on findings of the focus group report and on their experience and expertise in the area.

The Committee thought it was important that healthcare professionals discussed with the child or young person (if appropriate) and with their parents or carers their feelings and views on place of care and death. The decision on the preferred place should be based on a realistic appraisal of the individual circumstances and needs, and should take account of their feelings on the matter. When this was agreed this should be recorded as part of Advance Care Planning. The Committee agreed that it was important to recognise and to make clear in the discussion that such decisions were provisional and the preferred place might change for a variety of reasons.

The Committee discussed that an overriding principle should be regular communication in an individualised way for each family (see also the chapters on information, communication and care planning). Symptoms could evolve and therefore some choices could become inappropriate for the child or young person at different stages of their care. Importantly, it was clear that the child or young person and their parents or carers could change their mind about the place of care or death. The Committee also considered related evidence that this

could change if the child or young person or their parents or carers change their minds, if clinical needs evolve and change, and especially if there are service difficulties such as lack of day and night community support.

The Committee thought that ideally children and young people should be cared for and die in the place they (depending on their capacity to make these choices) or their parents or carers preferred, subject to other factors such as the trajectory of the condition, changes in care needs, and service availability.

The Committee emphasised that it was important for healthcare professionals to document preferred place of care and death in the Advance Care Plan. When developing and reviewing the Advance Care Plan, the child or young person and their parents' or carers' views should be explored and incorporated, as well as the input from healthcare professionals involved in the MDT caring for the child. Also, it was imperative that the Advance Care Plan should be regularly reviewed, taking into account the child's or young person's disease trajectory, their circumstances and possible needs in each stage of their illness.

The Committee also discussed the fact that the child or young person and the parents or carers should understand that while their choices were of central importance in determining the preferred place of care or death, they would not be expected to come to this decision alone or unsupported. The decision should be agreed in partnership with the relevant healthcare professionals in the multidisciplinary team.

The Committee also noted that evidence based on studies conducted among adults has shown that an important issue during end of life care was pain management. The group therefore agreed that this would also be a priority in consideration for children and young people because the place of care may have an impact on pain management (see symptom management question).

#### *6.2.8.5.1 Other considerations related to the TFSL focus group findings*

Regarding preferred place of care, the Committee noted that findings from the study commissioned and conducted among children and young people living with life-limiting conditions in the UK was largely consistent with what was indicated in the limited evidence identified from the literature review. This showed that although children and young people preferred to be cared for at home because of the familiarity, easy access to technology, and equipment for their specific needs, the majority of them also preferred to be cared for at hospitals when they felt unwell because of the availability of specialised medical care and the reassuring feelings that brought to them. These points were taken into consideration by the Committee when they wrote the recommendations.

The Committee discussed that there was a gap in directly applicable evidence that would inform choices in place of care and place of death. They therefore decided that future research would be important to inform guidance in future.

### 6.2.8.6 Key conclusions

The Committee concluded that possible options regarding preferred place for care/death should be explained and discussed with the child/young person and with their parents or carers as appropriate. Ideally the wishes of the child or young people and of their parents or carers should be met if this is possible. It was important to understand however that a range of factors needed to be taken into consideration, including their clinical needs as well as service availability. Moreover, decisions would need to be reviewed at intervals and when circumstances demanded it. In the study conducted for this guideline, children and young people understood that there would be situations when it would be in their best interest to be in hospital. Regular discussion and good communication and planning were paramount.

The Committee emphasised the importance of decision-making in partnership with the families of children and young people about places for care and death, to reduce the burden of the responsibility. An active effort should be made to establish the wishes of the child or young person if they have the capacity to make this choice. In addition, the Committee concluded that preferred place of care or death should be documented in the Advance Care Plan and reviewed regularly, taking into account of the child or young person's condition, the overall circumstances and needs of the child or young person and their family.

### 6.2.9 Recommendations

**40. Discuss with children and young people with life-limiting conditions and their parents or carers where they would prefer to be cared for and where they would prefer to die.**

**41. Agree the preferred place of care and place of death with children and young people and their parents or carers, taking into account:**

- their wishes, which are personal and individual
- their religious, spiritual and cultural values
- the views of relevant and experienced healthcare professionals
- safety and practicality.

**42. If possible, services should ensure that children and young people can be cared for at their preferred place of care and die at their preferred place of death.**

**43. Explain that the place of care or place of death may change, for example:**

- if the child or young person and their parents or carers change their minds or
- for clinical reasons or
- due to problems with service provision.

### 6.2.10 Research recommendations

**1. When planning and managing end of life care, what factors help children and young people with life-limiting conditions and their parents or carers to decide where they would like end of life care to be provided and where they prefer to die?**

| Research question | When planning and managing end of life care, what factors help children and young people with life-limiting conditions and their parents or carers to decide where they would like end of life care to be provided and where they prefer to die? |
|---|---|
| Why this is needed | |
| Importance to patients, service users or the population | In the past, CYP with life-limiting conditions had little choice in terms of place of care and more specifically place of death as this was invariably within the hospital setting. However, with the advent of children's hospices, increasing technology that can be used within the community, and increasing levels of skill in palliative care within the community (both medically and in terms of nursing), the choices for place of care and death have increased. In order to be able to offer this choice, it will need to be established what factors parents view as the most important in influencing their decisions. |
| Relevance to NICE guidance | High: there is very limited research available on where children and their parents want end of life care to happen. This research will inform the direction of future developments. |

| Research question | When planning and managing end of life care, what factors help children and young people with life-limiting conditions and their parents or carers to decide where they would like end of life care to be provided and where they prefer to die? |
|---|---|
| Relevance to the NHS, public health, social care and voluntary sectors | An evidence based understanding about the factors that influence parents managing the end of their child's life will allow focused service development into the areas that will support these choices. Caring for CYP at their or their parents' preferred place and having appropriate support to do so, will increase parental quality of life and satisfaction with service. Being proactive about this in care planning and enabling this preference to be achieved should therefore be of relevance as well as be a priority for the NHS. |
| National priorities | Several government reviews have highlighted the need for better support and choice in palliative care. These are for instance:<br>Craft, A. & Killen, S. 2007. Palliative care services for children and young people in England: an independent review for the Secretary of State for Health.<br>Department of Health. 2006. Our health, our care, our say: a new direction for community services: A brief guide Available: http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_4127602<br>Department of Health. 2008. End of Life Care Strategy – promoting high quality care for all adults at the end of life Available: http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_4127602 |
| Current evidence base | There is very limited published evidence on preferences for end of life care in children. |
| Equality | This research would address the current inequality by providing evidence behind choices for children and their families. |
| Feasibility | Access to children and their families would be needed at a sensitive time. This could be managed by using practitioners known to the families. Sensitively conducted qualitative studies may be able to address the reasoning behind the preferences. |

# 6.3 Organ and tissue donation

## 6.3.1 Review question

**What aspects of communication and information provision facilitate or hinder discussions between children and young people with a life-limiting illness and their family members or carers with healthcare professionals to make decisions on organ or tissue donation?**

## 6.3.2 Introduction

Organ transplantation is a widely accepted life-saving intervention for people with end-stage organ failure. Currently there are many more people who might benefit from organ and tissue donation than there are existing donors. The decision by the child or young person, their families and their carers to consider the donation of organs or tissue, can result in direct benefit for other patients and in positive memories of their child's legacy through organ and tissue donation.

Not all children are able to donate, even if this is the wish of the child or young person or their parents or carers. The child may have received aggressive medical therapies, or have suffered from an illness that precludes successful transplantation of their organs. In addition, organ and tissue donation is only possible when a child dies in hospital, so children who, for example, die at home will not be able to become donors. Tissue donation or donation for research may be alternatives under such circumstances.

Although approaching the parents or carers of a potential organ or tissue donor can be challenging for clinicians, with compassion and sensitivity, discussions about donation can be an integral part of end of life care planning. Where this is consistent with the child, young person or their parents' wishes, values and beliefs discussions should take place in a timely manner. The principles that should be maintained are those of being sensitive to the family's needs for time and privacy, and providing them with sufficient information in an understandable format which anticipates their likely concerns, giving them a realistic impression of whether donation will be possible, and if so, what it will entail.

This review seeks to determine the aspects of communication that support or hinder children and young people and their families and carers in making decisions about organ or tissue donation.

## 6.3.3 Description of clinical evidence

The aim of this review was to identify what aspects of communication and information provision influence the attitudes of the parents or carers of a child or young person with a life-limiting condition towards organ and tissue donation.

We looked for studies that collected data using qualitative methods (such as semi-structured interviews, focus groups, and surveys with open-ended questions) and analysed data qualitatively (such as thematic analysis, descriptive phenomenology, content analysis and so on). Survey studies restricted to reporting descriptive data that were analysed quantitatively were excluded.

Given the nature of qualitative reviews, findings/themes are summarised from the literature and were not restricted to those identified as likely themes by the Committee (including altruism, organ and tissue donation as part of care plan, religious beliefs, family influences, impact on siblings, cultural influences, body integrity, death rituals and so on).

Only 1 study conducted in the USA was identified for inclusion in this review. A total of 13 parents were interviewed and their experiences of consenting or not consenting to donate their child's organs after the child's death was described. Thematic analysis was used to analyse the qualitative data in the study.

A brief description of the studies is provided in Table 33

To include the views of children and young people with life-limiting conditions and direct experience of the health service in the UK, a focus group was commissioned specifically for this guideline. However, due to ethical and practical reasons, the Committee decided not to directly ask children and young people living with life-limiting conditions about tissue or organ donation during interviews. Therefore this topic was not covered in this research.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix and in the GRADE profiles below and in Appendix J. For presentation of findings, 2 theme maps were generated according to the themes emerged from studies (Figure 7 and Figure 8).

The mapping part of the review was drafted by 1 researcher but the final framework of themes was further shaped and when necessary re-classified through discussions with at least 1 other researcher. Due to the qualitative nature of these studies, evidence is summarised in adapted GRADE-CERQual tables within the evidence report. Therefore no separate Appendix is provided for this.

### 6.3.4 Summary of included studies

A summary of the included study is presented in Table 33.

**Table 33: Summary of included studies**

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| Hoover 2014 | Interviews | N=13 parents (11 consented to donate their child's organs, 2 did not consent) | To describe parents' experience of organ donation decision-making in the case of donation after circulatory determination of death. | • Parents were recruited from a single children's hospital in the Western USA<br>• The majority of parents (11/13) were European American and Christian (9/13). |

This study reported on 2 main themes/categories:

- factors that contributed to parents' decision-making regarding the donation of their child's organs
- factors that facilitated their communication with the healthcare professionals about their child's organ donation, or those that could improve their experience in the process.

### 6.3.5 Clinical evidence

The clinical evidence profile for facilitators and barriers for organ and tissue donation of the child or young person living with life-limiting conditions is presented in Table 34 and Table 35 (adapted GRADE-CERQual Tables for qualitative findings).

#### 6.3.5.1 Theme maps: Figure 7 and Figure 8

**Figure 7: Theme map 1: Individual reasons/factors contributing to organ donation of the child**

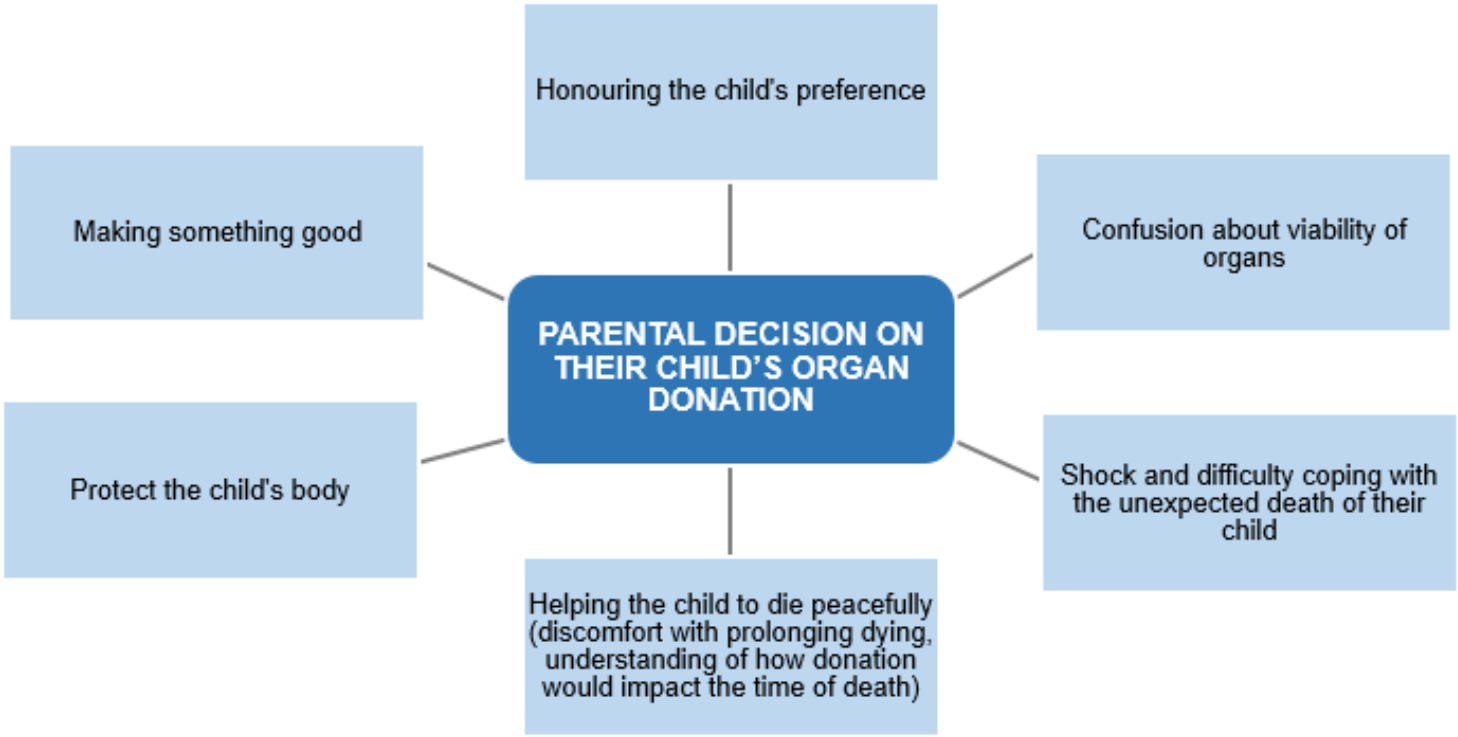
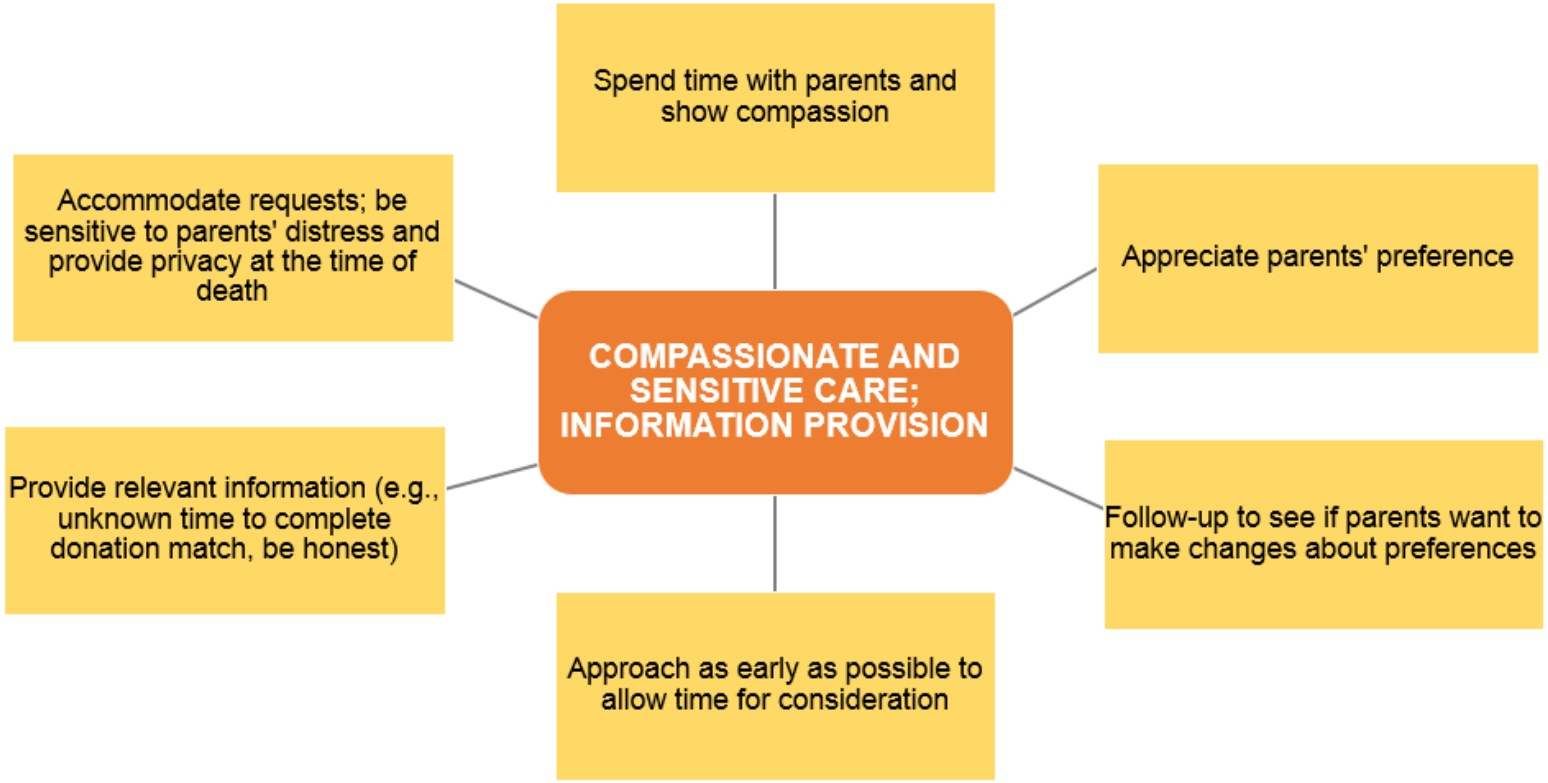


**Figure 8: Theme map 2: compassionate and sensitive care**




### 6.3.6 Evidence Summary

**Table 34: Summary of evidence (adapted GRADE-CERQual): Theme 1 – Individual reasons/factor**

| Study information | Description of theme or finding | Quality assessment |
|---|---|---|

| Number of studies | Design | | Criteria | Rating | Overall |
|---|---|---|---|---|---|
| **Subtheme 1: Factors that contributed to parental decision-making** | | | | | |
| 1 study (Hoover 2014) | 1 study used interviews; | In 1 study where parents were interviewed they reported reasons that contributed to their decision of donating their child's organs after the child's death. These included:<br><br>**Wanting to making something good out of the tragedy of their child's death:**<br>*"I mean she meant a great deal to us, and I loved her with everything in me, but I wanted her to be able to make more of an impact on somebody else's life by being able to donate, something that we would save somebody, you know?"*<br><br>Similarly, another parent explained:<br>*"That was largely my reasoning for organ donation, because I was going to make sure that something good could come out of a tragedy."*<br><br>**Wanting to honour their child's preferences:**<br>In addition to parental desire to help others, many believed that their child would have wanted to help others. One parent shared, *"I think this is what she had wanted me to do for her."*<br>*"I know what I need to do. I've had this conversation with my son. I know what needs to be done."*<br>*"If he were able to talk, then he would have totally said, 'take everything.' I know that."*<br>In the decision to consent to donate, their child's stated preferences were honoured.<br><br>**Confusion about viability of organs:**<br>Several families had some difficulty understanding whether or not their child could donate certain organs due to the trauma they had suffered.<br>*"And it went over across our mind a little bit such trauma that I* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *don't think that it would have been good at that point, you know, because they had to do CPR on her several times, I just didn't feel that that was the way to go, but I wanted to do her kidneys and her liver."*<br><br>In contrast, some parents assumed that donation was medically viable and then learned that it was not. Parent cited:<br>*"I mean [age] healthy younger girl, I mean you'd think after, you know, if someone needed a heart that that wouldn't-but I guess it has to be pretty, those things have to be pretty, they have to pretty careful."*<br><br>Another parent expressed her distress about learning that some organs could not be donated for transplantation.<br>*"I only thing I remember is that doctor...had told me that her body went without oxygen for so long that they would be afraid that they were too tainted to put into somebody else and so that they couldn't use her organs, and I remember that upset me, and I started crying."*<br><br>**Wanting to protect their child's body:**<br>Another factor that influenced parental decision-making was the desire to protect their child's body.<br>*"Because she she'd be through too much." …."When you're in this situation you're thinking, 'okay, she's going to have this casket and she's going to be in there, and I want her to be as pretty as she can for as broken and bruised as she is."*<br>Parents' desire to protect their child's body also influenced parents to limit specific organ donation. Many parents wanted their child's dead body to be *"whole."*<br><br>**Wanting to help their child die peacefully:** | | | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | | Criteria | Rating | Overall |
| | | This influenced how long parents were willing to maintain life support. When considering how to donate, some parents had discomfort regarding prolonging dying while seeking potential recipients.<br>*"just trying to get it done quick. We just didn't want to drag it out."*<br><br>**Shock and difficulty coping with the sudden unexpected death of the child:**<br>The factors that influenced the decision-making process revolved around the child's unexpected and sudden death. Parents emphasised their shock and difficulty coping with the sudden unexpected death of their child.<br>*"partially you never really think your kids are going to go before you. So you never think about it."*<br>*Another parent, when asked about the most difficult part of the decision to donate, she stated "letting her go."*<br><br>**Donation as meaningful contribution:**<br>*"She's living on in somebody else."*<br>Some parents expressed that donation to research was less satisfying:<br>*"I would definitely rather it goes to somebody than [be] used for research. But they also need research to make things better and to help somebody else.. [but], when it's used for research, it's done."* | | | |

**Table 35: Summary of evidence (adapted GRADE-CERQual): Subtheme 1 – Compassionate and sensitive care; information provision**

| Study information | Description of theme or finding | Quality assessment |
|---|---|---|

| Number of studies | Design | | Criteria | Rating | Overall |
|---|---|---|---|---|---|
| **Subtheme 1: Compassionate and sensitive care; information provision** | | | | | |
| 1 study (Hoover 2014) | 1 study used interviews; | Parents interviewed in 1 study offered recommendations about the donation process for HCPs involved in the process. Parents' recommendations were based by their experience and included those:<br><br>**Informed by positive experiences:**<br>• accommodate requests, such as spending time with deceased child after donation procedure<br>• HCPs spending time with parents and show compassion;<br>• allow parent to stay with their child throughout hospital experience;<br>• appreciate parents' preferences about organ donation; provide relevant information and updates, not providing irrelevant information; communicate honestly;<br><br>**Informed by improvable experiences:**<br>• Be sensitive to parents' distress and provide privacy at the time of death<br>• approach as early as possible to allow time to consider donation<br>• provide information about unknown time to complete donation match;<br>• follow-up to see if parents want to make changes about donation preferences after parents provide initial consent<br>• provide information about success rates and need for organ donation online;<br>• provide information about what kind of research is conducted with donated organs | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Unclear | |

1

### 6.3.7 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis.

### 6.3.8 Evidence statements

#### Factors that contributed to parental decision on their child's organ donation

In 1 study where parents were interviewed, they reported several Individual factors contributing to parental decision-making regarding whether to donate their child's organs. These included: making something good; honouring the child's preference; confusion about viability of organs, protecting the child's body, helping the child to die peacefully (low quality evidence).

#### Compassionate and sensitive care

In 1 study where parents were interviewed, they recommended that communication about organ and tissue donation should be delivered in a compassionate and sensitive way, as this would be helpful for them when approached and asked to make their decision. Specifically, their recommendations included: accommodating parents' requests, allowing parents time to stay with their child throughout the hospital experience, appreciating parents' preferences and following up to check whether parents want to make changes to these. They also recommended, being approached as early as possible about organ and tissue donation, and providing information about the unknown time to complete donation match (Low quality evidence).

### 6.3.9 Linking evidence to recommendations

#### 6.3.9.1 Relative value placed on the themes considered

The Committee agreed that all themes identified during the protocol stage were important to this review. These themes included the perspectives and experiences of the parents/carers of the children and young people living with life-limiting conditions, the children or young people living with life-limiting conditions, and the healthcare professionals involved in the end of life care. It was agreed that perspectives from different populations would each provide an important, informative and unique angle to this topic.

#### 6.3.9.2 Consideration of clinical benefits and harms

The evidence identified suggested a number of factors that contributed to parents/carers decision-making regarding whether to donate their child's organs or tissues. It also identified several factors that parents reported helpful in the process, if appreciated and accommodated by healthcare professionals. The Committee thought that the evidence was in accordance with their clinical observations particularly with regard to the emotional impact that organ donation can have (as in the theme 'making something good'). They commented that organ and tissue donation was about the wishes of the child and the parents or carers. The fact that many families asked in retrospect whether their child would have been suitable for donation, suggests that good communication and information provision is important and beneficial in this process. Only through this could the families' wishes or desires to become organ and tissue donors be identified and discussed, and the appropriateness of organ and tissue donation be assessed and explained.

The Committee commented that because organ and tissue donation is an emotive topic and because of the complexities involved when a child with life-limiting conditions is approaching

the end of life, healthcare professionals should explore the possibility of organ and tissue donation with the parents or carers in a sensitive and compassionate way, noting that it may not always be appropriate to ask parents or carers about this.

The Committee commented that it is important to find out whether the child is able to be a donor before raising the issue with child, young person or their parents or carers. If a request has already been made by the parents or carers, it is important to establish whether donation is possible given the child's circumstances and the other wishes around end of life care. The Committee considered it important to discuss with the child/young person as appropriate, and their families/carers, whether or not they could donate. When necessary, the specialist nurses for organ and tissue donation should be involved to help develop and convey the information, as some conditions or circumstances could preclude the possibility of organ and tissue donation.

The Committee noted that solid organ donation could only take place if death occurred in a setting able to provide appropriate clinical care for this during and after death. They also noted that in reality, most children donors are from those who die of trauma rather than those living with life-limiting conditions, because organ and tissue donation is more difficult to proceed outside of a paediatric intensive care unit. Therefore tissue donation could be considered and may be more appropriate under certain circumstances. However, the Committee noted that the families' choices around end of life care, including choices around place of care or death, should be absolutely respected and consideration around donation should not override a decision about treatment and place of care. In addition, the Committee noted that information about the possibility of organ or tissue donation should be provided to families whose children receive end of life care in different settings, including those who choose to receive end of care at home or in the community.

When, after consideration, organ or tissue donation is not possible, the Committee concluded that healthcare professionals should clearly explain the reasons to the child/young person (if appropriate) and their parents or carers. The parents or carers should also be alerted to any possibility of tissue donation or donation of samples for research.

When organ or tissue donation might be possible, the Committee concluded that healthcare professionals should discuss with the child/young person (if appropriate) and their parents or carers, whether the choice would have an impact on the care plan or place of care for the reasons mentioned above. Involving the organ donation team, the child/young person and their parents or carers should also be informed of the practical policies and procedures for organ and tissue donation. The Committee also noted that emotional support should be provided throughout the process, and this could be done by encouraging the child/young person or their parents or carers to discuss how they feel about the organ or tissue donation. As suggested by the evidence, families' wishes could change in the process and any change should be explored, noted and respected.

The Committee also discussed the circumstances where the child or young person lacks the capacity to make the decision. They agreed that parents or carers should be allowed to be the decision-maker regarding the child's organ or tissue donation, with the pre-condition that this was not against the best interest of the child or young person. However, they noted that if the child or young person's interest was compromised, then the donation should not happen.

The Committee noted the possible influence from the families' cultural and religious background in this context. They agreed that awareness of this should be raised in different care settings, and faith representatives from the community where the families came from could be involved in the communication as well. However, it was important not to see faith or religion as a barrier to donation.

#### 6.3.9.3 Quality of evidence

The quality of evidence ranged from moderate to low in this review. Evidence was downgraded mainly because of a lack of all relevant perspectives. Only experiences of mothers who have lost a child due to life-limiting conditions were interviewed and all participants were of one particular religious background. No studies were identified that explored the opinions of children or young people or of healthcare professionals. It was not clearly reported whether data saturation in the data analysis was achieved.

#### 6.3.9.4 Economic considerations

Organ and tissue donation is provided on the NHS and is considered cost-effective. However, the primary purpose of the guideline recommendations was not to increase the number of organ donors but rather to set the framework for discussing whether it is an option, and the practicalities, if so, in order that the preferences of children, young people and their families can be met. There are small costs associated in facilitating these discussions and information provision but the Committee considered these worthwhile in order to ensure that the wishes of the child, young person and their parents or carers were fulfilled as far as possible, and that where donation was not possible, the reasons were clearly explained.

#### 6.3.9.5 Other considerations

The Committee were aware of the guidance on effective communication contained within the NICE guideline on Patient experience in adult NHS services as well as NICE guidance on organ donation. The Committee considered these recommendations and how they apply to this guideline's population and some recommendations were based on Committee members' clinical experience, expert opinion and existing guidance.

Whether to draft a research recommendation was considered by the Committee, but even though the evidence was limited they concluded that there is sufficient guidance on organ donation already published which can be adapted to this guideline's population. The topic was therefore deprioritised.

#### 6.3.9.6 Key conclusions

The Committee concluded that discussions of organ and tissue donation should focus primarily on the wishes of the child or young person and their parents or carers. Their wishes should be respected and followed up to check whether they changed as circumstances changed. Information about organ and tissue donation should be provided at an early stage so families could consider it properly. Organ donation could not occur in all settings, this should be communicated to families and planned in advance. The advice and support of the organ and tissue donation team should be utilised in information provision and assessment of the appropriateness of the organ donation. The decision for organ or tissue donation should not override decisions about treatment, and should be separated from decisions about treatment withdrawal. All decisions should be made in the best interest of the child which is understood to include respect for their wishes and values, as well as their direct clinical interests.

### 6.3.10 Recommendations

**44. Discuss with the child or young person and their parents or carers whether or not they are eligible to donate organs or tissue.**

**45. Involve the organ donation service if needed. If organ or tissue donation is not possible, explain why.**

**46. If the child or young person is eligible to donate organs or tissue, discuss this with them and their parents or carers when they are ready and as part of Advance Care Planning, and:**

- provide written information leaflets if needed
- discuss how deciding to donate could affect their care, for example by changing their place of care and place of death
- explain the practical policies and procedures involved.

**47. If the child or young person does not have the capacity to decide about organ and tissue donation, ask their parents or carers to make the decision.**

**48. For further information on organ donation, including donor identification and consent, see the NICE guideline on organ donation for transplantation.**

# 7 Provision of care

## 7.1 Multidisciplinary teams

### 7.1.1 Review question

**In infants, children and young people living with life-limiting conditions, what is the clinical and cost effectiveness of receiving care from different models of MDT care including team composition and working arrangements?**

### 7.1.2 Introduction

Children and young people with life-limiting conditions have multiple needs which are addressed by a variety of healthcare professionals. There are often other multidisciplinary teams (MDTs) already involved with the child's care before the palliative care team become involved. As a result, as the child enters their end of life phase there is often a merging of different teams.

The question then arises as to who is required within a defined MDT to help each specific child through this aspect of their journey. There may be a need for additional clinical input, but many members of the original MDT may no longer be required to have an active role and may therefore need to adopt a supportive function and be called in only when necessary.

With the large variation in the composition of MDTs, the question arises as to what would represent an ideal MDT. This is of increasing importance as commissioners of services and service development need to be assured of the cost effectiveness of any defined MDT, balanced with the welfare of the child and support for his or her family. Finally, we must address the issue of whether the cost of developing a defined MDT is economically viable and whether the services could be run just through separate independent input.

### 7.1.3 Description of clinical evidence

The aim of this review was to determine the effectiveness of receiving care from a defined MDT of a particular composition compared with one of a different composition, or receiving care without a defined MDT, among years with a life-limiting condition. Evidence was sought which compared different MDT compositions.

No relevant studies were identified to meet the inclusion criteria for this review and therefore no evidence table was generated. Studies were excluded mainly because they were opinion pieces or narrative reviews without data analysis, with another study excluded because it was a non-comparative description / evaluation of a supportive program in paediatric palliative care. This study mainly reported on the services received by children and young people involved in the supportive programme which was not of interest to this review, therefore it was excluded. Published abstracts were also checked due to the scarcity of evidence, but none met the inclusion criteria.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

### 7.1.4 Summary of included studies

No evidence was found which met the inclusion criteria for this review.

### 7.1.5 Clinical evidence

No evidence was found which met the inclusion criteria for this review.

### 7.1.6 Economic evidence

This question was prioritised for health economic analysis, but no evidence was found. The Committee anticipated that the composition of the MDT would depend on the individual characteristics of the child or young person, and therefore a cost analysis of alternative MDT configurations was not undertaken.

### 7.1.7 Evidence statements

No evidence was found which met the inclusion criteria for this review.

### 7.1.8 Linking evidence to recommendations

#### 7.1.8.1 Relative value placed on the outcomes considered

Critical outcomes considered by the Committee were prevention of unplanned hospital admissions, quality of life of the child or young person, and quality of life of the parent or carer. Discharge time, satisfaction of the child or young person, satisfaction with care of the parent or carer (for example level of care and Improved communication), and control of symptoms (pain, dyspnoea, nausea/vomiting) were considered as important outcomes.

#### 7.1.8.2 Consideration of clinical benefits and harms

Despite the absence of clinical evidence, the Committee unanimously agreed on the importance of MDT working arrangements for the care of children and young people living with life-limiting conditions, throughout the course of their lives. Team working could ensure that the appropriate healthcare professionals and other agencies required, were involved in the care of the child or young person and that there would be effective and coordinated care strategies in place. The MDT composition and arrangements needed for each child and young people will differ and the needs of each individual would probably change during the course of the illness. The key consideration of the Committee was that needs will arise as early as when the life-limiting illness was diagnosed, and that MDT working arrangements should be in place throughout. The Committee considered that children with life-limiting conditions would (like others with chronic illnesses) be looked after by a multidisciplinary team with expertise in managing their specific condition. However, the team would also need to include expertise in those aspects of the condition specific to the life-limiting nature of their disease. For example, discussing early on in the course of their illness the nature of the condition and making an Advance Care Plan if appropriate. The team involved in their care would inevitably change in different care settings, if, for example they were admitted to hospital, or if they were to be cared for in an intensive care setting. In those who were approaching the end of life, or who were actually dying, the healthcare professionals and others in the MDT would inevitably change once more, in particular to include professionals with specific expertise in the management of end of life care. The Committee agreed that the essential consideration throughout the child or young person's life was that there should be continuity during such changes, and that there should always be a team of appropriate and clearly identified individuals who know the child, young person and their parents' or carers as members of the team. Clarity of communication with the parents or carers and between members of the team were also considered centrally important. The Guideline Committee considered that effective care via the MDT could assist in effective management, for example by avoiding unnecessary repeated hospital visits and potentially by avoiding the need for hospital admissions.

The Committee discussed the general principles for the composition of expertise in MDTs. They agreed that the MDT should involve relevant professionals from different disciplines depending on the needs of the individual child or young person. As the child or young person approached the end of life, involvement of those with expertise in palliative care might be increasingly important. If needed, the Committee considered that the MDT would need to be wider than health and may also include social care, education and the voluntary sector, as well as religious and spiritual support. Involvement of these individuals would depend on the needs of individual the child or young person and their parents or carers, especially towards the end of life.

The Committee also discussed the mechanism by which MDTs could work effectively with the child or young person and their parents or carers. They thought it was essential to inform the child or younger person and their parents or carers of which healthcare professionals are on the team and what their roles are in relation to the child or young person's care. This information as well as relevant contact details should also be provided in writing. If there are changes happening to the care or care settings, depending on the child or young person's prognosis of disease and stages of care, the child or young person and their parents or carers should be informed as to how the membership of the MDT could change during the disease trajectory accordingly, in order to accommodate the needs involved.

The Committee considered that it was essential to nominate a named co-ordinator to be the first point of contact throughout the end of life care from the point when an MDT was established. This could not only facilitate the young person and parents or carers' access to the MDT, but also improve the communication and coordination among healthcare professionals at different stages. However, the Committee noted the difficulties in naming a specific healthcare professional for different MDTs because of the various life-limiting-conditions, disease trajectories, and the changes needed accordingly during the course of end of life care. They noted the need to involve the GP throughout the process as they are likely to already be involved with the family, are be recipients of outcome letters from all specialists, and have a significant role in bereavement support for the whole family.

The Committee also discussed the importance of involving the child or young person and parents or carers in MDTs. They recommended that young people and their parents or carers should be offered the opportunity to participate in end of life care MDTs. They also thought that, where possible, children and young people parents and carers should be asked which professional group they would like to be involved in the end of life care MDTs. This would depend on the individual needs and circumstances of the child, young person, parents and carers, and it was recognised that the extent of involvement might vary and would be led by the wishes of the child or young person and their parents or carers.

#### 7.1.8.3 Economic considerations

The Committee considered that MDTs were important to ensure that all relevant healthcare professionals and agencies involved in the care of the child or young person, by working as a team, would promote effective and coordinated care. The Committee noted that an MDT was not necessarily more resource intensive in terms of staff time than more independent approaches, and that better coordination of services and care can lead to improved efficiency, of which better outcomes for the child or young person is an important part. The Committee also noted that MDTs are routinely used in the NHS for the management of chronic conditions.

#### 7.1.8.4 Quality of evidence

No clinical evidence was found for this review question.

#### 7.1.8.5 Other considerations

The Committee concluded that in the absence of relevant evidence, they would make recommendations that were based on Committee members' clinical experience, expert opinion and existing guidance.

#### 7.1.8.6 Key conclusions

The Committee emphasised that it was critical to have in place an MDT appropriate to the needs of the child or young person throughout the course of any life-limiting condition. They thought the establishment of an MDT could be as early as when the life-limiting condition is diagnosed in some cases, and that it should otherwise be formed when the need for it arises. The Committee recommended that professionals from different disciplines should be involved in the MDTs depending on the individualised needs of the young people and their families/carers. They also concluded that the composition of the MDTs, the roles of the healthcare professionals in the team, and possible changes on the composition during the trajectory of diseases should be communicated to the child/young person and their parents or carers. It was considered essential to have a named care coordinator to be the first point of contact with a deputy to cover absences. The Committee concluded that it was important to involve the young people with life-limiting conditions where feasible and according to their wishes, and was essential to involve parents or carers in the MDT and seek their opinions about which professional group should be involved in their care where possible, depending on their individualised needs.

### 7.1.9 Recommendations

**49. Children and young people with life-limiting conditions should be cared for by a defined multidisciplinary team.**

**50. As the child or young person's circumstances change (for example if they change from having care primarily to manage their condition to having end of life care), the membership of the multidisciplinary team should be adjusted accordingly.**

**51. Depending on the needs of the child or young person, the multidisciplinary team may include:**

- healthcare professionals from primary, secondary or tertiary services, including those with specialist expertise in palliative care
- social care practitioners
- education professionals
- spiritual or religious advisors
- hospice professionals.

**52. Explain to children and young people and their parents or carers:**

- who the multidisciplinary team members are and how they are involved in their care.
- how the multidisciplinary team membership will change if the care that is needed or the care setting changes.

**53. Think about involving children and young people and their parents or carers in multidisciplinary team meetings (when appropriate).**

**54. Think about having a named individual from the multidisciplinary team to act as a first point of contact and coordinate care for the child or young person and their parents or carers.**

## 7.2 End of life care around the clock

### 7.2.1 Review question

**What is the effectiveness of day and night specialist telephone health care professional support or parents/carers support, day and night community nursing support, and the combination of the 2 for the needs of infants, children and young people with life-limiting conditions, and for the needs of their family members and carers during this time and after death as part of service delivery?**

### 7.2.2 Introduction

Children and young people with life-limiting conditions often require complex management from a medical and nursing point of view. The rarity of many conditions means that primary care may not have much knowledge of the natural history or management of these illnesses. Primary-care physicians may only deal with the death of 1–2 child or young person in their career, and so are concerned with their abilities to manage such cases. Adult community nursing services have well-defined structures for managing older adult patients with terminal illness at home, but they are often concerned about their ability to deal with younger adults. Community children's nursing provision is variable throughout the country, and many teams struggle to provide round-the-clock care for terminally ill children.

In this section, we look at the evidence (or lack of evidence) for the effectiveness of round-the-clock specialist telephone healthcare professional support. The support can be either medical or nursing, and can be provided not only to the healthcare professionals in the community but also to the parents or carers of the child. We also consider the benefits of providing round-the-clock community nursing support to the child or young person and their parents or carers not only during the terminal phase of the illness but also after death.

Throughout this discussion we need to consider the medico-economic costs and benefits of providing care in the community compared with the hospital environment.

### 7.2.3 Description of clinical evidence

There were 3 objectives of this review. First was to assess the effectiveness of day-and-night specialist telephone support from healthcare professionals for parents/carers who are providing for the needs of children or young people living with life-limiting conditions. Second was to assess the effectiveness of day-and-night community nursing support services in providing for the needs of children or young people living with life-limiting conditions, and the needs of their families/carer. Third, the effectiveness of a combination of day-and-night telephone support plus community nursing support was assessed, in terms of how they provided for the needs of child or young person living with life-limiting conditions and their families/carers.

Systematic reviews of randomised control trials (RCTs) and cohort studies, RCTs, cohort studies and uncontrolled studies were searched for inclusion for this review. For interventions, we looked for studies that specifically examined the effectiveness of day and night specialist telephone healthcare profession support or/and day and night community nursing support. No directly relevant clinical study comparing day and night telephone advice or/and day and night community nursing versus no such services was identified.

One systematic review (Bradford 2013) on home-based telehealth was identified and its included studies on paediatric patients were cross-checked. Individual studies included in this systematic review that had been carried out among paediatric patients were checked, however, none of them specifically examined the effectiveness of day and night specialist telephone advice from healthcare professionals nor was day and night community nursing support, therefore none were included in this review.

We found no evidence on day and night community nursing support or the combination of day and night specialist telephone advice and day and night community nursing support.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

### 7.2.4 Summary of included studies

No evidence was found which met the inclusion criteria for this review.

### 7.2.5 Clinical evidence

No evidence was found which met the inclusion criteria for this review.

### 7.2.6 Economic evidence

A global health economic search was undertaken across the whole guideline. This identified a total of 4,156 papers. After reviewing titles and abstracts 38 full copies were obtained as potentially relevant to the questions under review in this guideline. A single health economic paper (Noyes, 2013) was identified from this search as relevant to the review question on day and night community nursing support and day and night specialist advice. This paper estimated an additional cost of £336,000 per year (or £14,000 per child) to provide 1 week of day and night end of life care at home to 24 children in North Wales. This paper is reviewed in more detail in the health economics chapter.

A costing model was produced for this guideline to compare the costs of a day and night community nursing support and day and night specialist telephone advice for children and young people receiving home care and approaching the end of life. This model is briefly summarised below but is described in full in the health economics chapter (see Appendix A).

The model was developed in Microsoft Excel® and sought to compare the costs of providing day and night community nursing support and day and night telephone specialist advice to children and young people being cared for at home and approaching the end of life with an alternative of inpatient hospital care. Model inputs could be varied as part of a ‘what-if’ analysis in order to assess the cost impact with different service configurations. The population covered by the service could also be varied to reflect that it has been suggested that such services would typically need to be provided for populations much bigger than those typically served by Clinical Commissioning Groups (CCGs). In addition the Committee thought that, while the service would typically be used for a relatively short period, there would be considerable variation in duration across different children and young people. Thus the mean duration of use of day and night service was highly uncertain and the ‘what-if’ approach allowed the implications of different service duration on costs to be explored.

The base case costing was based on commissioning for a population of 1.5 million, of which 337,500 were aged 0-18 years, as an independent report (Palliative Care Funding Review, 2011) suggested that this was likely to be an optimal population when commissioning palliative care services for children and young people. Staffing levels in the base case analysis reflected those used in a published cost exemplar (Noyes, 2013) although a small amount of consultant paediatric input was additionally included for the provision of specialist

telephone advice. The base case costing assumed a mean duration of 3 weeks for use of a day and night community nursing support and day and night specialist telephone advice service.

The model suggested that the cost of providing a day and night service for a population of 1.5 million would be £439,000 (£8,699 per child using the service). However, the model suggested that it would provide a net saving when compared with an alternative of inpatient care on a paediatric ward, costing £1,026,000 (or £20,265 per child).

In the published cost exemplar (Noyes, 2013) the population was considerably lower than 1.5 million and just 24 children used day and night service per annum. A sensitivity analysis was undertaken using this lower population and this suggested that day and night would remain cheaper providing the mean duration of service use was 17.5 days or more. With a duration of less than 17.5 days the costs of providing a day and night service were not completely offset by savings from reduced hospitalisation.

In order to populate the model and to assess the validity of the results, we were able to obtain data through a Committee member on the day and night community nursing service and day and night telephone specialist advice operated by the East Anglia Children's Palliative Care Managed Clinical Network (MCN). This service was available across 11 CCG covering a population of 3 million including just over 700,000 children. They reported that the managed clinical network had a Band 5 network co-ordinator working 22.5 hours per week (£16,000 per annum) but their duties would extend beyond activities relating to a day and night service. In the 2014/15 business case produced by the East Anglia Children's Palliative Care MCN to support their continuation they reported that the annual cost of providing "specialist medical advice at all times", a day and night specialist telephone advice service, would be £10,000 per annum. This was based on a trial period with an on-call rota of 4 specialists (3 paediatric consultants and 1 nurse consultant). They separately reported that their day and night specialist telephone advice service is now delivered by an on-call rota of 5 specialists (4 paediatric consultants and 1 consultant nurse) and that the total cost of the MCN specialist on-call telephone service for 2015/16 was £4,150. This figure is substantially less than the cost of day and night specialist telephone advice derived from our base case costing and for a much larger population. This would seem to suggest that the cost model may over-estimate the costs of providing such a service in practice.

They also report that the 2015/16 Symptom Management Nursing Team budget for 8 Band 7 nurses was £554,000 and that 6 Band 7 nurses participated in the out of hours on-call rota. They reported that the fixed cost of having a Band 7 nurse on standby out of hours was £16,425 per year but, in addition, the on-call team worked 295.75 hours overtime associated with work delivered while on call, which was paid at time plus 30%. This does not suggest that the cost model has under-estimated the staffing costs of providing day and night community nursing support based on the service configuration in East Anglia Children's Palliative Care MCN.

It should be noted that both the volume of out of hours calls for day and night specialist telephone advice and the number of out of hour visits made in response to those calls is relatively small even across this relatively large population. So for example, in 2015, 126 out of hour calls were made to the Symptom Management Nursing Team and that only 56 hours of out of hours visits were made in response to those calls. Most of the out of the hours calls were addressed with a telephone response from the community nurse specialist.

### 7.2.7 Evidence statements

One cost analysis reported that providing one week of day and night community nursing support and day and night specialist advice cost £14,000 per child. This was assessed as applicable with major limitations.

Original “what-if” analysis conducted for the guideline suggested that providing three weeks of day and night community nursing support and day and night specialist advice for a population of 1.5 million could save £11,072 per child compared to hospitalisation. This was assessed as applicable with major limitations.

## 7.2.8 Linking evidence to recommendations

### 7.2.8.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were quality of life of the child or young person and their families or carers, and satisfaction with the care of the child or young person and their families/carers. Change in health resources utilisation, change in level of distressing symptoms, and change requirement for home visits by nurses were rated as important outcomes.

### 7.2.8.2 Consideration of clinical benefits and harms

The Committee discussed the importance of enabling the child or young person to be cared for at home if this was their and their parents’ or carers’ preference. However, during the discussion of this topic it was also acknowledged that there may be clinical or other circumstances where it is not in the child or young person’s best interest to be looked after at home. In this respect the Committee also referred to the evidence from the interviews conducted for this guideline by Together for Short Lives in which children were realistic about having to go to hospital if this is necessary even if it goes against their preference. Sometimes the move from home to an institution often relates to unmet needs in the community, unsuccessful symptom management and most specifically to unsuccessful pain control. Balancing these options at the end of life is not straightforward since children with life-limiting conditions may have complex needs that, even with the best day and night care, may not be met in their home. It is important to acknowledge that these preferences can change with time and may need to be revisited. Identifying the place that can provide care that is in the best interest of the child or young person while respecting their and the parents’ wishes is therefore critical and will have to be discussed in advance and documented in their Advance Care Plan. The role of the children’s’ hospice should also be considered as they may be able to meet needs for nursing and medical care while avoiding the need to be cared for in a hospital environment. In particular, hospices often have more experience of symptom management (see 9, ‘Managing distressing symptoms’).

### 7.2.8.3 Economic considerations

No evidence was found in the review with respect to the effectiveness of day and night community nursing support and day and night specialist telephone advice for children and young people receiving home care and approaching the end of life. This is not surprising given the context in which such services would be provided. Nevertheless it is reasonable to assume that day and night services provide net benefits, as it facilitates home care where that is the preferred place of care and death.

The Committee were aware of a number reports supporting the provision of day and night services which helped inform guideline recommendations. In the End of Life Care strategy (Department of Health, 2008) it was stated that “Primary Care Trusts and Local Authorities will wish to consider how to ensure that medical, nursing and personal care and carers’ support services can be made available in the community 24/7”, and “that provision of day and night services can avoid unnecessary emergency admissions to hospital and can enable more people at the end of their life to live and die in the place of their choice”. An independent report (Palliative Care Funding Review, 2011) recommended that “Community services should be built up, to provide 24/7 access to community care across the country.

Availability of 24/7 care in the community is crucial to enable people to be cared for at home if they wish to do so."

There are approximately 39,000 children or young people in England with a life-limiting condition. Of these it is estimated that 18,000 per annum would be receiving some form of palliative care. The Committee agreed that the provision of day and night community nursing support and day and night specialist telephone specialist advice would predominantly be for a service for children and young people who are approaching end of life and having primary or secondary care. Using a published estimate (Lowson, 2007) which suggested that approximately 10% of children and young people in receipt of palliative care would die per annum, then in the region of 1,800 children and young people might be expected to make some use of a day and night community nursing support and day and night specialist telephone advice during the course of the year. This might be considered an 'upper bound' estimate of the number of service users as not all children and young people with a life-limiting condition will be receiving home care at the time of death. It is important to note that the duration a child or young person would use home nursing services is anticipated to be relatively short, typically a few days or weeks. Telephone services may be needed for several months but can cover a large geographical area so may be made more viable if the service is commissioned for a whole region These numbers represent a very small population from a commissioning perspective which will tend to limit the resource impact of recommendations on such a service.

Due to the relatively small number of children and young people with life-limiting conditions it is generally recognised that the optimal population size for commissioning paediatric palliative care services has to be much larger than the populations typically covered by clinical commissioning groups. A recently published review (Palliative Care Funding Review, 2011) suggested populations of 300,000 to 1.5 million for commissioning levels for palliative care services. The review also stated that for children the population is more likely to be closer to the maximum level.

In the absence of published evidence and limited information on actual service configuration a what-if costing model was developed to allow difference service configurations to be assessed. The base case costing, with a mean duration of day and night provision of 3 weeks, suggested that day and night services would be cheaper than an alternative where children and young people are cared for in hospital. This is because the costs of providing day and night services are more than offset by savings from reduced hospitalisations over this period. In the what-if analysis, day and night services remained cheaper providing that the cost of a hospital paediatric bed was more than £428 per day.

Clearly the level of staffing was an important determinant of cost-effectiveness, and if staffing levels were increased a point would be reached where day and night community nursing support and day and night specialist telephone advice services cease to be cheaper than hospitalisation. However, data from the East Anglia Children's Palliative Care Managed Clinical Network (MCN), covering a population of over 700,000 children, suggested that, if anything, day and night services could be provided with less staffing input per child using that service than was assumed in the base case analysis. Given what was stated earlier regarding the preferences of children and young people, a day and night service only has to demonstrate cost neutrality for cost-effectiveness to be established.

#### 7.2.8.4 Quality of evidence

No clinical evidence was identified.

#### 7.2.8.5 Other considerations

Due to the absence of evidence on clinical effectiveness in this review, recommendations were mainly based on the experience and expert opinions of the Committee. Because health

economic analysis showed that day and night specialist telephone advice and day and night nursing support would be cost saving, the Committee decided to recommend offering a flexible and responsive day and night specialist telephone and day and night nursing support to children and young people who are approaching their last days or weeks of life and are being cared for at home.

The Committee firstly considered the composition of such service delivery to enable it to happen. They agreed that a combination of telephone support and home visits would be needed and the services should be delivered by adequately trained staff, particularly with specific expertise in palliative care. They also discussed the importance of being able to provide parenteral drug therapy for those receiving their care at home. In particular, provision of support, training and equipment for subcutaneous infusions of, for example, opioids or anti-seizure medications, would be important for some patients. They concluded that the composition of day and night services for those cared for at home should include: specialist medical advice at any time; paediatric nursing support at any time; home visits for symptom management by a healthcare professional with expertise in palliative care; and practical support and equipment for interventions such as oxygen, enteral tube feeding, and subcutaneous therapies. In addition, the Committee also noted that proactive office hours planning regarding the care of the child or young person could have an impact on the care provided after hours. They agreed that when necessary anticipatory planning during office hours should take place so as to effectively manage symptoms that might arise when the child or young person was receiving care at home in their last days of life. However, they noted that extra support in the community was also needed in circumstances where additional treatment may be unexpectedly required for symptom management.

The Committee noted the importance of the local structure required to facilitate the proper day and night service delivery when the child or young person living with life-limiting conditions may be approaching the end of life and is receiving home care. They discussed that local structures are usually charities and hospices, however many of them are not funded by the NHS. They agreed that it was important to make sure that the child or young person’s Advance Care Plan is up-to-date and shared appropriately with the members of MDT teams, GPs, community nursing teams, and ambulance services.

The Committee agreed that clinical networks in collaboration with care planning and service delivery should be established so as to properly cover a population of an appropriate size and that these networks might aspire to formalised partnership working between the statuary and voluntary sector.

#### 7.2.8.6 Key conclusions

Although there was an absence of evidence, the Committee strongly recommended the provision of day and night service delivery for children and young people living with life-limiting conditions when they approach the end of life and receive care at home. They discussed the benefits that a day and night service could bring to those children and young people and their parents/carers, and what should be in place to make this possible: such as composition of the service delivery and skills needed, Advance Care Planning and anticipatory prescribing. Local structures should be in place to allow the services to be delivered, and collaboration needs to be established between clinical networks to cover the population of appropriate size in regions.

They also acknowledged that the child or young person may need or wish to move between settings in their last days of life. However, this choice needs to be balanced with careful consideration of circumstances where hospital admission may be necessary and preferable for all involved. Children and young people and their parents were found to be realistic about this. This information needs to be communicated sensitively and decisions need to be made in partnership and documented in the Advance Care Plan (see Shared decision-making and Advance Care Planning).

### 7.2.9 Recommendations

**55. For children and young people with life-limiting conditions who are approaching the end of life and are having home care, services should provide (when needed):**

- specialist medical advice at any time (day and night), for example telephone advice
- paediatric nursing care at any time (day and night)
- home visits by a healthcare professional with expertise in palliative care, for symptom management
- practical support and equipment for interventions including oxygen, enteral nutrition, and subcutaneous and intravenous therapies
- anticipatory prescribing for children and young people who are likely to develop symptoms.

**56. Services should have agreed strategies and processes to support children and young people who are approaching the end of life and are having home care. These services should be based on established clinical networks, and should collaborate on care planning and service delivery.**

# 7.3 Rapid transfer

## 7.3.1 Review question

**What services have to be in place to make rapid transfer available to take infants, children and young people with a life-limiting illness to their preferred place of care in their last days of life as part of service delivery?**

## 7.3.2 Introduction

When a child or young person enters the last days of life it is sometimes necessary to transfer them from 1 setting to another. This is normally from a hospital environment into either the child or young person's home or into a hospice. Many factors need to be considered to be able to carry out a rapid transfer seamlessly. Any transfer is subject to availability, even if there are long-standing advance plans already in place.

There are key issues to consider in terms of transport arrangements and particularly ambulance transfers, within the timescales necessary. In addition there needs to be a good care package in place and discussions regarding Advance Care Planning, parallel planning and care after death need to be considered. Equipment and medications have to be readily available in the location the child or young person is transferred to.

Management of the child or young person at home or hospice requires healthcare professionals from a variety of services as well as social and spiritual support. In complex cases the child or young person will require not only normal community support, but also support from hospital specialist and paediatric palliative care healthcare professionals. Throughout we must also consider the needs of the parents or carers around the child or young person.

We need to consider the special issues raised around any plan to undertake compassionate removal of a breathing tube (extubation) in the home or hospice. Success in this requires close collaboration between several different teams who would often need to work outside their normal environments. Parallel planning needs to be made in case the child or young person survives after the extubation.

Throughout this process we must also not forget all the other needs of the child or young person and their parents or carers.

## 7.3.3 Description of clinical evidence

The aim of this review was to determine the effectiveness of a rapid transfer programme (including from neonatal or paediatric intensive care), compared with standard transfer programme or care without such arrangement in facilitating children and young people with a life-limiting condition to die in their preferred place of care and/or death.

As an integrated part of the rapid transfer programme, particular consideration was given to children and young people who need compassionate extubation, also including planned withdrawal of all life-sustaining treatment (for example, non-invasive ventilation) in the preferred place, and looking at what services should be in place to facilitate this.

The aim was to include systematic reviews of randomised controlled trials (RCTs), RCTs, cohort studies and uncontrolled studies, but no evidence was found which met the inclusion criteria for this review.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

### 7.3.4 Summary of included studies

No evidence was found which met the inclusion criteria for this review.

### 7.3.5 Clinical evidence

No evidence was found which met the inclusion criteria for this review.

### 7.3.6 Economic evidence

This review question was prioritised for economic analysis.

A systematic search undertaken for this guideline did not identify any relevant economic literature relating to rapid transfer to take children and young people with a life-limiting illness to their preferred place of care in their last days of life.

A costing model was produced for this guideline to compare the costs of providing a rapid transfer service with an alternative where no such service was provided. This model is briefly summarised below but is described in full in Appendix K.

The model was developed in Microsoft Excel® and extensive use was made of 1-way and 2-way sensitivity analysis to reflect the fact that considerable uncertainty existed with respect to many model inputs.

Using the model's default input values, the analysis suggested that a rapid transfer service could be provided for an incremental cost of £700,000 per annum based on a population of 3,600 children and young people in their last days of life and where transfer would be in accordance of the wishes of the child and/or parents/carers, clinically appropriate and feasible. However, 1 of the key areas of uncertainty was with respect to the size of the population, and sensitivity analysis unsurprisingly suggested that the total costs of a rapid transfer service was sensitive to the size of the population.

Another, key input influencing net costs was the mean hospitalisation averted by rapid transfer. Holding other model inputs constant at their default value, a threshold analysis suggested that a rapid transfer service could become cost saving if the mean days of hospitalisation averted by rapid transfer was 0.2 days higher than indicated in the base case.

The modelling work undertaken for this guideline suggests that the population of children and young people who would use a rapid transfer service to their preferred place of care in their last days of life, is relatively small. There is some uncertainty as to whether the service would be cost saving or increasing at the individual level, but the relatively small population means that the resource impact of providing such a service is likely to be fairly limited.

### 7.3.7 Evidence statements

A cost analysis conducted for the guideline suggested that providing a rapid transfer service for England could cost £700,000 per annum relative to the alternative where a rapid transfer service was not provided. This was assessed as applicable with major limitations.

## 7.3.8 Linking evidence to recommendations

### 7.3.8.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were quality of life/ death of the child or young person, quality of life of the parents or carers, and satisfaction of the child or young person and their parents or carers; whereas transfer time, waiting time prior transfer, unexpected re-admission to hospital, and access of the family to their child were rated as important outcomes. No clinical evidence was identified.

### 7.3.8.2 Consideration of clinical benefits and harms

The Committee discussed the importance of taking into account the child or young person and their parents' or carers' wishes in relation to place of death. Accommodating their wishes should be a key aspect to consider in palliative care, as the Committee members agreed this diminishes suffering for both the children and young people and their families or carers.

The difficulties identified by the Committee in relation to rapid transfer were also raised. These were mainly related to costs and resource impact, and included: availability of ambulances, time taken to complete transfer, additional staff needed during transfer, and impact of ward functioning during staff absence. It was also mentioned by the Committee that although the number of people making use of this service is currently quite low, the number of people requesting this service is increasing steadily, as more parents are becoming aware of its availability and feasibility. The Committee therefore agreed that the comfort that the dying child or young person and their parents or carers would gain from being at the preferred place of death would outweigh the possible logistic difficulties of making this possible.

### 7.3.8.3 Economic considerations

In the absence of any studies reporting the effectiveness of a rapid transfer service to take children and young people to their preferred place of death, it was not possible to formally assess the cost-effectiveness of a service. Although the benefits of rapid transfer cannot be quantified at this moment in time it is reasonable to assume that a service would provide net benefits, as the Committee recognised that accommodating the wishes of children, young people and their families or carers in the context of palliative care helped to ameliorate suffering.

A service that provides net benefits and is cost neutral or cost saving can be described as cost-effective. The results from the model were equivocal as to whether a rapid transfer service would fall into that category. However, services which increase costs to the NHS can still be considered cost-effective if the additional benefit is commensurate with the increase in costs.

The model did not show that a rapid transfer service was not cost-effective and there were scenarios where the model suggested it could be cost saving, such as when rapid transfer would avert more than 1.2 days of inpatient care. Rapid transfer may also facilitate earlier withdrawal of burdensome care with concomitant improvements in welfare and reductions in costs. There is considerable uncertainty with respect to the number of children who would use such a service although that uncertainty is bounded within relatively small numbers. Therefore the resource impact of offering rapid transfer to children and young people is likely to be quite limited.

The Guideline Committee reported that such services were part of current practice in most of England and therefore did not think that the provision of rapid transfer would require an appreciable increase in NHS costs. They also noted that the provision of a rapid transfer service is also consistent with previously stated Department of Health objectives.

The Guideline Committee considered that, subject to limitations in the evidence, a rapid transfer service to the preferred place of care or death in the last days of life was likely to represent a good use of NHS resources.

#### 7.3.8.4 Quality of evidence

No studies were included in the review.

#### 7.3.8.5 Other considerations

Given the absence of evidence, the recommendations were based on Committee's expert opinion and a costing model produced for the guideline. The Committee agreed that the pathway proposed by Together for Short Lives could also be useful to draft the recommendations. The Committee agreed that it was important to establish a rapid transfer service to help children and young people to die in their preferred place. However, they could not be prescriptive about the details of the service because it would vary according to the collaborations (between already existing services, between hospitals and hospices for instance) that would be possible in a given area as well as vary according to the individual child or young person's condition and their particular circumstances.

The Committee agreed that the first step in the provision of this service would be to explain to the parents or carers whether rapid transfer is an option available to them, depending on the current setting of care and the preferred place of death and the child's individual circumstances. The Committee stressed the importance of not making assumptions, and the need to have discussions with the parents or carers in order to provide them with a meaningful choice where possible.

The second step would be to communicate with the MDT and to liaise with the relevant services (such as community nursing, their GP, hospice, ambulance service and so on) in order to ensure that everything is in place before commencing the transfer.

The third step would be to ensure that there is a current Advance Care Plan in place to manage the last hours or days of life. The Committee emphasised that having accommodated 'preferred place of care' in the child or young person's Advance Care Plan can be helpful, as it can facilitate having the necessary services in place.

In relation to the above, the Committee agreed that it is important to have an agreed treatment plan for the last hours or days of life. The main focus should be on symptom control, including pain management. In those children and young people requiring extubation, it is very important to agree the roles and responsibilities of the team, as well as the timescales. The Committee highlighted that parallel planning should be in place in case the child survives for longer than anticipated, as this is frequently seen in children and young people following withdrawal of life-sustaining treatment or other interventions. This uncertainty should be discussed with the parents or carers, and the child if appropriate.

The fourth step would be to plan for the events following the death of the child or young person, including aspects such as legal requirements and practical issues (for example confirmation and certification of death; as well as transport and care of the body after death). The Committee emphasised that it is particularly important to agree who will be responsible for the administrative aspects following the death of the child or young person.

The Committee agreed that there was much uncertainty about the decisions that procedures that need to be in place to make this service as effective as possible. They therefore thought that a research recommendation should be included with the aim of providing more clarity about this topic.

#### 7.3.8.6 Key conclusions

The Committee concluded that for some children and young people and their parents or carers, it is important to be transferred to their preferred place of care when they are entering the last days of life, and it should be explained to families whether this is an option that is available and suitable for them. For families for whom a transfer to the preferred place of care has been agreed, a plan should be made ahead of transfer to make sure that everything is in place. This plan should include details about the coordination with the relevant services, symptom management, timescales for the withdrawal of life-sustaining treatment (if relevant) and steps to follow after the death of the child.

### 7.3.9 Recommendations

**57. If it is suspected that a child or young person may die soon and they are not in their preferred place of death, think about whether rapid transfer is possible and in their best interest. Discuss this with them and their parents or carers.**

**58. When planning rapid transfer to the preferred place of death, review and if necessary update the Advance Care Plan in discussion with the child or young person and their parents or carers and with the healthcare professionals who will be involved following the transfer. The updated Advance Care Plan should include a record of:**

- any intended changes to care and when they should happen
- care plans that cover:
  - the final hours or days of life
  - what will happen if the child or young person lives longer than expected
  - support for the family after the child or young person dies
  - care of the child's or young person's body after death.
- the professionals who will be involved and their responsibilities
- the professionals who will help with the practical and administrative arrangements after the death.

**59. When planning rapid transfer of a child or young person to their intended place of death:**

- be aware that the course of their condition may be unpredictable, and that they may die sooner or later than expected
- discuss any uncertainties about the course of their condition and how this could affect their care with them and their parents or carers
- ensure that relevant changes to the Advance Care Plan are implemented.

**60. Think about using the rapid transfer service to allow the child or young person to be in their preferred place of death when withdrawing life-sustaining treatments, such as ventilation.**

**61. Before rapid transfer, agree with the parents or carers where the child's or young person's body will be cared for after their death.**

**62. In collaboration with local hospitals, hospices, and community, primary care and ambulance services, establish a rapid transfer service for children and young people with life limiting conditions to allow urgent transfer to the preferred place**

**of death (for example from the intensive care unit to their home, or other locations [such as a children's hospice]).**

### 7.3.10 Research recommendations

**2. Do protocols for rapid transfer of children and young people with life-limiting conditions help ensure that they are able to die in their preferred place of death?**

| Research question | Do protocols for rapid transfer of children and young people with life-limiting conditions help ensure that they are able to die in their preferred place of death? |
|---|---|
| Why this is needed | |
| Importance to 'patients' or the population | When a child or young person enters the last days of life it is sometimes necessary to transfer them from 1 setting to another. This is normally from a hospital environment either into the child or young person's home or into a hospice. Many factors need to be considered to be able to carry out a rapid transfer seamlessly (such as transport availability, equipment, availability of nursing staff). Research in this area could help by providing children or young people and their families/carers with the option to die in the place they prefer. A protocol will potentially decrease the length of hospital stay in specialist areas, for example NICU/PICU. It may also provide an opportunity for an audit to assess the needs for services according to the current clinical practice. |
| Relevance to NICE guidance | This should be considered a high priority as there is currently no clinical evidence to support the current recommendation for a rapid transfer policy/protocol. Research in this area would inform future guidance. |
| Relevance to the NHS | The availability of rapid transfer has a potential net saving to the NHS by decreasing the length of stay in areas of specialist input, for example PICU/NICU. At a time of emotional distress it will also increase parental satisfaction with services by being supported in their choice of place of death for their, baby, child or young person. |
| National priorities | Better care, better lives (Department of Health, 2008) suggests families should have a choice about place of care. |
| Current evidence base | In this guideline no clinical evidence was identified and it is therefore unclear whether or not having a rapid transfer protocol means that CYP and their families/carers are afforded their preferred place of death. |
| Equality | Many national reports have now highlighted that there is an inequity in choice of preferred place of death and having clear protocols will overcome this by having clear criteria for transfer. |
| Feasibility | This study can be carried out in several ways. It could be a comparative study between similar (well-matched) centres where one has a protocol and another does not. An alternative would be to assess current practice in areas without a protocol and then implement one and see if there is any change in outcome. |
| Other comments | |

# 7.4 Care based in the child or young person's home

## 7.4.1 Review question

**What is the clinical and cost-effectiveness of a home-based programme of care compared with care in other settings?**

## 7.4.2 Introduction

Care of a child or young person at home has always been seen as best practice, allowing the child to live within the community that they know. The benefits in terms of maintaining family life, saving on travel time to institutions and the disruption of living a life while residing within an institution such as a hospital, have been cited as reasons to try to develop home-based programs of care. There may also be financial benefits for commissioners in terms of cost of care. With the improvements of technology and the ability of the NHS to provide support outside of hospital to children with interventions such as gastrostomy, tracheostomy, and home ventilation, even children with very complex needs can now be managed at home.

However, the issue of how children can be clinically looked after on a home-based programme of care in terms of the skill sets of doctors, nurses and allied professionals needs to be assessed. Care of children within residential homes, children's hospices or hospitals have varying benefits and difficulties.

## 7.4.3 Description of clinical evidence

The objective of this review was to determine the clinical and cost effectiveness of a home-based programme of care compared with care in other settings for children and young people with a life-limiting condition who are approaching the last days of their life.

The aim was to include systematic reviews, randomised controlled trials, cohort studies and uncontrolled studies.

There were 3 observational studies which were included in this review (Arland 2013, Groh 2013, Postier 2014):

- 2 used an uncontrolled study design (Groh 2013, Postier 2014)
- 2 were retrospective cohort studies (Arland 2013, Friedrichsdorf 2015).

There were 3 studies conducted in the USA (Arland 2013, Friedrichsdorf 2015, Postier 2014) and 1 in Germany (Groh 2013).

With regard to the population, 2 studies included all paediatric patients receiving palliative care (Groh 2013, Postier 2014), 1 included oncology paediatric patients dying of a brain tumour (Arland 2013) and 1 study included bereaved parents of children who died of cancer (Friedrichsdorf 2015). All of the studies included indirect populations, as the life expectancy of the children was beyond 2 months.

With regard to the intervention and comparators included, all the studies looked at participants who received specialised palliative home/ hospice care compared with usual care provided by a non-specialised team.

Of the outcomes listed in the protocol and agreed by the guideline Committee:

- 2 studies reported on admissions to hospital (Arland 2013, Postier 2014);
- 2 studies reported on control of symptoms (Friedrichsdorf 2015, Groh 2013);
- 2 studies reported on children and young people's quality of life (Friedrichsdorf 2015, Groh 2013);

- 1 study reported on family or caregivers' quality of life (Groh 2013);
- 1 study reported on parents or caregivers' stress or distress (Groh 2013).
- no results were found for children and young people's satisfaction and control, nor for parents or caregivers' satisfaction and control.

A summary of the included studies is presented in Table 36.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix and in the GRADE profiles below and in Appendix J.

### 7.4.4 Summary of included studies

A summary of the studies that were included in this review is presented in Table 36.

**Table 36: Summary of included studies**

| Study | Intervention/ Comparison | Population | Outcomes | Comments |
|---|---|---|---|---|
| Arland 2013<br>USA<br>Retrospective study | • In-home end of life programme<br>○ Comprehensive end of life discussion<br>○ Medications for symptom control<br>○ Primary family liaison: a specific healthcare provider to be the contact person for the family and for the hospice or home-care agency.<br>○ Home visits: to assess the patient's symptoms by 1 or 2 healthcare providers from the team.<br>• Usual care<br>○ No details given | 166 paediatric patients aged 1 to 19 years dying of a brain tumour | • Unplanned/ precipitous admissions to hospital<br>○ Number of hospital admissions.<br>○ Length of hospital stay. | • Retrospective review of the patients' medical records.<br>• Data from the control group was extracted from medical records.<br>• Intervention and control groups were not comparable, as treatment of brain tumours may have improved during the 15-year period. |
| Friedrichsdorf 2015<br>USA<br>Retrospective study | • Home-based palliative care programme<br>○ Inpatient and clinic paediatric palliative care +<br>○ Home/ hospice visits by paediatric palliative care nurses, social workers, | 60 bereaved parents of children who died of cancer<br>Children median age at diagnosis: 7.7 | • Prevalence of symptoms (proxy for ICYP control of symptoms)<br>• ICYP quality of life | • Retrospective data<br>• Survey<br>• Parents of children with cancer only<br>• Enrolment in home-based palliative care programme based on eligibility criteria |

| Study | Intervention/ Comparison | Population | Outcomes | Comments |
|---|---|---|---|---|
| | chaplaincy<br>○ Some children received home visits from a paediatric palliative care physician and/ or paediatric oncologist or an oncology advanced practice registered nurse<br>• Usual care<br>○ Inpatient and clinic paediatric palliative care only | | | |
| Groh 2013<br>Germany<br>Uncontrolled study | • Palliative home care (PPHC)<br>○ Multi-professional PPCH team consisting of 3 paediatricians, 2 nurses, a social worker and a chaplain, all with special training in palliative care.<br>○ The main tasks of the team were the provision of palliative medical and nursing care, including day and night on-call service, as well as psychosocial support and coordination of professional assistance in cooperation with the local Health Care Professionals.<br>○ The participants had no additional support service added to their care during PPCH involvement that was not a direct result of the PPHC team's work. | 40 primary caregivers of severally ill-children aged 1 month to 18 years old | • Control of symptoms<br>• Children or young person's quality of life<br>• Caregivers' quality of life<br>• Parents or caregivers stress and distress<br>○ Burden relief for caregivers.<br>○ Caregivers stress and burden. | • Intervention group data was collected prospectively.<br>• Low internal validity due to study design, the children's health would be expected to deteriorate. |

| Study | Intervention/ Comparison | Population | Outcomes | Comments |
|---|---|---|---|---|
| | • Usual care<br>○ No details given | | | |
| Postier 2014<br>USA<br>Uncontrolled study | • Home-based paediatric care and hospice care (PPC)<br>○ No details given<br>• Usual care<br>○ No details given | 425 children aged 1 to 21 years old | • Unplanned/ precipitous admissions to hospital<br>○ Number of hospital admissions<br>○ Length of hospital stay | • Retrospective data obtained from hospital records.<br>• Low internal validity due to B-A study, as the children's health would be expected to deteriorate. |

1

## 7.4.51 Clinical evidence

2 **Table 37: Summary clinical evidence profile – Comparison 1: home-based palliative care versus usual care**

| **Home-based palliative care compared with usual care for end of life care** | | | | | | |
|---|---|---|---|---|---|---|
| Outcomes | Illustrative comparative risks* (95% CI) | | Relative effect (95% CI) | No of Participants (studies) | Quality of the evidence (GRADE) | Comments |
| | Assumed risk | Corresponding risk | | | | |
| | usual care | Home-based palliative care | | | | |
| Number of patients admitted to hospital<br>Assessed with: Hospital records<br>Follow-up: 5 years | 545 per 1000 | 295 per 1000<br>(180 to 480) | RR 0.54<br>(0.33 to 0.88) | 114<br>(Arland 2013)<br>Retrospective cohort study | ⊕⊖⊖⊖<br>very low[1,2,3] | |
| Total number of admissions<br>Assessed with: Hospital records<br>Follow-up: 5 years | 909 per 1000 | 409 per 1000<br>(309 to 545) | RR 0.45<br>(0.34 to 0.6) | 114<br>(Arland 2013)<br>Retrospective cohort study | ⊕⊖⊖⊖<br>very low[1,2] | |
| Average number of admissions<br>Assessed with: Hospital records<br>Follow-up: 24 months | The mean average number of admissions in the control group was 3.09±3.6 | The mean average number of admissions in the intervention groups was 0.09 higher<br>(0.44 lower to 0.62 higher) | | 425<br>(Postier 2014)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low[3,4,5] | |
| Average length of stay (days)<br>Assessed with: Hospital records<br>Follow-up: 5 years | The mean average length of stay (days) in the control groups was<br>4.04 days | The mean average length of stay (days) in the intervention groups was 1.01 higher | | 114<br>(Arland 2013)<br>Retrospective cohort study | ⊕⊖⊖⊖<br>very low[1,2] | Imprecision not calculable |
| Average length of stay (days)<br>Assessed with: Hospital records | The mean average length of stay (days) in the control groups was | The mean average length of stay (days) in the intervention groups was 10.06 lower | | 425<br>(Postier 2014)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low[4,5] | |

| | | | | | | |
|---|---|---|---|---|---|---|
| **Home-based palliative care compared with usual care for end of life care** | | | | | | |
| Follow-up: 24 months | 20.97 days | (14.65 to 5.47 lower) | | | | |
| Burden relief for caregivers<br>Measured with: own scale; range of scores 0 to 10 (better indicated by higher values)<br>Follow-up: 7.5 months | The median burden relief for caregivers in the control group was<br>9.0 (3) | The median burden relief for caregivers in the intervention groups was<br>2.0 (3) | P<0.001 | 40<br>(Groh 2013)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low[6,7] | Imprecision not calculable |
| Caregiver stress and burden<br>Measured with: HADS; range of scores 0 to 21 (better indicated by lower values)<br>Follow-up: 7.5 months | The median caregivers stress and burden in the control group was<br>7.0 (3) | The median caregivers stress and burden in the intervention groups was<br>10.0 (2) | P<0.001 | 40<br>(Groh 2013)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low[6,7] | Imprecision not calculable<br>HADS: Hospital Anxiety and Depression Scale |
| Control of symptoms<br>Measured with: range of scores 0 to 10 (better indicated by higher values)<br>Follow-up: 7.5 months | The median control of symptoms in the control group was<br>5.0 (3) | The median control of symptoms in the intervention groups was<br>9.0 (2) | P<0.001 | 40 (Groh 2013)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low[6,7] | Imprecision not calculable |
| Child or young person's health-related quality of life<br>Measured with: own scale; range of scores 0 to 10 (better indicated by higher values)<br>Follow-up: 7.5 months | The median children's health-related quality of life in the control group was<br>4.0 (4) | The median children's health-related quality of life in the intervention groups was<br>2.5 (2) | P<0.001 | 40<br>(Groh 2013)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low[6,7] | Imprecision not calculable |
| Child or young person's health-related quality of life: having fun<br>Measured with: own scale; nominal scale great deal/ a | 300 per 1000 | 699 per 1000<br>(387 to 1000) | RR 2.33 (1.29 to 4.23) | 60<br>(Friedrichsdorf 2015)<br>Retrospective cohort study | ⊕⊖⊖⊖<br>very low[3,8] | |

| Home-based palliative care compared with usual care for end of life care | | | | | | |
|---|---|---|---|---|---|---|
| lot/ some vs little/ none<br>Follow-up: not reported | | | | | | |
| Child or young person's health-related quality of life: feeling peaceful<br>Measured with: own scale; nominal scale great deal/ a lot/ some vs little/ none<br>Follow-up: not reported | 467 per 100 | 499 per 1000<br>(294 to 845) | RR 1.07 (0.63 to 1.81) | 60<br>(Friedrichsdorf 2015)<br>Retrospective cohort study | ⊕⊖⊖⊖<br>very low[8,9] | |
| Child or young person's health-related quality of life: feeling afraid<br>Measured with: own scale; nominal scale great deal/ a lot/ some vs little/ none<br>Follow-up: not reported | 267 per 1000 | 435 per 1000<br>(211 to 891) | RR 1.63 (0.79 to 3.34) | 60<br>(Friedrichsdorf 2015)<br>Retrospective cohort study | ⊕⊖⊖⊖<br>very low[8,9] | |
| Child or young person's health-related quality of life: enjoying meaningful events<br>Measured with: own scale; nominal scale great deal/ a lot/ some vs little/ none<br>Follow-up: not reported | 633 per 1000 | 798 per 1000<br>(576 to 1000) | RR 1.26 (0.91 to 1.75) | 60<br>(Friedrichsdorf 2015)<br>Retrospective cohort study | ⊕⊖⊖⊖<br>very low[3,8] | |
| Parents or caregivers quality of life<br>Measured with: QOLLTI-F; range of scores 0 to 10 (better indicated by higher values)<br>Follow-up: 7.5 months | The median parents or caregivers QoL in the control group was<br>5.8 (1) | The median parents or caregivers QoL in the intervention group was<br>7.2 (1.3) | p<0.001 | 40<br>(Grogh 2013)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low[6,7] | Imprecision not calculable<br>QOLLTI-F: Quality of Life in Life-Threatening Illness--Family Carer Version |
| *The basis for the assumed risk (for example, the median control group risk across studies) is provided in footnotes. The corresponding risk (and its 95% confidence interval) is based on the assumed risk in the comparison group and the relative effect of the intervention (and its 95% CI). | | | | | | |

**Home-based palliative care compared with usual care for end of life care**

CI: Confidence interval; RR: Risk ratio;

*<Insert Note here>*

*1 This is an observational study and the quality of the evidence was further downgraded by 2 due to high risk of selection and performance bias and unclear risk of attrition and detection bias*

*2 The quality of the evidence was downgraded by 1 because the sample is limited to children with brain tumours. Also sample includes home and hospice care.*

*3 The quality of the evidence was downgraded by 1 because the CI crosses 1 default MID*

*4 This is an observational study and the the quality of the evidence was further downgraded by 2 due to high-risk of selection bias and performance bias*

*5 The quality of the evidence was downgraded by 1 because the ICYP participants in this sample have a life expectancy >2 months (24 at least) (indirect population)*

*6 This is an observational study and the the quality of the evidence was further downgraded by 2 due to high risk of selection, performance and detection bias*

*7 The quality of the evidence was downgraded by 1 because the life expectancy in this sample is beyond 2 months (indirect population)*

*8 This is an observational study and the the quality of the evidence was further downgraded by 2 due to high risk of performance and detection bias*

*9 The quality of the evidence was downgraded by 2 because the CI crosses 2 default MIDs*

### 7.4.6 Economic evidence

This question was prioritised for health economic analysis but no evidence was found. No additional modelling was undertaken partly because of the limitations of the clinical evidence but more importantly because of the substantial overlaps between home-based programmes of care and the provision of day and night community nursing and telephone support which the Committee considered a vital component of any home-based programme of care.

An alternative to home-based care is hospital care and Table 38 lists illustrative NHS Reference costs associated with neonatal and paediatric hospitalisation.

**Table 38: Hospital critical and palliative care costs per bed day**

| Description | Unit Cost [a] | Currency Code |
|---|---|---|
| Neonatal Critical Care, Intensive Care | £1,176 | XA01Z |
| Neonatal Critical Care, High Dependency | £847 | XA02Z |
| Neonatal Critical Care, Special Care, without External Carer | £533 | XA03Z |
| Neonatal Critical Care, Special Care, with External Carer | £424 | XA04Z |
| Neonatal Critical Care, Normal Care | £464 | XA05Z |
| Paediatric Critical Care, Advanced Critical Care 5 | £4,824 | XB01Z |
| Paediatric Critical Care, Advanced Critical Care 4 | £1,783 | XB02Z |
| Paediatric Critical Care, Advanced Critical Care 3 | £1,967 | XB03Z |
| Paediatric Critical Care, Advanced Critical Care 2 | £1,924 | XB04Z |
| Paediatric Critical Care, Advanced Critical Care 1 | £1,662 | XB05Z |
| Paediatric Critical Care, Intermediate Critical Care | £1,297 | XB06Z |
| Paediatric Critical Care, Intermediate Critical Care | £988 | XB07Z |
| Paediatric Critical Care, Intermediate Critical Care | £849 | XB09Z |
| • Inpatient Specialist Palliative Care | £388 [b] | SD01A |

*(a) NHS Reference Costs 2014-15*
*(b) This is based on patients aged 19 years and over because the equivalent cost for patients aged 18 years and under is based on a single data submission*

A "what-if" analysis reported in section 7.2.8.3 suggested that day and night services to facilitate home care could be cheaper than the hospital alternative providing the cost of hospital care was greater than £428 per day. This finding depended on the particular configuration of the day and night service as well as features of the population covered by the service.

### 7.4.7 Evidence statements

**Unplanned/ precipitous admissions to hospital**

Very low quality evidence from 1 retrospective cohort study with 114 paediatric patients dying of a brain tumour, showed that when specialised home-based palliative care was in place, a clinically significant lower number of children had to be admitted to hospital at 5-years follow-up. There was uncertainty around this estimate effect.

Very low quality evidence from 1 retrospective cohort study with 114 paediatric patients dying of a brain tumour, showed that when specialised home-based palliative care was in place, the total number of admissions was clinically significant lower at 5 years follow-up.

Very low quality evidence from an uncontrolled study with 425 paediatric patients showed a clinically significant higher average number of admissions after the home-based paediatric care programme was introduced at 24 months follow-up. There was uncertainty around this estimate effect.

Very low quality evidence from 1 retrospective cohort study with 114 paediatric patients dying of a brain tumour showed an increase in the average number of days that children stayed in hospital after the home-based palliative care programme was implemented at 5 years follow-up. The clinical significance of this outcome could not be calculated with the data reported.

Very low quality evidence from 1 uncontrolled study with 425 paediatric patients showed a clinically significant reduction in the average number of days that children stayed in hospital after the home-based palliative care programme was implemented at 24 months follow-up.

**Parents or caregiver's stress and distress**

Very low quality evidence from 1 uncontrolled study with 40 primary caregivers of severely-ill children showed that palliative home care reduced caregivers' burden (as measured with the Hospital Anxiety and Depression scale, HADS) when compared with previous usual care at 7.5 months follow-up. The clinical significance of this outcome could not be calculated with the data reported.

Very low quality evidence from 1 uncontrolled study with 40 primary caregivers of severely-ill children showed there was an improvement in parents' or caregiver's reported stress and distress (as measured with the Hospital Anxiety and Depression scale, HADS ) when palliative home care was in place at 7.5 months follow-up. The clinical significance of this outcome could not be calculated with the data reported.

**Children and young people's satisfaction and control**

No evidence was found for this outcome.

**Parents or caregiver's satisfaction and control**

No evidence was found for this outcome.

**Control of symptoms**

Very low quality evidence from 1 uncontrolled study with 40 primary caregivers of severely-ill children showed that there was an overall improvement in symptom management (as measured with study own scale) when palliative home care was in place, compared with previous usual care at 7.5 months follow-up. The clinical significance of this outcome could not be calculated with the data reported.

**Children and young people's health-related quality of life**

Very low quality evidence from 1 uncontrolled study with 40 primary caregivers of severely-ill children and young people showed that the child or young person's quality of life (as measured with study own scale) improved when palliative home care was in place, compared with previous usual care at 7.5 months follow-up. The clinical significance of this outcome could not be calculated with the data reported

Very low quality evidence from 1 retrospective cohort study with 60 bereaved parents of children who died because of cancer showed that parent-reported quality of life of their children was clinically significant higher for the domain "having fun" (as measured with study own scale, follow-up not reported).

Very low quality evidence from 1 retrospective cohort study with 60 bereaved parents of children who died because of cancer showed that parent-reported quality of life of their children was clinically significant higher for the domains “feeling peaceful” and “feeling afraid” (as measured with study own scale, follow-up not reported). There was considerable uncertainty around this estimate effect.

Very low quality evidence from 1 retrospective cohort study with 60 bereaved parents of children who died because of cancer showed that parent-reported quality of life of their children was clinically significant higher for the domain “enjoying meaningful events” (as measured with study own scale, follow-up not reported). There was considerable uncertainty around this estimate effect.

**Parents or caregiver’s health-related quality of life**

Very low quality evidence from 1 uncontrolled study with 40 primary caregivers of severely-ill children showed that the caregivers’ quality of life (as measured with the Quality of Life in Life-Threatening Illness--Family Carer Version, QOLLTI-F) improved when palliative home care was in place, compared with previous usual care at 7.5 months follow-up. The clinical significance of this outcome could not be calculated with the data reported

## 7.4.8 Linking evidence to recommendations

### 7.4.8.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were control of symptoms and parents or carer’s stress and distress; whereas admissions to hospital, child or young person’s satisfaction and comfort, parents or carers’ satisfaction, child or young person and their parents or carers’ quality of life were rated as important outcomes.

### 7.4.8.2 Consideration of clinical benefits and harms

The Committee discussed the potential advantages and disadvantages of the different models of care.

They pointed out that although it is widely assumed that community-based care is better, in reality it is not always the case. The results from the indirect evidence presented in this review showed that access to home-based care can reduce the number of admissions to hospital and the length of stay. However, evidence from other studies did not corroborate these findings. The Committee, based on their knowledge and experience highlighted that in adults only a small percentage of people had adequate pain control at home. They highlighted that this was very likely a consequence of inadequate resources to deliver effective symptom control at home and rather than concluding that home-based care was inevitably associated with poor pain management, the necessary expertise, resources and support should be provided. The Committee concluded that there were clear advantages for adequately supported home-based care rather than hospital care where this was clinically appropriate and the child or young person and parent or carer preferred it (as highlighted in the Preferred Place of Care and Preferred Place of Death section in this guideline).

The consensus of the Committee was that home-based care can be preferable to hospital care as long as an adequate package of care is in place. Based on this, the Committee concluded that a recommendation should be made to recommend comprehensive packages of home care to support palliative care at home, where appropriate. They agreed that a home-based programme of care should always include day and night access to specialist palliative medical advice and specialised palliative nursing support, although this may sometimes need to take the form of telephone specialist advice supporting bed side care from local teams, in addition to universal services. Some children may also require access to specialised ancillary support and appropriate equipment and maintenance.

The Committee recognised based on their own experience that many parents would like to take their child home, as long as an adequate package of care was in place. However they also recognised that some prefer care to be given in hospital. They all agreed that, where appropriate, parents would like to be offered the choice, following informed discussion. They also mentioned that it is important to take into account the cultural, religious and spiritual perspective of the child or young person and their parents or carers.

Although no evidence was found in relation to satisfaction, the Committee discussed parent's views and experiences and concluded from their combined experience that some may find it excessively burdensome and stressful to have their child at home when they are dying. However, others feel better if their child is at home. This is particularly the case when children have a complex package of care already in place. In relation to the child's satisfaction, it would be difficult to show an improvement in their condition or overall care.

#### 7.4.8.3 Economic considerations

In the absence of direct evidence, the Committee hypothesised that home-based community care was likely to be cost-effective, whereas hospital care would be the most expensive option. Some support for this view comes from a report, on adults, that the estimated cost for a day of community care at the end of life is £145, substantially less than the £425 for a specialist palliative inpatient bed day in hospital (https://www.mariecurie.org.uk/globalassets/media/documents/commissioning-our-services/publications/understanding-cost-end-life-care-different-settingspdf ). However, they also noted that a home-based community care would not always be preferable to care delivered in a hospital or hospice setting.

Home-based community care usually involves the substitution of care in a hospital setting with care in a home setting. Therefore, while there are costs associated with delivering a home-based programme of care, there would be some off-setting reduction in hospital costs from reduced hospital admission.

Key components of the costs of providing a home-based community service are addressed in Section 7.2.6 and Section 7.2.8.3 which suggested that round-the-clock community nursing and telephone support could be cost saving relative to hospitalisation subject to the precise configuration of the service.

Key components of the costs of providing a home-based community service are addressed in 7.2.6 and **Error! Reference source not found.** which suggested that round-the-clock community nursing and telephone support could be cost saving relative to hospitalisation subject to the precise configuration of the service.

#### 7.4.8.4 Quality of evidence

The quality of the evidence was initially graded as low as all studies were observational, and it was further downgraded due to the methodological flaws inherent to uncontrolled study design, population indirectness and reporting bias (recall and desirability bias). So the overall evidence was of very low quality as assessed by GRADE.

The Committee also noted that the evidence did not cover the aim of the review, as it did not actually compare home versus hospital or hospice care, but compared different programmes of home care. They also mentioned that there were issues regarding the generalisation of the results. Firstly, 2 of the studies were carried out in the USA, and their healthcare system is not comparable with that in the UK. In this sense, the study carried out in Germany was agreed to be more relevant to the UK setting. Secondly, the programmes of care described in the studies were much more comprehensive than the current programmes of home care used in the UK.

#### 7.4.8.5 Other considerations

The Committee emphasised that healthcare professionals should discuss, in advance, with the parents or caregivers the support needs that might be necessary, such as house adaptations, equipment or social support.

They also noted that the healthcare professionals should know what services can be provided to parents, and that a recommendation was needed about this. In this sense, the Committee raised issues regarding equality between the care provided in hospital and at home, and also inequities between different settings. For example, in some regions there is no out of hours support or specialist care for symptom management or end of life care.

Finally, the Committee agreed that, in the absence of evidence, a research recommendation was needed to assess the effectiveness of home-based care and the impact on satisfaction, quality of life and symptom control.

#### 7.4.8.6 Key conclusions

The guideline Committee concluded that in addition to universal services, home-based programmes of care should include: access to specialist medical advice, nursing and ancillary support at any time, and access to appropriate equipment.

### 7.4.9 Recommendations

**63. When discussing possible places of care or places of death with children and young people and their parents or carers, provide information about:**

- the various care settings (for example home, hospice or hospital care)
- the care and support available in each setting
- practical and safety issues.

**64. If the child or young person and their parents or carers prefer home care, take into account and discuss the practical considerations with them, such as the possible need for:**

- home adaptations
- changes to living arrangements
- equipment and support.

**65. Services for children and young people who are approaching the end of life and are being cared for at home should be able to support parenteral drug administration (for example, continuous subcutaneous opioid or anticonvulsant infusions).**

### 7.4.10 Research recommendations

**3. What is the effectiveness of a home-based package of care as opposed to hospital or hospice care?**

| Research question | What is the effectiveness of a home-based package of care as opposed to hospital or hospice care? |
|---|---|
| Why this is needed | |
| Importance to 'patients' or the population | Children with life-limiting conditions and their families and carers at times prefer to have end of life care in the home. Due to lack of availability they may at times have to attend hospitals acutely and require emergency admission to hospices when they could be managed at home with the appropriate support. This could potentially avoid a hospital admission. |

| Research question | What is the effectiveness of a home-based package of care as opposed to hospital or hospice care? |
|---|---|
| Relevance to NICE guidance | It is a high priority for research in this area to guide future recommendations, because there was no evidence available at the time of the original guideline being written. |
| Relevance to the NHS | Economic modelling undertaken during the guideline development process suggested that there would be net cost savings to the NHS when there was an effective home-based package of care. This would also decrease pressure on acute hospital beds in specialist areas. |
| National priorities | In the End of Life Care strategy (Department of Health, 2008) it was stated that "PCTs and LAs will wish to consider how to ensure that medical, nursing and personal care and carers' support services can be made available in the community 24/7" and "that provision of day and night services can avoid unnecessary emergency admissions to hospital and can enable more people at the end of their life to live and die in the place of their choice". An independent report (Palliative Care Funding Review, 2011) recommended that "Community services should be built up, to provide day and night access to community care across the country. Availability of day and night care in the community is crucial to enable people to be cared for at home if they wish to do so."<br>Two of the aims of the document Better care, Better lives (Department of Health, 2008) for children with life-limiting conditions were to:<br>• "ensure that all children have a choice on location of care, 24-hour access to multidisciplinary community teams and, when needed, specialist palliative care advice and services."<br>• Have "Access to specialist end-of-life care and 24-hour support and advice should be available." |
| Current evidence base | No evidence was identified with respect to the effectiveness of any home-based package of care, day and night community nursing support and day and night specialist telephone advice for CYP receiving home care approaching the end of life. |
| Equality | Children with life-limiting conditions should have the opportunity to participate in research that could improve their quality of life. |
| Feasibility | This would need to be a multicentre national study as the numbers of children with life-limiting conditions are relatively small. There would be no additional expense if areas that already had day and night community and specialist services were included. If looking at new services being set up then there would be an initial cost in expanding the community and specialist services but this has been shown by economic modelling to have a net saving to the NHS in the long term. Outcomes to be measured could include satisfaction, quality of life and symptom control. |
| Other comments | |

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# 8 Support

## 8.1 Emotional and psychological support and interventions

### 8.1.1 Review question 1

**Are psychological interventions effective for infants, children and young people with life-limiting conditions and what factors influence the attitudes of children and young people and the family's involvement and decisions about choices of those interventions?**

### 8.1.2 Review question 2

**Are psychological interventions (including short-term bereavement therapies) effective for family members and carers of infants, children and young people and what factors influence their attitudes about those interventions before and after the death of an infant, child or young person with a life-limiting condition?**

### 8.1.3 Introduction

The immense diversity and variety of psychological needs of children and young people with life-limiting conditions cannot and should not be underestimated. Differences in age, ability, disability, symptoms, condition type, illness progression, phase of condition, stage of social and intellectual development, culture, family relationships, support network and financial resources all impact on psychological experience and possibilities for engaging with different types of psychological intervention. Additionally, the needs of the individual child vary with changes in health needs, social and emotional development and key transition points on the care journey.

Some life-limiting conditions affect children's cognitive and social communication abilities directly and, there is a high prevalence of learning disabilities and progressive neurological changes that can impact on an individual's capacity to communicate about medical symptoms and understand their condition and medical interventions.

For children with life-limiting conditions, the timeliness of interventions are particularly important as their changing health may create time-limited windows of opportunity to engage in therapy and to live life well.

There is a broad range of specialist psychological interventions that may be indicated for individuals who are able to engage in talking therapies. These may include preparation for medical procedures, promoting adherence to care plans, pain management, managing trauma, developing adaptive strategies for coping with difficult feelings and thoughts, adjusting to diagnosis, adjusting to loss of skills and abilities, and seeking change in relationships in anticipation of death. Specialist psychological interventions can also address the needs of children at a pre-verbal level of development who demonstrate high levels of distress or behavioural difficulties.

As in the case of psychological interventions for other family members, this question specifically addresses the provision of psychological and psychotherapeutic interventions and therapies delivered or directly supervised by qualified psychological practitioners or psychotherapists with professional accreditation and registration.

In our society, children dying through illness is outside of most peoples' expectations and experience. Families of children with life-limiting conditions face extraordinary psychological circumstances during the journey from diagnosis, through living with illness and medical

interventions, deterioration, dying, death and bereavement. Families have busy and unpredictable lives with frequent health appointments, hospital admissions and changes in the child's health that can prevent them from accessing clinic-based mental health services. Childhood life-limiting conditions affect the whole family at an individual and systematic level and this can challenge health systems to be mindful of the needs of all members of the family when the focus is on the needs of the dying child.

In these circumstances, and given the general high rates of prevalence of mental health difficulties across the life span in child and adult populations, it is highly likely that some family members and carers will be vulnerable to experiencing significant mental health difficulties which require specialist intervention as recommended.

While emotional support and compassion provided by all members of the child's MDT are essential, this question specifically addresses the provision of psychological and psychotherapeutic interventions and therapies delivered or directly supervised by qualified psychological practitioners or psychotherapists with professional accreditation and registration. Practitioners providing interventions for families affected by childhood life-limiting conditions need to be skilled in both the evidence-based therapeutic approach and in adapting therapies to themes of medical decision-making, loss, death, dying, bereavement and early intervention to develop resilience and supportive family relationships.

Psychological interventions provided by practitioners not positioned in pathways to be able to provide continuity across the journey from diagnosis to after-death care, introduces a risk that professional support may be withdrawn at the time of death, compounding losses at a time of bereavement.

### 8.1.4 Description of clinical evidence

Two mixed-methods reviews were carried out for this chapter. One focused on children and young people living with life-limiting conditions, and the other on their family and carers, respectively. The mixed-methods approach was taken because it allows for the inclusion of different study designs (both quantitative and qualitative) in order to investigate both the effectiveness of interventions as well as to explore peoples' perspectives on related to this topic.

#### 8.1.4.1 Description of evidence on children and young people living with life-limiting conditions

**For the quantitative review, the objectives were:**

- To assess the effectiveness of psychological interventions/therapies for improving psychological well-being (such as resilience, depression, fear, or anxiety) in children and young people living with life-limiting conditions and approaching the end of life.
- To assess the effectiveness of psychological interventions/therapies for reducing physical symptoms (such as pain) associated with a life-limiting condition in children and young people who are approaching the end of life.
- To look for systematic reviews, randomised control trials, and observational comparative studies.

**For the qualitative review, the objectives were:**

- To identify and describe the factors that influenced children, young people, and their parents/carers' attitudes in making choices about psychological therapies, who are living with a life-limiting condition and approaching the end of life.
- To identify and describe children, young people, and their parents/carers' experiences with psychological therapies.

- To look for studies that collected data using qualitative methods (such as semi-structured interviews, focus groups, and surveys with open-ended questions) and analysed data qualitatively (including thematic analysis, framework thematic analysis, content analysis and so on). Survey studies restricted to reporting descriptive data that were analysed quantitatively were excluded.

#### 8.1.4.2 Description of evidence on the family and carers of children and young people living with life-limiting conditions:

**For the quantitative review, the objective was:**

- To assess the effectiveness of psychological interventions/therapies for improving psychological well-being such as resilience, depression, fear, or anxiety in carers/families (including siblings) of children and young people with life-limiting conditions before and after the child's death.
- To look for systematic reviews, randomised control trials, cohort studies and uncontrolled studies.

**For the qualitative review, the objectives were:**

- To identify and describe factors that influenced carers/families' (including siblings) attitudes towards psychological therapies, whose child (or sibling) living with a life-limiting condition and/or approaching the end of life, before and after the child's death.
- To identify and describe carers/families' (including siblings) experiences with psychological therapies, challenges faced, and unmet needs (such as access, resources, burdens due to the lack of adequate psychological therapy either provided to them or to their child (sibling) with life-limiting condition).
- To look for studies that collected data using qualitative methods (such as semi-structured interviews, focus groups, and surveys with open-ended questions) and analysed data qualitatively (including thematic analysis, framework thematic analysis, content analysis and so on). Survey studies restricted to reporting descriptive data that were analysed quantitatively were excluded.
- One study (Jennings 2014) was identified.

## 8.1.5 Summary of included studies

### 8.1.5.1 Children and young people

**Quantitative review: summary of included studies**

No evidence was found which met the inclusion criteria for this review

**Qualitative review: summary of included studies**

No evidence was found which met the inclusion criteria for this review

### 8.1.5.2 Parents and carers

**Quantitative review: summary of included studies**

No evidence was found which met the inclusion criteria for this review for children and young people living with life-limiting conditions nor their family and carers.

**Qualitative review: summary of included studies**

For the family and carers of children and young people living with life-limiting conditions, only 1 qualitative study (Jennings 2014) conducted in Ireland among mothers (N=10) whose child died from a life-limiting condition was included. Participants in this study had received formal and informal bereavement support following the death of their child. The study collected data using unstructured interviews and content analysis was employed to analyse qualitative data.

This single study reported on mothers' attitudes toward bereavement support received. Except for this, no evidence on other themes considered important by the Committee was identified.

A brief description of this study is provided in Table 39.

Due to the qualitative nature of these studies, evidence is summarised in adapted GRADE-CERQual tables within the evidence report. Therefore no separate Appendix is provided for this.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A summary of the studies identified is reported in a selection flow chart in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included study studies is summarised in the evidence tables in Appendix G and in the adapted GRADE profiles below and in Appendix J. The TFSL focus group report can be found in Appendix L.

A summary of the study that was included in this qualitative review is presented in Table 39.

**Table 39: Summary of included studies**

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| Jennings 2014 | Interviews | N= 10 mothers in Ireland whose child died of a LLC | To examine 10 mothers' experiences of bereavement following the death of their child from a life-limiting condition in Ireland. | • Very small sample study.<br>• Formal sources of bereavement support included hospital organised bereavement group meetings, bereavement days, and voluntary organisations.<br>• Unclear whether data saturation was achieved in terms of both data collection and analysis.<br>• Researchers' role in the analytical process not critically reviewed. |

## 8.1.6 Clinical evidence

### 8.1.6.1 Quantitative review

No evidence was found which met the inclusion criteria for this part of the review.

### 8.1.6.2 Qualitative review

- Clinical evidence profile

Summary of Clinical Evidence in Table 40

**Table 40: Summary of evidence (adapted GRADE-CERQual): Theme 1 – Companionship and being understood**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Parents' (mothers') experience with bereavement support: companionship and being understood** | | | | | |
| 1 study (Jennings, 2014) | 1 study used unstructured interviews; | One study conducted in Ireland among mothers who lost a child to LLCs reported that they perceived accessing formal sources of bereavement support helpful. The mothers felt supported by attending group meetings, through meeting other parents who had also experienced the death of their child:<br><br>**Companionship and being understood:**<br>*" It was good hearing other people's stories and they had the same kind of feelings...I don't know, it's kind of a general companionship or something being with other people that you don't feel like you are the only one"* | Limitation of evidence | Major limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

### 8.1.7 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis.

### 8.1.8 Evidence statements

#### 8.1.8.1 Quantitative review: evidence statements

No quantitative evidence was identified.

#### 8.1.8.2 Qualitative review: evidence statements

**Parents' (mothers') experience with bereavement support: companionship and being understood**

Low quality evidence from 1 study carried out among mothers who had a child that died of life-limiting conditions, showed that mothers felt supported by accessing formal sources of bereavement support through attending group meetings, meeting other parents who had also experienced the death of their child.

### 8.1.9 Linking evidence to recommendations

The Committee wrote recommendations that applied to children and young people as well as their families.

#### 8.1.9.1 Relative value placed on the outcomes and themes considered

##### *8.1.9.1.1 Outcomes and themes considered in the review question for children and young people living with life-limiting condition*

**Outcomes of the quantitative review:**

For the quantitative review, critical outcomes considered were psychological well-being, quality of life of the child or young person and changing clinical symptoms, whereas satisfaction of children and young people and their parents or carers and adherence to care plan or management of condition would be important outcomes. No evidence was identified for the quantitative review

**Themes for the qualitative review:**

In the context of children and young people's attitudes and views about psychological therapies, the Committee anticipated some themes but also considered other themes were they to emerge from the evidence. These anticipated themes included children and young people's perceptions of treatments effectiveness; their attitudes toward psychological therapies; their experiences with psychological therapies; experiences with the therapist; unmet needs; challenges experienced; and others. No evidence was found for the qualitative review.

#### *8.1.9.1.2 Outcomes and themes considered in the review question for parents and families of children and young people living with life-limiting conditions*

**Outcomes considered in the quantitative review:**

The Committee considered that psychological well-being of carers/families before and after the child's death; quality of life of carers/families before and after the child's death, and family function before and after the child's death would be critical for decision-making, whereas satisfaction of carers/ families coping of carers/ families and activity of daily living and parenting would be important outcomes. No evidence was identified.

**Themes for the qualitative review:**

In the context of parental and familial attitudes and views about psychological therapies before or after the child or young person's death, the Committee anticipated some themes but would also consider other themes were they to emerge from the evidence. These anticipated themes included bereavement for carers/families (including siblings) before and after child's death; families' perceptions of treatments effectiveness; families' attitudes toward psychological therapies; unmet needs; parents' attitudes about disclosure to siblings; and challenges experienced. Only limited evidence with only one theme was identified. In this theme it described that mothers liked the companionship that bereavement support groups provide and found these meetings therefore helpful. The Committee considered this theme important in their discussions when drafting the recommendations.

### 8.1.9.2 Consideration of barriers and facilitators (for children and young people, and parents and families)

For children and young people no evidence was identified. The Committee acknowledged the emotional burden and distress caused in relation to end of life care for children and young people with life-limiting conditions. They agreed that offering psychological support to the children and young people with life-limiting conditions and their family members would improve their end of life care and the quality of life. Although there was a lack of data from well-conducted quantitative or qualitative research, the Committee agreed that psychological support and interventions would benefit the child or young person and their families/carers and guidance was therefore needed.

#### *8.1.9.2.1 Barriers and facilitators highlighted in the TFSL report*

Due to the absence of published literature, the Committee's discussion focused around the experiences and opinions that were expressed by children and young people in the focus groups conducted for this guideline.

"Better emotional care" was a key theme in the TFSL children and young people consultation report. The young people stressed that living well and dealing in an emotionally healthy way with their illness or condition, alongside their other developmental tasks. One of themes that emerged was "talking it through", that is, having conversations with someone who really understands was identified as important to many participants. This varied across the interviewed children and included friends, family, teachers, online forums, other young people with a similar condition, carers and professional support from a psychologist, which for some young people provided an opportunity to share feelings they would not discuss with others. Not everyone in the focus group reported having someone to talk to who understands, and while some young people believed this would help them, other young people were reluctant to seek professional support for emotions.

Children and young people reported that they were often coping as best they could on their own, largely using distraction and avoidance techniques to manage difficult feelings and stop themselves from overthinking. The Committee discussed that these techniques have a place

but they are not optimal coping strategies as they are avoidant rather than positive strategies.

"Being seen as an individual person" and "living life well" were important over-arching themes in the TFSL consultation of relevance to planning psychological support and interventions to children and young people and their families. Children and young people spoke about the central importance of being seen as an individual person first, rather than the condition or illness being their defining characteristic. The young people emphasised the importance to them of living well with the condition, rather than the focus being on deteriorating health and preparing to die. The Committee discussed that all healthcare professionals should be aware of the psychological importance of a focus on being enabled to live well with a life-limiting condition and also consider referral to specialist psychological services that can provide therapies focused on living life well, having good relationships and memories, alongside managing the emotional challenges of living with a life-limiting condition.

Based on these themes the Committee discussed that integrating specialist psychological input into the care and care plan at an early stage may help children and young people and families to access such help and ensure that they could benefit from this resource.

#### 8.1.9.3 Economic considerations

The Committee appreciated that there was a lack of clinical evidence for specific psychological interventions in children and young people with life-limiting conditions and their parents and carers. However, they also noted the difficulties of undertaking quantitative research in this area and stressed that the lack of evidence should not be interpreted as a lack of effectiveness. The recommendations reflected this lack of evidence while recognising that psychological and emotional support is important for the well-being of children and young people with life-limiting conditions and their parents and carers. The Committee were strongly of the view, based on their clinical experience and evidence in other contexts, that psychological and emotional support was likely to be cost effective.

While the recommendations are not prescriptive regarding the provision of specific interventions, the Committee were aware that there was inequity in the provision of psychological services across regions and, therefore, by highlighting good practice the expectation was that some uplift in NHS resource use would be required to redress the inequity in access to psychological and emotional support. The Committee were aware that a shortage of appropriately trained staff contributed to the inequity in provision of services.

#### 8.1.9.4 Quality of evidence (for children and young people, and parents and families)

Although the evidence review was conducted for 2 separate questions, the Committee agreed to write 1 set of recommendations for both children and young people as well as their parents/carers and family members because no published evidence was found for the review of psychological interventions for children and young people.

**Quantitative reviews: quality of the evidence**

No evidence was found either for children and young people or their family and carers.

**Qualitative reviews: quality of the evidence**

No qualitative evidence was identified for children and young people's experiences with psychological interventions.

For parents and families, low quality evidence was identified from 1 study carried out among mothers who had lost their child to a life-limiting condition in a UK NHS setting. The quality was graded low because there was no discussion on whether saturation was reached in the

thematic analysis. Similarly, the analytical process was not described in detail, as there was no description of how 'themes' were arrived at. The researchers also did not critically review their own roles in the process, nor the relationship between them and the respondents.

#### 8.1.9.5 Other considerations

The Committee was aware of the scarcity of evidence about effectiveness of psychological interventions or qualitative research with children and young people who have a life-limiting condition as well as their families. Conducting research in this area is difficult due to the variety of psychological conditions and disorders, the small number of subjects available for trials, and the range and complexity of psychological interventions employed. It was also noted that there are currently small numbers of psychological services providing specialist interventions specifically for children and young people with life-limiting conditions and their families, and that this would further limit opportunity to evaluate the effectiveness and families' experience of these interventions for this population.

The Committee discussed the following issues in current practice. They acknowledged an inequity of psychological services provision across regions for children and young people and their families and talked about the emotional needs at different times of change.

The Committee considered the enormity of the psychological impact on children and young people with a life-limiting condition and their families particularly about the awareness of shortened life, approaching death and bereavement and the importance of all healthcare professionals involved in their care being mindful and sensitive to this. The Committee also discussed the wide range of other stressful and distressing circumstances experienced by children and young people with a life-limiting condition and their families, and the impact this can have on lives and relationships for the whole family.

The Committee thought that healthcare professionals should also be aware that some children and young people with life-limiting conditions and their families may need support from healthcare professionals or specialist interventions to prevent the risk of developing psychological difficulties or disorders and to enhance their quality of life. They also noted that particularly during times of change in end of life care (for example, changes in care setting or staff or deterioration of the condition) timely/immediate interventions were needed which could help children and young people deal with their distress, learn to cope and build resilience in the process. They acknowledged that families' needs for psychological support can vary widely and that not everybody would need complex interventions.

The Committee agreed and stressed that all healthcare professionals (including primary care) had a responsibility to provide the families with some emotional support and information with regard to psychological support or interventions that they may have access to and are available. They emphasised the importance of the availability and accessibility of psychological support/interventions.

Families and carers of children and young people with life-limiting conditions often encounter gaps in services with regard to trying to access psychological services. The Committee identified a need for parents/carers assessed as having psychological support needs in their own right to have access to therapeutic support to enable them to cope and build resilience to go on in providing support and care to their children. They recommended that a link to appropriate psychological services should be established in the MDT for the care of the children and young people with life-limiting conditions and their families. These psychological services should not only be able to provide advice, but also to accept referrals when needs arise and to offer flexibility to enable access.

The Committee noted that while most healthcare professionals should be aware of the psychological impact on families, they may need to develop confidence to talk about these issues with children and young people and their families, and to offer families the opportunities for open discussions and review of their psychological needs at regular

intervals. The Committee discussed the importance of training, support, supervision and access to psychological consultation for healthcare professionals around talking with children and young people and families about their emotional and psychological needs but acknowledged that it is beyond the scope of the guidance to make recommendations on these themes, and the evidence for the effectiveness and experience of training and supervision of healthcare professionals was not looked at in the evidence review.

The Committee considered that review of psychological support needs would be important at key moments, such as at diagnosis, during deterioration, at times of change in personal circumstances, at transition to nursery/school/college/employment, and at times of significant change in goals for management and care. Moreover, the healthcare professionals need to be aware that the need for access to psychological services is not the same across different conditions because of varying trajectories and prognoses in the child's life-limiting condition. The access to services may therefore need to be individualised for each child and family.

The Committee noted that an important issue in practice was raising awareness and understanding of emotional and psychological supports that are available and which may be helpful for children and young people and their parents or carers. Healthcare professionals should inform families how to access those services, if they are available.

The Committee also discussed equality of access for children and young people with learning disabilities, communication difficulties or other developmental conditions. There are particular specialist psychological interventions that can be tailored to support the needs of children and young people with learning and social communication difficulties.

The Committee discussed bereavement support for parents/carers and family members when the child or young person is approaching the end of life. They noted that provision of information about bereavement support was identified as helpful by parents and carers in the previous review on information provision. They also thought that the findings of this review were in line with their observations in practice. As suggested by the evidence, they agreed that bereavement support groups could enable parents or carers to share their feelings with other families who have had similar experience.

They thought that information on bereavement support should be offered for parents/carers and families after the child's death. Furthermore, although no evidence was found on the effectiveness of psychological interventions among bereaved parents or carers, the Committee agreed it was appropriate for healthcare professionals to inform them of the availability of psychological support group meetings.

The Committee acknowledged that some children and young people with a life-limiting condition and/or their family members may experience significant mental health problems and may need support from mental health services. The Committee noted that in practice sometimes the access to such services could take a long time and this is of concern when life expectancy is short or uncertain.

The Committee discussed the skills and competencies healthcare professionals need in order to carry out the psychological assessments and interventions with children and young people and their parents/carers and family members. They also noted the importance of family involvement in decision-making and consent with regard to emotional support, psychological assessment or more complex interventions. An awareness of cultural and religious differences in the acceptance of psychological interventions was also viewed as important by the Committee.

The Committee discussed whether a research recommendation should be made because of the lack of evidence. There was agreement that future research should explore the emotional support needs of children and young peoples as well as parents or carers with the aim to find better ways to address these needs.

#### *8.1.9.5.1 Other considerations related to TFSL in emotional and psychological support*

The Committee also discussed relevant findings from the TFSL's report. They noted that findings from this report were in line with their experiences. They also noted that "Better emotional care" was a key theme in the report. And another relevant theme identified was young people coping by "Talking it through" (with, for example, friends, family, carers, teachers, in online forums, other young people with a similar condition and psychologists). However not everyone reported having someone available to talk to. Furthermore, some children and young people reported that they were often coping as best they could on their own, largely using distraction and avoidance. This, from another perspective, highlighted the need for optimal strategies provided by the professionals when the needs arise, and as discussed earlier, particularly at times of change.

### 8.1.9.6 Key conclusions

Mainly based on discussions about the findings from the children and young peoples' focus groups conducted for this guideline, the Committee concluded that, psychological support and interventions have to be individualised and that the needs for psychological support are likely to change during end of life care. However, not all children and young people with life-limiting conditions and their families would need or want psychological intervention. The need for information, support and regular discussions about psychological wellbeing as well as bereavement support as well as the needs of people with identified mental health problems and children with developmental problems or learning difficulties were highlighted as the most important issues. Special consideration should be given to those with special needs such as children and young people with learning disabilities, social communication difficulties and other developmental conditions.

## 8.1.10 Recommendations

**66. Be aware that children and young people with life-limiting conditions and their parents or carers may have:**

- emotional and psychological distress and crises
- relationship difficulties
- mental health problems.

**67. Be aware that children and young people and their parents or carers may need support, and sometimes expert psychological intervention, to help with distress, coping, and building resilience.**

**68. Be aware that children and young people may experience rapid changes in their condition and so might need emergency interventions and urgent access to psychological services.**

**69. Be aware of the specific emotional and psychological difficulties that may affect children and young people who have learning difficulties or problems with communication.**

**70. Provide information to children and young people and their parents or carers about the emotional and psychological support available and how to access it.**

**71. Regularly discuss emotional and psychological wellbeing with children and young people and their parents or carers, particularly at times of change such as:**

- when the life-limiting condition is diagnosed

- if their clinical condition deteriorates
- if their personal circumstances change
- if there are changes to their nursery care, school or college arrangements, or their employment
- if there are changes to their clinical care, for example if their care changes focus from treating the condition to end of life care.

### 8.1.11 Research recommendations

**4. What emotional support do children and young people with life-limiting conditions and their parents or carers need, and how would they like these needs to be addressed?**

| Research question | What emotional support do children and young people with life-limiting conditions and their parents or carers need, and how would they like these needs to be addressed? |
|---|---|
| Why this is needed | |
| Importance to 'patients' or the population | The findings of such research could contribute to guidance on how to improve identification of psychological needs of families of ICYP with life-limiting conditions and also contribute to planning to enable access to services providing interventions adapted to the specific needs and goals for psychological interventions for this population.<br>This is important because The Big Study identified significant unmet need in this area (approximately one-third of parents feel that the psychological needs of the child with a life-limiting condition, their siblings and their own needs as parents were not met by current UK service provision). |
| Relevance to NICE guidance | High: Only 1 research paper met criteria for evidence for this very broad topic in the current guidance, so it has only been possible to make very general recommendations based on expert opinion for this area which is of concern given that this area was considered of high importance by stakeholders in the scoping process. |
| Relevance to the NHS | It is unclear what the impact would be as the findings are unknown and there is no current evidence base specific to this population. However, it could be cost saving to the NHS because the costs of the interventions and trained staff could be offset by better symptom management (for example, levels of anxiety, agitation and maybe pain) as well as better health related quality of life and satisfaction with care. |
| National priorities | • NHS England (2016) The 5-year forward view for mental health. A report from the independent Mental Health Taskforce to the NHS in England.<br>• Department of Health (2015) Improving mental health services for young people: Report of the work of the Children and Young People's Mental Health Taskforce.<br>• Department of Health (2011) No Health Without Mental Health: The mental health strategy for England |
| Current evidence base | No evidence was found in the process of developing guidelines for psychological interventions for ICYP with life-limiting conditions. Previous UK studies have explored broad themes of psychological support needs and identified unmet need for psychological services for this population. However, these studies have not used standardised measures of psychological and relationship distress to understand the severity and nature of the distress and have not been robust enough in their design and reporting to be considered in the NICE review process. |
| Equality | The research will explore to what extent this population do have or are currently excluded from accessing psychological services and what are the barriers and facilitators for different members of the family when an ICYP has a life-limiting condition. |

| Research question | What emotional support do children and young people with life-limiting conditions and their parents or carers need, and how would they like these needs to be addressed? |
|---|---|
| | |
| Feasibility | **Population:**<br>**ICYP with life-limiting conditions and their parents/carers and siblings.**<br><br>To be representative, participants recruited from:<br><br>(1) Multiple sites across the UK to ensure sufficient numbers of participants are recruited and are representative of UK diversity as culture and social circumstance may have significant impact on psychological needs, preferences for services and access needs.<br>(2) Areas both with and without specialist children's palliative care psychology services (necessary because large sections of the country do not have these specialist services and the experience and needs of people in both situations may be very different due to differences in what they can and can't access)<br>(3) Both NHS and 3rd Sector Hospice services (necessary because an exclusively hospice sample would exclude families who do not feel comfortable with accessing / being referred to a hospice and also selectively include families with relatively higher levels of social support which they access via the hospice)<br><br>Participants should also be representative of the range of ICYP ages, ability/disability and type of life-limiting conditions.<br><br>A qualitative approach is proposed:<br>The aim is to explore the experience and attitudes of participants in addressing the following questions:<br><br>What is the impact of living with life-limiting conditions on the psychological wellbeing, family functioning and quality of life experienced by ICYP with life-limiting conditions and their parents/carers and family?<br>What range of types and severity of psychological and relational distress are experienced by ICYP with life-limiting conditions and their parents/carers and family members across their journey from recognition of shortened life expectancy to end of life care and bereavement? What are the predictors of forms and levels of psychological and relational distress and coping in this diverse population?<br>What types of psychological services and psychological interventions are ICYP with life-limiting conditions and their parents/carers and family members currently accessing? What are the barriers and facilitators to access? How useful do families find the interventions that they access? What ideas do families have about how the interventions they access could be improved? What outcome goals do families have for accessing psychological services/interventions? To what extent are these goals achieved?<br><br>In order to assemble sufficiently large samples, participants would need to be recruited across a number of centres.<br>Not all families will be willing or able to participate immediately after disclosure of diagnosis / prognosis, but a majority may be prepared to be invited and may engage/opt to participate within the first 6 months. |
| Other comments | Together for Short Lives carried out a focus group study to inform this guideline, which partially covered CYP's views on their emotional needs. However, it did not cover the topic in detail or include family members. It shows that this is a feasible study to carry out given the small population. |

# 8.2 Social and practical support

## 8.2.1 Review question

**What factors of social and practical support (including care of the body) are effective in end of life care of infants, children and young people with life-limiting conditions and their family members or carers and what influences attitudes about these before and after death?**

## 8.2.2 Introduction

The impact of life-limiting conditions on children, young people and their families are significant. They are at risk of practical and social isolation and exclusion, for example in relation to housing, transport and access to education. Parents are at risk of feeling emotionally overburdened and physically exhausted. The practical demands of care can create barriers in accessing employment or education which can in turn compound social isolation and effect financial resources.

Social and practical support that takes into account the cultural and religious background of the family has the potential to reduce the impact on their health needs and disabilities on daily life and development. It may also ameliorate the effects of these challenges and demands on parents/carers and siblings/other children in the family who may themselves be at risk of becoming young carers.

It is important to note that the support needs for children with life-limiting conditions and family members are not static but fluid and will change throughout their lifetime.

The impact on families does not stop when a child dies. Parents may not have experience or knowledge of the legal framework and multiple practical tasks that are bestowed on the next-of-kin when anyone dies. These have to be managed alongside coping with the impact of a major loss in the family and social network. Bereaved parents may experience social isolation and stigma or feel silenced when they attempt to talk about their loss and face the challenge of supporting others in the family while managing their own grief.

Bereaved siblings also have ongoing needs for parenting and social support that need to be considered in terms of understanding their loss and, to the social reactions of peers who may need guidance to be supportive towards a bereaved child.

## 8.2.3 Description of clinical evidence

The mixed-methods approach was taken because it allowed for the inclusion of different study designs (both quantitative and qualitative) in order to investigate both the effectiveness of interventions as well as to explore people's perspectives on related to this topic.

The aim of the quantitative review was to:

- Assess the effectiveness of social and practical support interventions for children and young people who are approaching the end of life and their family members or carers.

For the qualitative review, the aim was to:

- identify and describe factors that influence children and young people and their families or carers attitudes towards social and practical support interventions
- identify and describe children and young people and their family or carers' experiences with social and practical support interventions, challenges faced and unmet needs.

No evidence for the quantitative part of the question was found. The description here focused on the qualitative evidence included in this review.

For the qualitative part of the review question, studies were looked for that collected data using qualitative methods (such as semi-structured interviews, focus groups, and surveys with open-ended questions) and analysed data qualitatively (including thematic analysis, framework thematic analysis, content analysis and so on). Survey studies restricted to reporting descriptive data that were analysed quantitatively were excluded.

Given the nature of qualitative reviews, findings/themes were summarised from the literature and were not restricted to those identified as likely themes by the Committee.

A total of 22 studies (Brosig 2007; Cadell 2012; Champagne 2012; Contro 2002; Contro 2012; Davies 1996; Davies 2004; deCinque 2006; Einaudi 2010; Eaton 2008; Forrester 2008; Grinyer 2010; Jennings 2014; Konrad 2007; Malcolm 2008; Maynard 2005; Monterosso 2007; Price 2013; Robert 2012; Remedios 2015; Steele 2008; Weidner 2011) were identified for inclusion in this review. The majority of them focused on the perspectives of parents whose child had received or was receiving hospice or palliative care, or had passed away. Only 3 studies (Contro 2002; Price 2013; Remedios 2015) focused on the healthcare professionals' perspectives and 1 study focused on the perspectives of family members including siblings and grandparents (Grinyer 2010).

The majority of included studies collected data by semi-structured interviews or focus groups. The most common data analysis method employed across studies was thematic analysis. Four studies (Davies 1996; Einaudi 2010; Forrester 2008; Remedios 2015) collected data by open-ended questionnaires.

With regard to the setting of studies:

- 6 were conducted in the UK (Eaton 2008; Forrester 2008; Grinyer 2010; Malcolm 2008; Maynard 2005; Price 2013);
- 6 in the USA (Brosig 2007; Contro 2002; Contro 2012; Konrad 2007; Robert 2012; Weidner 2011);
- 4 in Canada (Champagne 2012; Davies 1996; Davies 2004; Steele 2008);
- 3 in Australia (deCinque 2006; Monterosso 2007; Remedios 2015);
- 2 in both Canada and in the USA (Cadell 2012); and
- 2 each in France (Einaudi 2010), and Ireland (Jennings 2014).

Evidence on all themes considered important by the Committee was identified. A number of further themes or sub-themes emerged from studies were also identified and incorporated in the review.

A brief description of the studies is provided in Table 41.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix G and in the GRADE profiles below and in Appendix J.

For presentation of findings, a theme map was generated according to the themes that emerged from studies (Figure 9 shows a theme map relating to social and practical support.

Figure 9). The mapping part of the review was drafted by 1 researcher from the guideline technical team but the final framework of themes was further shaped and when necessary re-classified through discussions with at least 1 other researcher. Due to the qualitative nature of these studies evidence is summarised in adapted GRADE-CERQual tables within the evidence report. Therefore no separate Appendix is provided for this.

### 8.2.4 Summary of included studies

A summary of the studies that were included in this review are presented in Table 41.

**Table 41: Summary of included studies**

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| **Interviews/focus-groups** | | | | |
| Brosig (2007) USA | Interviews | N=19 parents of deceased infants | To identify factors important to parents in their infant's end of life care. | • unclear whether data saturation in terms of collection or analysis was achieved<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Cadell (2012) Canada and the USA | Interviews | N=35 individual and couple interviews (47 people) | To explore the factors that allow parents who are caring for a child with a life-limiting illness to survive and to grow in the face of adversity | • The relationship between the researcher and the respondents was not reported |
| Champagne (2012) Canada | Interviews | N= 25 families (25 mothers and 8 fathers) | To analyse, from the parents' point of view, the effects of respite services offered at a children's hospice. | • the researchers' roles and potential influences in the analytical process was not critically reviewed |
| Contro (2002) USA | Interviews | N=68 family members of 44 deceased children | To analyse information from families about their experiences and their suggestions for improving the quality of end of life care, for developing a Paediatric Palliative Care Program | • Convenience sampling strategy used<br>• The relationship between the researcher and the respondents not clearly reported<br>• no discussion on whether saturation was reached for any of the themes reported |
| Contro (2012) USA | Interviews | N = 60 staff members from multiple disciplines | To examine the current state of bereavement care at a university-based children's hospital from the perspective of the interdisciplinary staff. | • sample selection procedure was not clearly reported<br>• the relationship between the researcher and the respondents was not clearly reported<br>• no discussion on whether saturation was reached for any of the themes reported |
| Davies (2004) | 1) face-to-face | N=18 families | To evaluate the | • the relationship |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| Canada | interviews and 2) mail-out surveys (questionnaire) | (50 family members): face-to-face interviews N=70 families: mail-out surveys | respite component of a broader project that examined the effect of the Canuck Place children's hospice program on the families it served. | between the researcher and the respondents was not clearly reported<br>• no discussion on whether saturation was been reached for any of the themes reported |
| deCinque (2006) Australia | Interviews | N=9: parents who had received hospital-based bereavement support following the death of their child from cancer | To explore the experiences and needs of 9 parents who had received hospital-based bereavement support following the death of their child from cancer, in Western Australia | • unclear whether data saturation in terms of collection or analysis was achieved<br>• researchers' role in and influences in the analytical process was not critically reviewed |
| Eaton (2008) UK | Interviews | N=11 families either receiving (n=5) or not (n=6) respite care at the hospice | To describe the experiences of families, whose children had life-limiting and life-threatening conditions and who had complex healthcare needs, of receiving respite care at home or in a hospice. | • convenience sampling strategy used<br>• The relationship between the researcher and the respondents was not clearly reported<br>• no discussion on whether saturation was been reached for any of the themes reported<br>• The researchers' roles and potential influences in the analytical process were not critically reviewed |
| Grinyer (2010) UK | Interviews | N=11 families - interviews (24 people interviewed) | To evaluate the views of 24 service users – parents, CYP, siblings, guardians and family carers – on their experiences of respite care in the of a children's hospice in northern England. | • data analysis methods were not clearly stated<br>• no discussion on whether saturation was been reached for any of the themes reported<br>• the researchers' roles and potential influences in the analytical process were not critically reviewed |
| Jennings (2014) Ireland | Interviews | N=10 Mothers | To report on research that examined | • convenience sampling strategy used<br>• the relationship |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| | | | mothers' experiences of bereavement support following the death of their child from a life-limiting condition. | between the researcher and the respondents was not clearly reported<br>• no discussion on whether saturation was been reached for any of the themes reported<br>• the researchers' roles and potential influences in the analytical process were not critically reviewed |
| Konrad (2007)<br>USA | Psychological phenomenological design | N = 12 mothers whose child was seriously ill or dying | This article described unexpected findings from a qualitative study with mothers of seriously ill and dying children who supported the value of parent-to-parent connection and mentorship. | • researchers did not critically review their roles in data collection and data analysis process<br>• unclear whether data saturation was achieved regarding data collection and analysis |
| Malcolm (2008)<br>UK | Interviews | A) N=5: Families using hospice services; B) N=44: Hospice staff and volunteers; C) N=18: Professionals associated with the hospice | To generate a list of priority topics for children's hospice care research in Scotland from the perspective of its key stakeholders. | • researchers did not critically review their roles in data collection and data analysis process<br>• unclear whether data saturation was achieved regarding data collection and analysis |
| Maynard (2005)<br>UK | Focus group interviews | N=29 parents from 22 families (of whom 6 were bereaved) | To describe a quality assurance initiative undertaken as 1 component of a clinical governance strategy. | • researchers did not critically review their roles in data collection and data analysis process unclear whether data saturation was achieved regarding data collection and analysis |
| Monterosso (2007)<br>Australia | Phase 1: questionnaires administered either by telephone or face-to-face; Phase 2: Interviews | N=134 parents and 20 service providers. | To explore parents and service providers to better understand the needs of families of children receiving palliative and supportive care about their care | • researchers did not critically review their roles in data collection and data analysis process<br>• unclear whether data saturation was achieved regarding data collection and analysis |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| | | | needs in hospital and in community settings | |
| Price (2013) UK | Focus groups (using the nominal group technique) | N=35 health and social care professionals | To investigate health and social care professionals' perspectives on developing services for children with life-limiting conditions at the end of life using issues identified by bereaved parents as priorities | • Researchers did not critically review their roles in data collection and data analysis process<br>• unclear whether data saturation was achieved regarding data collection and analysis |
| Robert (2012) USA | Focus groups | N= 14 parents from 9 families (out of 25 families who responded to contact attempts). | To describe and being to understand the experience of bereaved parents whose deceased child had received paediatric oncology services at a tertiary comprehensive cancer centre. | • small sample size but acceptable for qualitative study<br>• possible selection bias of participants<br>• participants may have been subject to recall bias but how this was affected by their emotions couldn't be assessed.<br>• researchers did not critically review their roles in data collection and data analysis process<br>• unclear whether data saturation was achieved regarding data collection and analysis |
| Steele (2008) Canada | Interviews | N=11 parents from 6 families | To describe the experiences of parents as their families transitioned in a Children's hospice in Canada | • researchers did not critically review their roles in the data collection and data analysis process<br>• unclear whether data saturation was achieved regarding data collection and analysis |
| Weidner (2011) USA | Interviews and focus groups | N= 29 parents representing 20 families | To identify and define the dimensions of paediatric end of life care that were important to | • participants may be subject to recall bias due to bereavement emotions;<br>• researchers did not critically review their |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| | | | parents of children or infants who died either in hospital or at home under hospice care as a result of an illness, chronic condition, or birth defect. | roles in the data collection and data analysis process<br>• unclear whether data saturation was achieved regarding data collection and analysis |
| **Questionnaires/surveys** | | | | |
| Davies (1996) Canada | Structured questionnaire | N= 15 families | To explore factors and how families cope over time with a child who has a neurodegenerative genetic disorder. | • convenience sampling strategy used<br>• -the relationship between the researcher and the respondents was not clearly reported<br>• no discussion on whether saturation was reached for any of the themes reported |
| Einaudi (2010) France | Questionnaire with open-ended response questions | N=11 parents of deceased children | To understand the parental response to perinatal death by describing the experiences of the families. | • the relationship between the researcher and the respondents not clearly reported<br>• no discussion on whether saturation has been reached for any of the themes reported |
| Forrester (2008) UK | Retrospective cross-sectional survey using a postal questionnaire | N=16 bereaved families whose child remained in a cold bedroom following the child's death | To explore how bereaved families experience the child remaining in a cold bedroom following the child's death | • convenience sampling strategy used<br>• the relationship between the researcher and the respondents not clearly reported<br>• no discussion on whether saturation has been reached for any of the themes reported<br>• The researchers' roles and potential influences in the analytical process were not critically reviewed |
| Remedios (2015) Australia | Questionnaires including standardised psychometric measures and open-ended questions | N=77 carers | To determine the impact of out of home respite care on levels of fatigue, psychological adjustment, quality of life and relationship satisfaction | • researchers did not critically review their roles in data collection and data analysis process<br>• unclear whether data saturation was achieved regarding data collection and analysis |

| Study | Data collection methods | Participants /respondent | Aim of the study | Comments |
|---|---|---|---|---|
| | | | among caregivers of children with life-threatening conditions | |

Four categories/themes of social and practical support during the palliative care, before and after the death of the child found to be helpful emerged from the evidence, which are: social and practical support, respite services, care around and after the child's death, and bereavement support and follow–up.

### 8.2.5 Clinical evidence

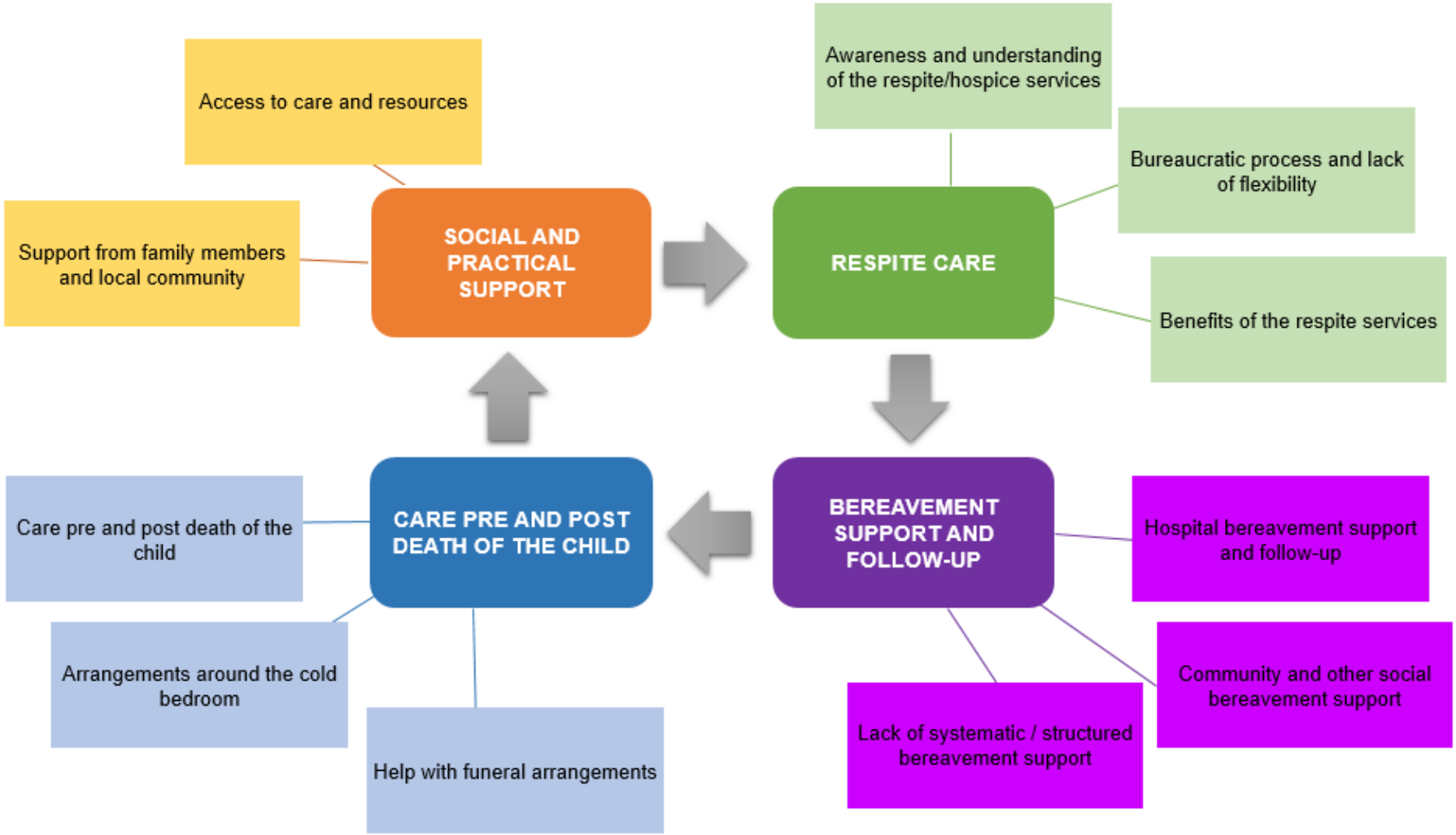
#### 8.2.5.1 Clinical evidence profile

Table 42, Table 43, Table 44 and Table 45 show summaries of clinical evidence of qualitative findings, from adapted GRADE-CERQual.

#### 8.2.5.2 Theme map

Figure 9 shows a theme map relating to social and practical support.

1 **Figure 9: Theme map – social and practical support**




2

3

### 8.2.5.31 Evidence summary

2 **Table 42: Summary of evidence (adapted GRADE-CERQual): Theme 1 – Practical and social support**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Subtheme 1: Access to care and resources** | | | | | |
| 2 studies (Weidner 2011; Remedios 2015) | 1 study used interviews; 1 study used surveys; | Two studies (Remedios 2015; Weidner 2011) from the USA in which interviewed parents reported that they were faced with financial pressure and cost of caring issue when caring their child living with life-limiting conditions. They reported that access to care and resources in terms of financial sources, paperwork, equipment and training would be helpful.<br><br>**Financial pressure and costs of caring:**<br>Income and financial pressure: free-text qualitative data revealed that financial costs of caring, coupled with an inability to work, posed a major difficulty for some families:<br>*"Taken on an extra job [started a business] for extra income as my financial situation is becoming dire. My daughter who attends VSK is having more seizures, waking at night and becoming heavier and taller. My home is not equipped properly for her condition and I cannot afford a larger more equipped house." (ID: 052)*<br><br>**Access to care and resources when the child is cared at home: (financial resources, paperwork, equipment and training),** (*parents*)<br>Many parents talked about the value of having their children at home at the end of life stage and stressed the importance of having enough resources to manage it.<br>Some talked about the financial resources they required and the help they needed to fill out forms and file paperwork. Others talked about the equipment and training | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | they needed to care for their child at home. They also indicated the importance of having help to coordinate these resources so that they could minimise the burden and maximise the time they spent with their child.<br>*"There was on number to call when you have problems, and they contacted the person that you needed at that moment...It wasn't like you had 10 numbers...it made it a lot easier for us."*<br>*"I guess they made you feel that our main concern is our child and being with our child...not coming up with the money for her to be here. They psychologist had contacted my insurance...she had already filled in my insurance company so I didn't have to reiterate the whole situation and try to figure how things were going to work out"* | | | |
| **Sub-theme 2: Support from family members and local community** | | | | | |
| 2 studies (Robert 2012; Konrad 2007) | 1 study used focus groups; 1 study used interviews; | In 2 studies where parents were interviewed they reported that support from their own family and connections with local community members, such as parent-to-parent support group, were helpful.<br><br>**Help and support from those close to the mother:**<br>Support from those close to the mother mainly consisted of positive emotional support, such as advice from the mother's own mother.<br><br>**Social support from the local community:**<br>A locally-based community support system was highly valued by parents.<br>*"He wanted to be with his friends and in classes as much as possible...I would wheel him out and put him in the car – literally pick him up and put him in the driver's seat, put the* | Limitation of evidence | Minor Limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *wheelchair in the back. He would drive to school, call his buddies from class and say, "Hey, I'm in the parking lot. Can you come get me?"...Tons of support in every teacher, principal and student."*<br>Parents emphasised the importance of discussing social support needs with providers and maximising social connections in the treatment plan.<br><br>**Local parent-to-parent support group:**<br>Mothers in this study strongly encouraged parents to seek out the support from other parents and take them as mentors and guides. Shared experience provided these mothers with both useful information and comforting reassurance that they could be competent in their child's care.<br>*"Try to talk to other people who are experiencing the same thing for two reasons: number one to get your hearts connected so that you know that you know, it, it's unbelievably helpful. And also to share the technical stuff or what's going on, um, physically with your kid."*<br>Mothers' stories told comfort generated through informal connections with families who had similar journeys.<br>*"one was a friend of a friend who knew that my son had [disease]…and then another one is someone who lives in town, um, that we were acquaintances with but they had heard our son had it. And I think parents do an incredible job supporting each other...I am not afraid to say to either one of these parties that I would ask a lot of questions...I would appreciate someone telling me what their experience was so I could at least get used to what we were dealing with."*<br>Similarly helpful and comforting connections with local parent–parent-organisations were noted by a few of the | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | mothers. | | | |

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3 **Table 43: Summary of evidence (adapted GRADE-CERQual): Theme 2 – Respite services**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| **Sub-theme 1: Awareness of the hospice and understanding of the services it provides** | | | | | |
| 2 studies (Malcolm 2008; Steele 2008) | 1 study used interviews; 1 study used focus groups; | Two studies reported on this theme. Both HCPs and parents' perspectives were incorporated and reported in the 2 studies.<br><br>**Awareness and greater understanding about hospices and the services they provide**<br>There was unanimous acknowledgement among both HCPs and parents on 1 study carried out in the UK that many myths and misconceptions concerning children's hospices continue to prevail among public and professionals alike. Recognition of the need to develop strategies that would promote a greater understanding of the hospice and assist to dispel existing misconceptions was made.<br><br>It was felt very strongly across all of the participant groups that actively promoting the wide range of care and support provided by the hospice was necessary to increase awareness among the public and professionals and thus improve access to the service, tapping into unmet need: | Limitation of evidence | Major limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *"There is a big issue in terms of getting children and families across the threshold of a children's hospice, a) because of the terminology and b) because of professional misconceptions or lack of education and information that professionals have about what children's hospices do...." (Professional)*<br>*"Well for everyone I would think the first priority is making the health professionals more aware of the service that the hospice offers." (Family)*<br>The same was reported by parents interviewed in another study conducted in Canada<br><br>Understanding and information about respite care services provided by hospices: that is, viewing hospice as a possible resource for family (not only for end of life care):<br>*"[I] never really paid much attention because we thought it was for end of life care. So I thought, well, if we reach that point with [child's name] we'll look into it then." (father)*<br><br>Once parents became aware that the hospice provided respite as well as end-of-life care, they considered CPCH a possibility:<br>*"I heard about it from a friend of mine ... and she said, Why don't you try that place?", but I think from what I knew I thought it was only end-of-life care. I didn't know that they provided respite care." (Mother)* | | | |
| **Sub-theme 2: Benefits of respite care** | | | | | |
| 5 studies (Davies 2004; | 1 study used interviews and | The benefits of respite care was reported in 5 studies conducted in Australia, Canada and the USA). According | Limitation of evidence | Major limitations | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| Champagne 2012;<br>Remedios 2015;<br>Steele 2008;<br>De Cinque 2006) | surveys;<br>3 studies used interviews;<br>1 study used surveys; | to the parents interviewed, apart from them and their child living with LLCs, siblings and other family members also benefited from it as well. For those who thought they did not receive sufficient respite service, they perceived this as an unmet needs.<br><br>**Benefits of respite:**<br>Benefits for parents include getting a break, a sense of freedom, and time for themselves and others:<br>*"When she is here, we can come and get her and take out to do stuff or we can just go and do what we want. I think it was more effective in that just had time to socialize with friends and be on my own so that I was a little sane. I found that when I was really stressed, I was obviously not very pleasant to be around. I mean, it is really to keep your cool when you are going through all these different stresses and then you have teenagers that are on your case about nothing. Just everything happens at once. So you tend to snap a lot faster. So it really was important get away from it. And keep some sense of balance." (Mother)*<br><br>**Benefits for children receiving respite: receiving care, relaxation and enjoyment, learning and socialising**<br>It [Canuck Place children's hospice] was more comfortable than a hospital providing "*less depressing*", surrounding and "better emotional" atmosphere, it was more "*like home*":<br>*"I mean they are [the staff] always getting them involved …Today, she is going to walk down to the corner and watch some film that is being produced in the corner. Little things like that…they went out to the UBC [nearby university] sports facility – they had these off-road wheelchairs that they get to try out. So she had a good* | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *time on those. And trick-or-treating on Halloween, they went all over the place". (mother)*<br>**Benefits for siblings:**<br>Because siblings could also attend school at Canuck Place and could stay overnight, all children in the family had time together away from parents, when they could about the illness and the ill child's prognosis. Parents believed such discussions benefited siblings/child relationship. (author's quote)<br><br>Parents also felt respite care helped them see future in perspectives and get prepared for changes.<br><br>**Dealing with future changes:**<br>"*Parents saw benefits for the future as well. They felt more comfortable dealing with future changes, for example, if the child's health deteriorated and they required further medical interventions. Parents were less afraid about end-of-life care because they realized that CPCH manages more comprehensive care than they could provide at home on their own."* (Author's quote)<br><br>**Unmet Needs – Respite and practical support during palliative phase:**<br>*"It would have given me a break, I could have done things. I could have been stronger for her, I could have fought the battles." (parent)* | | | |
| **Sub-theme 3: Bureaucratic process and lack of flexibility – things that could be improved** | | | | | |
| 3 studies (Grinyer 2010; Eaton 2008; | 3 studies used interviews; | In 3 studies where parents were interviewed, they reported problems they encountered when accessing respite care services. | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of | Coherent | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| Maynard 2005) | | | findings | | |
| | | **Lack of choice regarding respite: in terms of timing and frequency:**<br>"*There seemed to be little choice about when, how often, and for how long respite care was offered. […] what was offered was gratefully accepted, but the timing and frequency of the respite did not always fit with the family's plans or preferences and they felt unable to articulate this for fear of appearing ungrateful*." (author's quote)<br><br>**Inflexibility of the booking system:**<br>Although both hospice and home respite services use a booking system for care, parents can find this too inflexible to meet their needs:<br>*"When you have a crisis with a child like this, it's usually in the middle of the night, on a weekend, a bank holiday, when there's nobody around, or if they are there's a very limited service." (M7)*<br><br>**Practical problems of access:**<br>*Packing and transfer of equipment:*<br>The duration of the respite care was often very short and the complicated preparations necessary were thought by some to be disproportionate to the benefit<br>*'[it's] very difficult packing everything up just for the day – almost not worth the bother'. (mother)*<br>*"We have to trundle the equipment down." (M1)*<br>*"We have to take his potty chair, medication, clothes, nappies, chocolate." (M8)*<br><br>**Transport:**<br>No offers of support with travel to the hospice were | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | reported, and parents, particularly when on their own, could struggle with the practicalities of transporting a very disabled child along with heavy equipment and all their medications.<br>*"It just would have been great if they could have offered a transport service to and from, that would make life easier ... On your own with him in the car ... if he was having a fit in the car or needing oxygen, I would be driving and I would have to pull over.'" (stepmother)*<br><br>**Bureaucratic requirement regarding form filling:**<br>Although it was recognised that records need to be kept up to date, what was seen to be excessive and laborious form filling.<br>*"'More hassle than it's worth'. (mother)* | | | |
| **Sub-theme 4: Access and availability of respite services** | | | | | |
| 1 study:<br>(Monterosso 2007) | 1 study used interviews; | This theme was reported in 1 study carried out among parents in Australia.<br><br>**Lack of funding** to purchase respite and other healthcare services<br>*"Parents spoke frequently about difficulties in procuring funding for various forms of care and perceived a number of barriers and inequities to exist. Although adequate financial and practical assistance was central to care provision and contributed to the quality of life experienced by children and their parents, parents from the non-cancer group especially, articulated the burden they endured as a result of the lack of financial and practical assistance." (Authors quote)* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | Rigid criteria to be admitted:<br>*"Most parents from the non-cancer group used or attempted to access respite and felt this was crucial to the well-being of their children and other family members. However, many parents were hindered by lack of financial support and/or rigid criteria, which limited their access. In contrast, parents from the cancer group rarely felt the need to access respite." (Authors quote)* | | | |

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2 **Table 44: Summary of evidence (adapted GRADE-CERQual): Theme 3 – Care around and after the child's death**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: Care pre- and post-death of the child** | | | | | |
| 1 study:<br>(Forrester 2008) | 1 study used surveys; | This theme was reported in 1 study. Parents reported that they appreciated the continuity of care and of personnel, opportunity to be with their child, and care provided to other members of the family at this moment.<br><br>**Continuity of care and personnel:**<br>Some parents reported that continuity of care of personnel pre- and post-death of the child was important. They appreciated knowing who was caring for the child and eventually putting them in their coffin:<br>*"They popped in and tucked * [in] at night and kept the music on for * and cared for * physically … with grace and dignity as if * were their own child' (R 15).*<br>**Deterioration of the child's body:** | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *'A slightly surreal experience' (R 13)*<br>*'I found it very hard to be with * cold body'* (R 15)<br><br>**Opportunity to be close to the child** – importance that the child was not taken away (cover by communication review):<br>*"Easy access at all hours to go see, touch *, stroke * hair, talk to *" (R 9).*<br>*"To have had * taken away would have been unbearable" (R 5)*<br>*"We did not want to be parted from * until we had to" (R 14)*<br>*"We could take * from the security of the hospice to the crematorium without being parted" (R 13).*<br>*"There are no memories of a death at home and the difficulties that accompany that" (R 14)*<br>*"I wanted * not to die at home so that there was not a room I did not want to go in" (R 4).*<br><br>**Care for the family:**<br>The attention given to the families' physical needs (for example, ***meals being provided***). The importance of staying together as a family (for example, ***the opportunity to have accommodation at the hospice, a family room***):<br>*"Kept us together until we had to say goodbye" (R 13).* | | | |
| **Sub-theme 2: Arrangement around the cold bedroom after the child's death** | | | | | |
| 1 study: (Forrester 2008) | 1 study used surveys; | This theme was reported in 1 study carried out among parents. | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of | Coherent | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | **Opportunity to see or not to see the cold room:**<br>*"It felt like we were being shown another room; the full impact of what it would mean didn't register; the rooms were not chilled at that point" (R 10).*<br><br>**The décor of the room was important** (room needs feel warm, natural):<br>*"The room was homely, peaceful, like a child's bedroom…we were told it could be kitted out…really to represent one's own home"*<br>*"You could make the room into something your child would have loved…the room gave me comfort…"*<br><br>**Care for the family members around the cold room**: for example, provide family members with:<br>• warm jackets for parents to wear<br>• for family with another child: *"for the sibling to be able to go in and out of the room without restriction"*<br>• comforting music. | findings | | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |
| **Sub-theme 3: Help with funeral arrangement** | | | | | |
| 1 study<br>(Forrester 2008 | 1 study used surveys; | **Help with funeral arrangement**<br>Respondents commented on how they valued help with making funeral arrangements, including making appointments to register the death and with funeral directors. Ten respondents commented that the funeral directors visited them at the hospice. Five appreciated access to poetry/prose materials for use at the funeral service.<br>*"We managed to organise what we wanted"* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of findings | Applicable | |
| | | | sufficiency or saturation | Saturated | |

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2 **Table 45: summary of evidence (adapted GRADE-CERQual): Theme 4 – Bereavement support**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: Hospital bereavement support and the continuity of follow-up** | | | | | |
| 3 studies (Contro 2002; Contro 2012; De Cinque 2006) | 3 studies used interviews; | This theme was reported in 3 studies where parents and HCPs were interviewed. Both commented that bereavement follow-up from the hospital was helpful for parents after their child passed away.<br><br>**Hospital bereavement support** (for example, staff attending funeral)<br>Many parents felt that contact from oncology unit staff both during palliation and bereavement was important:<br>*"But then it would have been nice if they [hospital staff] had said, 'Come for a check-up' or just don't drop her like that. I think that's the biggest mistake you can do." (parent)*<br>*"I thought that I'd have the phone call and they'd [hospital staff] say, 'How are you coping?', and that sort of thing. So it was very different to what I expected." (parent)*<br>Bereavement follow-up: continued contact from the hospital staff<br><br>This was noted by both parents and HCPs.<br>Continued contact with hospital staff after their child's death was meaningful to the families who spent time at LSPCH. Follow-up by telephone, mail, and/or in person was desirable and appreciated:<br>"*The phone calls are important. When her doctor called, I thought, "Wow, you're still thinking of us!" The nurse practitioner still calls periodically. When your child is sick like that, it becomes your life and the doctors and nurses* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *become your extended family. If they can continue some kind of periodic contact, it's important*. (Unclear quotation owner)"<br><br>**Continuity of relationship as vital to the bereavement process (HCP's perspective):**<br>Although staff identified continuity of relationships as vital to the bereavement process for them and for the families, they could rarely maintain these connections. ... family members who felt alone and abandoned by their *"hospital family"* after the death of their child.<br>*"We need continuing support for families so they don't feel forgotten. If you have the choice between more or less, more is better because parents can always decline. But I think reaching out to families is best so they feel they are still remembered."*<br>*"Families often feel no one really understand their situation except people at the hospital – but then they are abruptly cut off from these very people they have come to rely on"* | | | |
| **Sub-theme 2: Community's and other social bereavement support** | | | | | |
| 4 studies<br>(Price 2013; Jennings 2014; De Cinque 2006; Contro 2012) | 1 study used focus groups;<br>3 studies used interviews; | ***Bereavement support from the community*** and other social relationships were reported as a theme in 4 studies. However, some parents noted that community bereavement support organised and provided by their local communities was not always helpful for them.<br><br>***Bereavement support for siblings and grandparents*** was reported to be important as well by parents.<br><br>**Community bereavement support:** | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | *"And I also strongly recommend that they be very careful about the counsellors that they go to because there is a lot of counsellors out there but there's very few, very few who can really assist. And I mean in a concrete way with skills and, and in a way that they can ask questions that go down deeper than just the bereavement because the bereavement is the top layer." (parent)*<br>*"Our parish priest was marvellous and Sister Margaret. They came down and helped organise the funeral and they knew what they were doing and that was a great help." (parent)*<br><br>**Keeping the memory of the deceased child alive:**<br>*"He'll never be gone from my memory. He will always be there and I think that's really important." (Participant 7)*<br>*It was also achieved by attending their (mothers) deceased child's grave on their own, which was important to their adjustment to bereavement:*<br>*'I would go to the grave twice a day; it was like it was her school, it was her time ... It was almost like a ritual. I went up in the morning and again in the afternoon.' (Participant 3)*<br>*"Bar his bed clothes nothing has changed in his room ... He's not in the house but he's everywhere ... I sleep with Tom's pyjamas under my pillow. Every night I take them out." (Participant 10)*<br>*"I have a massive memory box with all her stuff. And her first tooth ... I have her lock of hair and bits and pieces ... videos we had taken of her. I have all that upstairs and I think that will all just stay." (Participant 6)*<br><br>**Informal source of bereavement support:**<br>***Family and friends:*** | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | Some mothers said that family and friends were hugely supportive and helpful:<br>*"I think friends and family are the main ... my friends, that's what got me through ... friends and neighbours. 'Cos they're there, not the milestone moments, just the normal moments." (Participant 7)*<br>Others mentioned that websites or online chat rooms were supportive, especially in early bereavement.<br>*"When I went back to work ... no one asked me anything. Nothing. No conversations about her with anyone at all. They didn't say anything." (Participant 6)*<br><br>***Group meetings among parents:***<br>The mothers felt supported by attending group meetings, through meeting other parents who had also experienced the death of their child:<br>*"It was good hearing other people's stories and they had the same kind of feelings ... I don't know, it's kind of a general companionship or something being with other people that you don't feel like you're the only one." (Participant 1)*<br><br>**Contact with other bereaved parents:**<br>(covered by information review):<br>*"Other parents from the bereavement group would come out and have a coffee or have a chat and reassure me. I found that was very, very helpful to know that I could talk to somebody else who had lost their child and had experienced losing a child ... . You do hold back your feelings and you need somebody else that has been there." (parent)* | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | Some parents felt the oncology unit should link them with other bereaved parents who could offer support:<br>*"I think there should be someone tied up with the ward that has experienced it. I think at the end of the day it will help you cope with the situation better. There should be someone there who understands that it's a terrible thing to lose a child". (parent – mother)*<br><br>**Bereavement support and needs for siblings and grandparents:**<br>Most of the interviewed expressed deep concern about the paucity of services offered to siblings prior to, at the time of, and after the death of the child. When siblings did receive help, it was often because parents had requested it. Staff also identified other close to the child, for example grandparents, who experience great distress and yet rarely received services. | | | |
| **Sub-theme 3: Lack of systematic/structured bereavement support** | | | | | |
| 3 studies (Price 2013;<br>Einaudi 2010;<br>Contro 2012) | 1 study used focus groups;<br>1 study used surveys;<br>1 study used interviews; | In 3 studies where parents and HCPs were interviewed, they reported that lack of structured bereavement support system was an issue in providing support to parents. The barrier caused by language and culture differences was also reported by HCPs in 1 study.<br><br>**Structured bereavement support:**<br>"*Participants ranked structured bereavement support for families as the most important priority for service development … Significant professional and personal dilemmas arose when families expected bereavement support to be provided, often over the long term, by those previously involved in care. These dilemmas included: feelings of guilt at not being able to provide the support* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of findings | Applicable | |
| | | | sufficiency or saturation | Unclear | |
| | | | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *required; 'burn-out' and consequent diminution in the ability to provide effective nursing care; and the potential for families' 'recovery' through bereavement to be jeopardized through an over-dependency on individual care team members"* (Authors quote)<br><br>Lack of structured bereavement support was also reported by HCPs in another study (Contro 2012).<br><br>**Lack of systematic bereavement follow-up after the child passes away:** (HCPs' perspective)<br>Several mentioned that they gave written information to families about the grieving process and provided contact information for any available community resources. However, not 1 person interviewed knew how often families followed up on these referrals.<br>*"The gap in care is the follow-up. We do the immediate care, but often don't have time to follow up with families. They physician should offer an appointment 3 months out to answer any questions a family might have. Parents could always decline it, but at least they would have the opportunity."*<br>*"There is a lack of organisation and systematic follow-up with families after the death of a child. There needs to be funding and hospital support for bereavement activities."*<br><br>**Timing of bereavement follow-up and support after the child's death:**<br>Booklets about the grieving process should be distributed 2 months or more after the death of the child, when many families find themselves in a "social desert" after the support of the first few weeks has waned. | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | **Language and culture issue during bereavement follow-up:**<br>Concern for the care of non-English speaking families, particularly in bereavement follow-up, was frequently expressed. There is a descending level of care depending on the language of the family: English-speaking families receive the most care, followed by Spanish-speaking families (with the help of bilingual staff and interpreters).<br>Families who speak languages other English or Spanish receive little or no bereavement follow-up.<br><br>Several staff reported feeling helpless when trying to serve non-English speaking families, despite the fact that the hospital has an exceptional interpreter service.<br>*"Many [non-English speaking] families don't understand what is going on and it is very difficult for them. Many things are lost in translation and staff feels particularly helpless when they don't speak the same language as the family." (child-life specialist)*<br>*"The interpreters are very good, but it is very difficult to use interpreters when dealing with bereavement issues. Consequently, sometimes the follow-up for these families just doesn't happen" (social worker)* | | | |

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### 8.2.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis as it was thought that there would not be evidence on the effectiveness of competing alternatives.

Respite care is an important component of social and practical support and Table 46 provides illustrative unit costs for this service in alternative settings. The Committee agreed that £1,000 was a reasonable estimate of the per diem cost of hospice respite care.

**Table 46: Illustrative unit cost of respite care**

| Service | Cost per bed-day | Source |
|---|---|---|
| Paediatric respite care [a] | £657 | NHS Reference Costs 2014-15 |
| Hospice respite care [b] | £1,082 | PSSRU 2015 |

*(a) Currency code PX55Z; Based on a NHS Reference Unit cost of £1,505 for a mean length of stay of 2.29 days*
*(b) This is based on a costing for longer life illness trajectories (cardiac care) and a cited figure of £16,233 for 15 days of respite care per annum*

### 8.2.7 Evidence statements

A number of themes emerged from the interviews with parents or healthcare professionals. They were: social and practical support, respite services, care pre- and post-death of the child, and bereavement support and follow-up.

#### Practical and social support

Moderate to low evidence from 5 studies conducted among parents showed that parents thought that support to help them access care and resources available, and support from family members and local community such as parent-to-parent group was helpful.

#### Respite services

Moderate to very low quality evidence from 11 studies in which parents or healthcare professionals were interviewed, suggested that regarding respite services, raising the awareness and understanding of it would be helpful. Parents also thought that they and their child living with a life-limiting condition benefited from respite services greatly, and this benefit extended to other family members. However, parents and healthcare professionals both pointed out that there were things to be improved regarding respite services, which were mainly reflected by the bureaucratic process involved such as the booking system, and the lack of flexibility regarding the timing and frequency of respite services. Some parents also reported that they had financial difficulties in procuring all forms of services.

#### Care around and after the child's death

Moderate quality evidence from 1 study where parents were interviewed they reported that around the death of their child, they appreciated the continuity of the care and of personnel pre- and post- death of their child. They also appreciated the care provided to other family members at this moment. The same study also reported on parents' views and perspectives on the "cold bedroom" after the child's death. Opportunity to see the "cold bedroom" (where the child's body was kept soon after the child died), the "cold bedroom" feels warm, homely, and peaceful was deemed helpful. Also, parents appreciated care provided to themselves and the child's siblings in the "cold bedroom."

**Bereavement support**

Moderate quality evidence from 6 studies where parents and healthcare professionals were interviewed, they reported that bereavement support from hospital staff, such as follow-up calls and the continuity of relationship was very helpful for the bereavement process. Parents also found that bereavement support from their local community and other social bereavement support groups (such as contact with other bereaved parents) was helpful. However, some parents reported that they didn't always find bereavement support from their local community helpful, and commented on the need for counsellors to have appropriate skills and experience. Furthermore, lack of systematic and structured bereavement support after the child's death was noted as an area that needed to be improved. Some healthcare professionals also noted that there were language and cultural barriers during bereavement follow-up when providing follow-up support to parents from minority ethnic groups.

## 8.2.8 Linking evidence to recommendations

### 8.2.8.1 Relative value placed on the themes considered

Evidence on the majority of themes considered important during the protocol development as well as further themes that emerged from the evidence was identified. The Committee focused their discussion mainly on the following themes that were reported in the evidence review: respite support; continuity of care and of healthcare professional staff when the child or young person is approaching the end of life; caring for the parents or carers and family members when the child is approaching the end of life; bereavement support; information sharing among organisations after the child's death, and training for healthcare professionals involved in bereavement support.

### 8.2.8.2 Consideration of different practical and social support needs

Based on the evidence, the Committee thought it important to raise awareness of parents' or carers' individual needs when the child or young person is approaching the end of life. Depending on the child's or young person's conditions, those needs could include alterations to the family's home, supply of equipment and training in how to use it, respite care and financial support. Some of those needs also applied to the wider family, such as siblings.

Continuity of care and of care staff, in particular, was another theme that emerged from the evidence and was considered by the Committee. They agreed that there should be a plan in place to help families to receive care from professionals with whom they are familiar. This would also facilitate continuity of who communicates with the family and provides the relevant information. The Committee discussed that repeatedly having to tell different healthcare professionals the same information, was reported to be a cause of frustration and anger.

The Committee also discussed the role of healthcare professionals in enabling children and young people to access education and the importance of both the educational and social aspects of school and college for children and young people trying to live well with a life-limiting condition.

The Committee acknowledged that while NICE guidance does not extend to education, there are often significant barriers assessing to schooling when a child or young person has a life-limiting condition in some regions.

The Committee then discussed care for family members when the child or young person is approaching the end of life. As supported by the evidence, the Committee concluded that practical support around this time should include the provision of information and advice on practical issues related to the death of the child or young person (such as funeral arrangement, registration of death, and coroners processes).

The Committee recognised the importance of bereavement support to families, which was identified in the evidence review as a theme, and discussed how this could be provided and who should be involved. The Committee thought that it is important to make healthcare professionals aware that the process of bereavement starts before the death of the child or young person. They noted that it is important to identify a healthcare professional who has expertise in bereavement support and who is ideally already known to the family. The role of the family's general practitioner was also discussed. It was felt that general practitioners can play an important role in bereavement support for family members and that to do this well they need to be informed of what information and options for support have already been provided by healthcare professionals in the multidisciplinary team around the child or young person.

The Committee also discussed that there are different means of providing information about bereavement support to families. It was for instance, noted that information about bereavement should be provided both verbally and in written format because families may not always be able to process everything at the time they are told about it.

In the evidence, families reported a range of different approaches that they found helpful in dealing with their bereavement, such as meetings with staff who cared for their child, attending bereavement support groups in their local community, and also the provision of support to the child or young person's siblings. The individual differences in preferences were considered to be important by the Committee.

The Committee discussed the fact, which was also a theme in the evidence report, that there was a risk to families if relevant databases were not promptly updated following the death of a child. For example appointments might be mistakenly offered through automated processes causing upset to the family. They made a recommendation on this matter to avoid this risk.

The issue of training in bereavement support was also discussed. It was noted that staff do not always have the relevant expertise to support bereaved families and that this could lead to a breakdown in trust between the multidisciplinary team and the family. It is therefore important to refer bereaved parents or carers to those with the right skills and expertise to provide the service. The Committee also discussed training and on-going support and supervision for healthcare professionals to develop skills in providing compassionate bereavement support for families, but acknowledged that this was beyond the scope of the guidelines as the review did not address the effectiveness of different approaches to training, support and supervision of staff.

The Committee additionally considered and discussed the impact that the death of a child or young person can have on healthcare professionals who have provided end of life care. Feeling of stress and burn-out could result from this, and staff therefore need support to deal with these situations.

#### 8.2.8.3 Economic considerations

Social and practical support has resource implications. For example, the provision of material support for housing adaptations and access to respite care all have cost implications, although not all these costs would be met by the NHS. There are aspects of social and practical support which facilitate objectives such as the provision of home-based community care and therefore the Committee felt they warranted the expenditure.

The Committee agreed that the number of days of respite care that could be offered to children and young children was not unlimited and that in the absence of resource constraints more days/nights of respite care would be provided. The Committee also agreed referral to finite respite care was more straightforward for progressive conditions with a clearer diseases trajectory.

The Committee was also aware that there was a statutory duty to provide 'short breaks' for carers, in particular the Breaks for Carers of Disabled Children Regulations, 2011. While most of that duty rests with local authorities, advice from the Department of Education is explicit in stating that the NHS has a direct funding duty for breaks for children with complex needs, which includes the funding of children's hospice provision. It states: *"Health services have multiple roles to play in the provision of short breaks for disabled children in their areas. They will directly provide and commission some services, for example, short breaks for children with complex health needs. (For some children, this may involve spending some time in a hospice.)".*

#### 8.2.8.4 Quality of evidence

Moderate to very low quality evidence was found in the review. The main concerns with regard to the quality of the evidence were:

- Self-selection bias and recruitment bias: in many studies only about half or less than half of the people who were contacted consented to be interviewed. People who chose to participate may be different in many ways to those that did not want to take part.
- Lack of saturation: most studies did not report whether they collected sufficient data to explore the topic fully which means that there could have been other practical and social needs that were not reported. However, when considering the evidence as a whole saturation was achieved on some meta-synthesised themes.
- Lack of the critical review of the researcher's role in sample recruitment, data collection or data analysis process. Few studies clearly reported the relationship between researchers, interviewers and the respondents. This could be a problem because a pre-existing hypothesis may bias interviews and the analysis.
- Lack of verification of findings: few studies verified their findings with participants or external sources, nor reported the reason why verification was not necessary or applicable. This means that it was unclear whether the findings were applicable and generalisable to all people in similar situations.
- Applicability: findings from the majority of included studies are applicable to the UK setting because of the direct relevance of their populations, contexts, and the topics explored.

Due to the uncertainty in data saturation or sufficiency of many findings in this review, the Committee interpreted the evidence with caution.

#### 8.2.8.5 Other considerations

The Committee discussed that some of the social and practical support needs that were identified from the evidence were also consistent with some of the themes that were picked up in the focus groups that were run for this guideline. Particularly with regard to continuity of care. Children and young people were frustrated by having to tell different healthcare professionals the same information.

The Committee discussed whether they wanted to prioritise this topic for a research recommendation, but they concluded that the combination of the evidence (including the focus group report), their experience and their expertise provided sufficient information to base their recommendations on.

#### 8.2.8.6 Key conclusions

The Committee concluded that healthcare professionals should be aware of the parents' or carers' individual needs for practical support when their child has a life-limiting condition. They emphasised the importance of continuity of care and care for the extended family (including siblings and grandparents). Bereavement should be considered before the child or young person's death. Identification of a key professional in the provision of bereavement

support should be planned in advance. The evidence also highlighted that healthcare professional have support needs and the Committee agreed that guidance was needed to address this. The role of primary care, including GPs, in the support for families was also important. It is important for healthcare professionals to discuss with families whether aspects of their cultural and religious background have important implications for how healthcare professionals should provide for the individual needs of the child or young person and their family. There are different approaches to bereavement support and parents or carers should be informed about all available options.

### 8.2.9 Recommendations

**72. Be aware that continuity of care is important to children and young people and their parents or carers. If possible, avoid frequent changes to the healthcare professionals caring for them.**

**73. Be aware that children and young people with life-limiting conditions and their parents or carers have varied social and practical support needs, and that those needs may change during the course of their condition. This may include:**

- material support, for example housing or adaptations to their home, or equipment for home drug infusions
- practical support, such as access to respite care
- technical support, such as training and help with administration of drug infusions at home
- education support, for example from hospital school services
- financial support.

**74. Discuss with parents or carers the practical arrangements that will be needed after the death of their child, and provide this information in writing. This should cover matters such as:**

- the care of the body
- relevant legal considerations, including
  - the involvement of the child death overview panel
  - the involvement of the coroner
  - registration of the death
- funeral arrangements
- post-mortem examination (if this is to be performed).

**75. When a child or young person is approaching the end of life, discuss the bereavement support available with their parents or carers and provide them with written information.**

**76. When a child or young person is approaching the end of life, talk to their parents or carers about available psychological bereavement support groups.**

**77. Offer bereavement support to the parents or carers both before and after the death of a child or young person.**

**78. When planning bereavement support for parents or carers:**

- talk to them about the support that is available and explore with them what they would find helpful and acceptable

- think about what support different professionals could provide, for example:
  - their GP
  - healthcare professionals who know the child or young person and are involved in their care
- think about the role of individual healthcare professionals in providing specific aspects of support
- inform the multidisciplinary team about the support plan.

**79. When making a bereavement support plan with parents or carers, discuss possible options with them such as:**

- opportunities to talk to the professionals caring for the child or young person, to:
  - discuss memories and events
  - answer any concerns or questions they may have
- home visits from the healthcare professionals caring for the child or young person
- bereavement support groups.

**80. Give professionals involved in the care of the child or young person opportunities to talk about and explore their thoughts and feelings:**

- when the child or young person is approaching the end of life and
- after the child or young person has died.

**81. Following the death of a child or young person, ensure that relevant healthcare and other professionals are informed in a timely manner.**

**82. Update relevant documents and databases after the death of a child or young person (to avoid, for example, clinical appointments being offered by mistake).**

**83. Ensure that healthcare professionals providing bereavement support have the necessary expertise.**

# 8.3 Religious, spiritual and cultural support

## 8.3.1 Review question

**What factors of spiritual or religious support (including care of the body) are effective in end of life care of infants, children and young people with life-limiting conditions and their family members or carers and what influences attitudes about these before and after death?**

## 8.3.2 Introduction

Receiving a diagnosis of childhood life-threatening condition and facing death and bereavement often moves children, young people and family members to search for meaning in these events and to reflect on cultural, ethical, religious, faith or spiritual questions connected to the meaning and purpose of life, illness and death.

End of life care planning decisions may also generate ethical and value conflicts for individuals or between family members. Parents of children with genetically heritable life-limiting conditions may face additional dilemmas around future family planning options that affect them at a spiritual and cultural level.

Some families may have a strong connection to a belief system or community which provides clear support and guidance for managing end of life and after death care for the child. Healthcare professionals and systems need to enable families to honour, respect and follow religious and spiritual practices of life and death in a timely manner and in all places of care.

Children and young people and their families may also experience dilemmas, struggles, distress or “crisis” in relation to beliefs and values and may seek spiritual or religious guidance to express fears, doubts and anxieties and reflect on the ways in which illness and death may challenge spiritual beliefs. For other individuals and families their spirituality, values and beliefs may be less well defined or they may be trying to manage complexities of blended family belief systems. Individualised care for some children and families may involve supporting families seeking to make meaning of experiences and uncertainty at an ethical or meta-physical level when medicine can only offer explanation at a biological or material level.

Hospitals and hospices typically offer chaplaincy or multi-faith support services, which provide access, if individuals wish to, to both spiritual guidance and a space for prayer, meditation and reflection and to perform rites and rituals. In addition to this distinct service, all healthcare professionals can integrate respect and support for spiritual needs of the child and family with all aspects of care. However these needs may go unrecognised if professionals are uncomfortable with discussing these issues and avoid doing so.

## 8.3.3 Description of clinical evidence

The mixed-methods approach was taken because it allowed for the inclusion of different study designs (both quantitative and qualitative) in order to fully understand areas of concern. The aim of this review was to investigate both the effectiveness of interventions as well as to explore people’s perspectives related to this topic.

For the quantitative part of the review, the objective was:

- To assess the effectiveness of spiritual and religious support for children and young people with a life-limiting condition who are approaching the end of life, and their family members or carers

- To look for systematic reviews, randomised control trials, cohort studies and uncontrolled studies.

No evidence was found which met the inclusion criteria for this part of the review.

For the qualitative part of the review, the objectives were:

- To identify and describe the factors that influence children and young people living with a life-limiting condition and their families or carers attitudes towards religious and spiritual support.
- To identify and describe children and young people living with a life-limiting condition and their families or carers experiences with religious and spiritual support, challenges faced, unmet needs, ethical issues.
- To look for studies that collected data using qualitative methods (such as semi-structured interviews, focus groups, and surveys with open-ended questions) and analysed data qualitatively (including thematic analysis, framework thematic analysis, content analysis and so on). Survey studies restricted to reporting descriptive data that were analysed quantitatively were excluded.

Fourteen studies were identified (Boss 2008, Ebmeier 1991, Forrester 2008, Forster 2014, Foster 2009, Hexem 2011, Jones 2006, Lundqvist 2003, Meert 2005, Meyer 2006, Reder 2009, Robinson 2006, Talbot 1996, Zelcer 2010). Of them:

- 13 studies focused on the perspective of parents who were caring for a child with a chronic or a life-limiting condition or whose child had died due to an acute illness or a life-limiting condition (Boss 2008, Forrester 2008, Forster 2014, Foster 2009, Hexem 2011, Lundqvist 2003, Meert 2005, Meyer 2006, Reder 2009, Robinson 2006, Talbot 1996, Zelcer 2010)
- 1 study involved siblings (Foster 2009)
- 2 study involved healthcare professionals (Jones 2006, Reder 2009)
- 1 study involved children hospitalised for an acute illness or exacerbation of a chronic condition (Ebmeier 1991)

With regard to the countries in which the studies were conducted:

- 10 studies were conducted in the USA (Boss 2008, Ebmeier 1991, Foster 2009, Hexem 2011, Jones 2006, Meert 2005, Meyer 2006, Reder 2009, Robinson 2006, Talbot 1996,)
- 2 in the UK (Forrester 2008, Zelcer 2010)
- 1 in Australia (Forster 2014)
- One in Sweden (Lundqvist 2003)

With regard to the methodology of the studies:

- 7 studies collected data by interviewing the participants (Boss 2008, Forster 2014, Foster 2009, Hexem 2011, Meert 2005, Robinson 2006).
- 4 studies used surveys or questionnaires (Forrester 2008, Jones 2006, Meyer 2006, Talbot 1996)
- 2 studies used focus groups (Reder 2009, Zelcer 2010)
- 1 study used storytelling, based on the grounded theory qualitative approach (Ebmeier 1991)

The most common data analysis method employed across studies was thematic analysis.

Evidence on all themes considered important by the Committee was identified. A number of further themes or sub-themes emerged from studies were also identified and incorporated in the review.

A summary of the included studies is presented in Table 47

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

For presentation of findings, a theme map was generated according to the themes emerged from studies (Figure 10). The mapping part of the review was drafted by 1 researcher but the final framework of themes was further shaped and when necessary re-classified through discussions with at least 1 other researcher from the guideline technical team. Due to the qualitative nature of these studies, evidence is summarised in adapted GRADE-CERQual tables within the evidence report. Therefore no separate Appendix is provided for this.

### 8.3.4 Summary of included studies

#### 8.3.4.1 Quantitative review

No evidence was found which met the inclusion criteria for this part of the review.

#### 8.3.4.2 Qualitative review

A summary of the studies that were included in this review are presented Table 47.

**Table 47: Summary of included studies**

| Study | Data collection methods | Participants | Aim of the study | Comments |
|---|---|---|---|---|
| Boss 2008<br>USA | Interviews | 26 mothers of infants who died as a result of extreme prematurity or a lethal congenital anomaly | To explore parental decision-making regarding delivery room resuscitation for infants born extremely prematurely or with potentially lethal anomalies. | • The relationship between the researcher and the respondents not clearly reported<br>• The data collection process and discussion on whether saturation has been reached for of the themes was reported<br>• The researchers did critically review their own roles in the process. |
| Ebmeier 1991<br>USA | Storytelling | 28 children hospitalised for an acute illness or exacerbation of a chronic condition | To understand children's relationship with God during an illness experience | • Unclear sampling strategy used.<br>• The relationship between the researcher and the respondents not clearly reported<br>• Unclear discussion on whether saturation has been reached for any of the themes reported<br>• Researchers did not critically review |

| Study | Data collection methods | Participants | Aim of the study | Comments |
|---|---|---|---|---|
| | | | | their own roles in the process<br>• No details given about analysis saturation<br>• The researchers' roles and potential influences in the analytical process critically reviewed;<br>• Indirect study population, as < 50% were hospitalised due to a chronic condition |
| Forrester 2008<br>UK | Survey | 16 bereaved families whose child had been cared for in a cold room. | To describe how bereaved families experience the use of a cold room following the child's death. | • Convenience sampling strategy used. The Authors were unable to establish contact with many eligible families.<br>• The relationship between the researcher and the respondents not clearly reported<br>• No discussion on whether saturation has been reached for any of the themes reported<br>• Researchers did not critically review their own roles in the process<br>• Data analysis' methods not stated.<br>• Retrospective survey |
| Forster 2014<br>Australia | Interviews | 12 bereaved parents and 10 healthcare professionals | To describe the role of communication in the construction of meaning around post-mortem care. | • The relationship between the researcher and the respondents not clearly reported<br>• No details provided in relation to data collection methods<br>• No discussion on whether saturation has been reached for any of the themes reported<br>• Researchers |

| Study | Data collection methods | Participants | Aim of the study | Comments |
|---|---|---|---|---|
| | | | | critically reviewed their own roles in the process<br>• The researchers' potential influences in the analytical process were not clearly reviewed• |
| Foster 2009<br>USA | Interviews | 40 families of children who died of cancer (36 mothers, 27 fathers and 40 siblings) | To explore bereaved parents' and siblings' reports of legacies created by children with advanced cancer. | • The relationship between the researcher and the respondents was not reported<br>• No discussion on whether saturation has been reached for any of the themes<br>• Researchers did clearly review their own roles in the analytical process<br>Only study that includes siblings |
| Hexem 2011<br>USA | Interviews | 73 parents of children who had enrolled in the Decision Making in Paediatric Palliative Care study. | To describe the role of religion, spirituality and life-philosophy in the life of parents of children with life-threatening conditions. | • Sample selection clearly reported.<br>• The relationship between the researcher and the respondents not clearly reported<br>• No discussion on whether saturation has been reached for any of the themes reported<br>• Researchers critically reviewed their own roles in the process but was unclear whether saturation in terms of analysis was achieved.<br>• Parents who reported not having a religion and/ or spiritual 'feeling' were not interviewed further. |
| Jones 2006<br>USA | Survey | 131 members of the Association | To identify the needs of children | • Convenience sampling strategy |

| Study | Data collection methods | Participants | Aim of the study | Comments |
|---|---|---|---|---|
| | | of Paediatric Oncology Social Workers. | with cancer and their families at the end of the child's life. | used.<br>• The relationship between the researcher and the respondents not clearly reported<br>• No discussion on whether saturation has been reached for any of the themes reported<br>• Researchers did not critically review their own roles in the process, no details given about analysis saturation |
| Lundqvist 2003<br>Sweden | Interviews using standardised questionnaire | 11 Muslim women who had given birth in Sweden. | To explore Muslim women's views of neonatal end of life care in Sweden. | • The relationship between the researcher and the respondents not clearly reported<br>• No discussion on whether saturation has been reached for any of the themes<br>• Researchers did critically review their own roles in the process.<br>• This study only included Muslim women.<br>Indirect population, as not all women had experienced foetal impairment or neonatal death<br>• This study only includes Muslim women.<br>• Indirect population, as not all women had experienced foetal impairment or neonatal death |
| Meert 2005<br>USA | Interviews | 33 parents of children who died at the PICU | To explore parents' spiritual needs at the time of their children's death in the PICU and during bereavement. | • The relationship between the researcher and the respondents was reported<br>• Data collection process and |

| Study | Data collection methods | Participants | Aim of the study | Comments |
|---|---|---|---|---|
| | | | | discussion on whether saturation has been reached for any of the themes were both clearly reported<br>• The analytical process was described in detail; researchers did critically review their own roles in the process and saturation in terms of analysis was achieved<br>• Indirect population, data was not reported separately, 69% of children died as a result of a chronic condition and 31% of children died as a result of an acute illness or injury |
| Meyer 2006 USA | Open-ended questionnaire | 55 parents whose children had died after the foregoing of life-sustaining treatment | To explore the priorities and recommendations, from a parental perspective, regarding end of life communication. | • The relationship between the researcher and the respondents not clearly reported<br>• Data collection process and discussion on whether saturation has been reached for any of the themes reported<br>• Researchers did critically review their own roles in the process.<br>• Self-administered questionnaires<br>• Mixed religious backgrounds, although most of them were Catholic or Protestant<br><br>Same population as Robinson 2006, different themes reported<br>• Same population |

| Study | Data collection methods | Participants | Aim of the study | Comments |
|---|---|---|---|---|
| | | | | as Robinson 2006, different themes reported |
| Reder 2009<br>USA | Focus groups | 39 participants, including bereaved parents, paediatricians, and nurses | To investigate the concept of hope for families and paediatric healthcare professionals during a child's serious illness. | • The relationship between the researcher and the respondents was not reported<br>• Data collection process clearly reported; no discussion on whether saturation has been reached for any of the themes reported and about the roles of the researchers<br>• Researchers did clearly review their own roles in the analytical process<br>• Saturation in terms of analysis was not discussed |
| Robinson 2006<br>USA | Self-administered questionnaire | 56 parents whose children had died in the ICU after the foregoing of life-sustaining treatment | To identify the nature and the role of spirituality from the parent's perspective at the end of their child's life in the PICU. | • The relationship between the researcher and the respondents not clearly reported<br>• Data collection: process and discussion on whether saturation has been reached for any of the themes reported<br>• Researchers did critically review their own roles in the process.<br>• Mixed religious backgrounds, although most of them were Catholic or Protestant<br>• Same population as Meyer 2006, but different themes reported |
| Talbot 1996<br>USA | Self-report questionnaire and interviews | 80 bereaved mothers | To describe mother's attitudes about life 5 or more years after the | • Convenience sampling.<br>• The relationship between the |

| Study | Data collection methods | Participants | Aim of the study | Comments |
|---|---|---|---|---|
| | | | death of their only child. | researcher and the respondents not clearly reported<br>• No details given about data saturation<br>• Researchers did not critically review their own roles in the process<br>• Findings/results: Results were presented clearly. The researchers' roles and potential influences in the analytical process critically reviewed;<br>• Mostly protestants<br>• This study includes indirect population, as 73% of mothers had lost their child following an accident. |
| Zelcer 2010 UK | Focus groups | 25 parents of 17 children who had died of a brain tumour | To explore the end of life experiences of children with brain tumours and their families. | • Small sample size, gathered from a single institution. The Authors were unable to establish contact with many eligible families.<br>• The relationship between the researcher and the respondents not clearly reported<br>• Data collection process clearly reported<br>• No discussion on whether saturation has been reached for any of the themes<br>• Researchers critically reviewed their own roles in the process |

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## 8.3.5 Clinical evidence

### 8.3.5.1 Quantitative review

No evidence was found which met the inclusion criteria for this part of the review.

### 8.3.5.2 Qualitative review

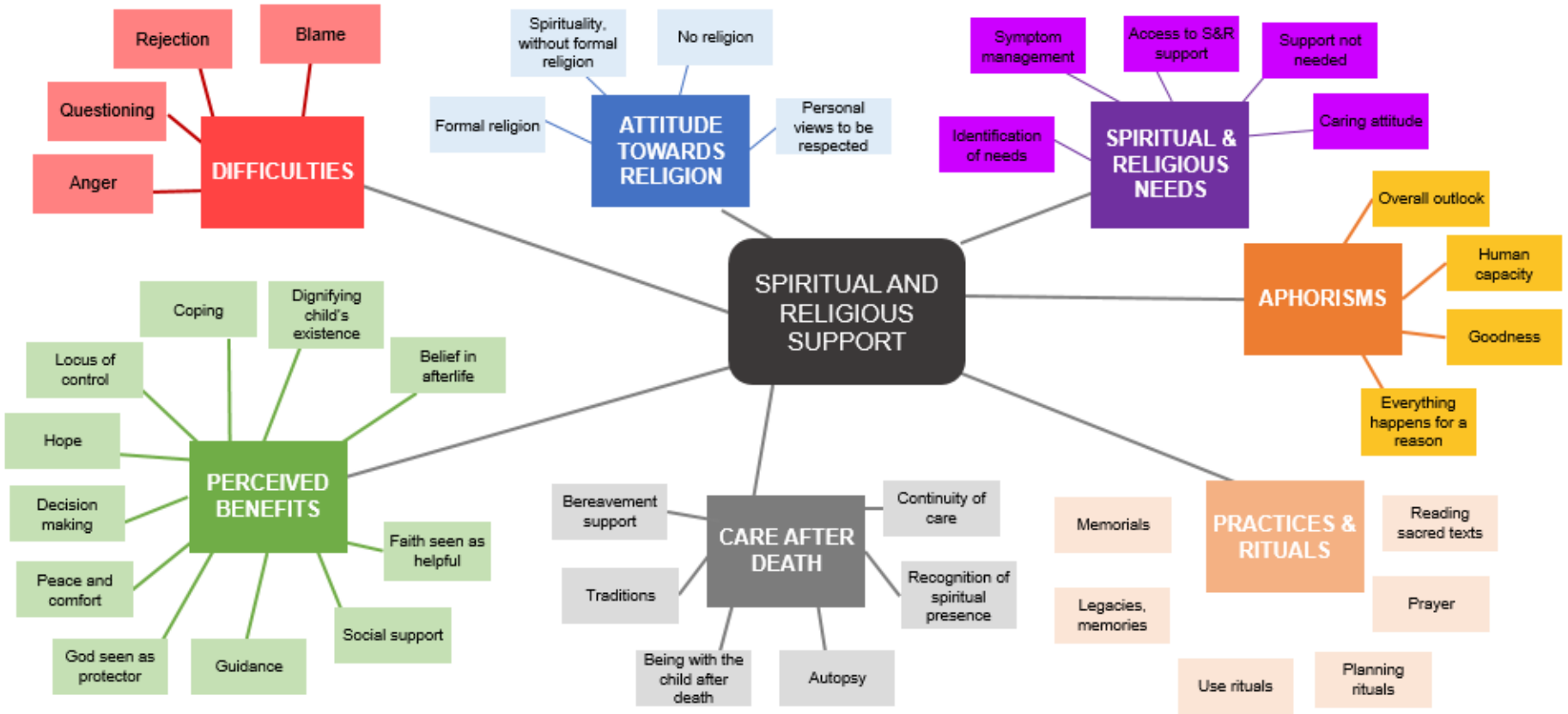
#### *8.3.5.2.1 Clinical evidence profile*

The clinical evidence (adapted GRADE-CERQual) for spiritual and religious support is presented in Table 48, Table 49, Table 50, Table 51, Table 52, Table 53 and Table 54

#### *8.3.5.2.2 Theme map*

The theme map for spiritual and religious support is presented in Figure 10.

**Figure 10: Theme map – religious, spiritual and cultural support**



1 **Table 48: Summary of clinical evidence (adapted GRADE-CERQual): Theme 1 – Attitude towards religion**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: having a formal religion** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, some parents identified themselves as members of a particular religious faith, and described their affiliations very positively:<br>• *"We're Presbyterian and we have a church that we're very involved in, and that's been a wonderful support." (parent)* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 2: spirituality of life philosophies, without formal religion** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, some parents described themselves as not regular church attendees still often felt a connection to God or sense of spirituality:<br>• *"If I want to talk to God, I just will." (parent)*<br>• "*I haven't been drifting toward any type of spirituality; I don't know what kind of spirituality it would be, but it would probably be my own*." (parent) | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: unwilling to discuss their views** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, while most parents reported some level of religious, spiritual, or other beliefs or observances, some answered the inquiry with a quick *"No," "No, not really," or "Umm, no"* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: no beliefs** | | | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| 1 study (Forrester 2008) | 1 survey | In 1 study conducted in the UK with families that had lost a child, some parents reported having no beliefs. | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 5: personal views to be respected** | | | | | |
| 1 study (Robinson 2006) | 1 survey | In 1 study conducted in the USA with parents whose child had died in the ICU, some parents refrained from offering specific advice to other parents, spiritual or otherwise, some noting that each person's situation was *"too personal and subjective."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

1

2 **Table 49: Summary of clinical evidence (adapted GRADE-CERQual): Theme 2 – Spiritual and religious needs**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: identification of needs** | | | | | |
| 1 study (Robinson 2006) | 1 survey | In 1 study conducted in the USA with parents whose child had died in the ICU, 1 parent specifically noted the pivotal role of healthcare team members in identifying when spiritual care might be beneficial: | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | • *"The nurse was extremely helpful ... making suggestions for a chaplain."* | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 2: support not needed** | | | | | |
| 1 study (Forrester 2008) | 1 survey | In 1 study conducted in the UK with families that had lost a child, some parents indicated that they did not need spiritual support. | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: access to spiritual and religious support** | | | | | |
| 2 studies (Meert 2005, Robinson 2006) | 1 interview 1 survey | In 2 conducted in the USA with parents whose child had died in the PICU, parents identified the importance of ready access to both their own familiar community clergy person and the hospital chaplain, as well as a chapel:<br>• *"The services of my rabbi [were most helpful]."*<br>• "Allowing our minister ... to have access to us."<br>• *"If someone is gonna come in and say a prayer, I would just have liked it to be someone of my religious persuasion. They had the wrong kind of collar walk in our room". (Parent)*<br>• *"[A] discussion with our pastor confirming we had the scriptural authority to make these decisions [withdrawal of life-sustaining therapies] was very helpful."*<br>• *"I think it is nice that there's a chapel available. I used it basically just as a place that was quiet". (Parent)* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: symptom management** | | | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| 1 study (Jones 2006) | 1 survey | In 1 study conducted in the USA, social workers identified that symptom management should be holistic:<br>• *"I think families and children need pain management that is physical, mental and spiritual"* | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 5: caring attitude** | | | | | |
| 1 study (Meert 2005) | 1 interview | In 1 study conducted in the USA with parents of children who died at the PICU, it was highlighted that parents valued care and empathy:<br>• *"Several times he would come in and check on my baby even though his part was done, and you just knew that he cared." (Mother)*<br>• *"That little personal titbit that he shared connected me to him. You know that he would think enough, feel enough, that he would share that with me." (Mother)*<br>• *"All he told me is, 'M___ expired.' And he turned around and went his way. And I said to myself, 'He's so cold.'" (Mother)*<br>• *"I remember one nurse taking me by the hand and she prayed with me, and talked to me, gave me a hug and told me it was going to be all right." (Mother)*<br>• *"'He probably can't see much anyway because of all the medication he's on, it's probably just a blur.' I know she didn't mean anything by it and maybe that's truthful but it seemed a little insensitive. My son is sick and dying and I didn't need to know that probably he can't focus on anything anyway." (Mother)* | Limitation of evidence | Major limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |



1

**Table 50: Summary of clinical evidence (adapted GRADE-CERQual): Theme 3 – Common aphorisms**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: overall outlook** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, parents offered statements pertaining to their overall outlook on the situation:<br>• *"That's just life"*<br>• *"What's going to happen is going to happen"*<br>• While some phrases referenced the sacred:<br>• *"It's in God's hands"* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 2: goodness** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, parents frequently mentioned the quality of goodness:<br>• *"God is always good."*<br>• *"I just believe in God and I try and find the good in things."*<br><br>Additionally, some parents described their children's presence in the world as a gift:<br>• *"Every day is a gift, because she was only given three days [to live]. So every other day with her is a gift."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: human capacity** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, parents spoke about their sense of human capacity, or how a given parent expected to function in the situation:<br>• *"We're not given more than we can handle."*<br>• *"One day at a time, one step at a time, one mile at a time."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: everything happens for a reason** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, a statement that parents used most often was that: *"Everything happens for a reason."*<br>Parents seemed to identify their religion with that statement:<br>• *"I do believe in that higher faith, so I believe that there was a reason why [our child] was put here, given to us."*<br><br>Just because parents believed there were reasons, however, did not mean they always found those reasons easy to accept:<br>• *"I think there's a reason for everything. I'm not always happy about it."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

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2 **Table 51: Summary of clinical evidence (adapted GRADE-CERQual): Theme 4 – Practices and rituals**

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: prayer** | | | | | |
| 4 studies (Ebmeier 1991, Forrester 2008, Hexem 2011, Robinson 2006) | 1 interview, 2 survey, 1 story telling study | In 2 studies conducted in Australia and the USA with parents of children with parents of children receiving end of life care, most parents reported praying for their children, both alone and in prayer groups.<br>• *Prayer was found to be a helpful coping strategy, and parents would advise other parents to pray:*<br>• *"We ... prayed a tremendous amount."* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | • "Pray for strength."<br>• *"Be strong and pray."*<br>• *"Pray and don't be afraid to ask the staff questions."*<br>• *"Pray!"*<br><br>One important aspect of prayer was that it could happen anywhere:<br>• *''The chapel is here, but I feel like you don't have to be in a chapel to pray.'' (parent)*<br><br>Similarly, another study conducted in the UK following the death of the child, parents using cold rooms referred to the importance of praying:<br>• *"I can pray anywhere and at any time" (R10)*<br><br>In 1 study conducted in the USA with 28 hospitalised children, the children in the study referred to the children in their stories as praying to God in a formal sense:<br>• *"He/ she would say a prayer".*<br>• *"God could you please make me feel better"*<br>• *"Please help me not to be afraid"*<br>• *"Thank you God, for helping me get well"*<br>• *"He's praying to God that, well, I hope it does not hurt and I hope I get out pretty soon".*<br><br>They also referred to children praying informally or just talking to God:<br>• *"Please help me"*<br>• *"Make me better"* | saturation | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | • *"Please help me get through without getting hurt"*<br>• *"Why do I have to go through it, I don't understand"*<br>• | | | |
| **Sub-theme 2: reading the sacred text** | | | | | |
| 2 studies (Hexem 2011, Meert 2005) | 2 interviews | In 1 study conducted in the USA with parents of children who died at the PICU, parents viewed meditation on sacred text as an important activity<br>• *"I needed my Bible and that's why I always work with" (parent)*<br><br>In another study conducted in Australia with parents of children receiving paediatric palliative care, many parents reported reading the Bible in response to stressful life events.<br>For example, 1 parent chose to read the Bible stories of Job and of Abraham, saying, *"All the trials they went through in life and how their faith in God brought them through – that helps me a lot." (parent)* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: planning rituals** | | | | | |
| 1 study (Jones 2006) | 1 survey | In 1 study conducted in the USA, social workers said that many parents describe the importance of rituals:<br>• *"[Families need] spiritual support and involvement in planning rituals around death"* | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: use of rituals: candles, music** | | | | | |
| 2 studies (Forrester 2008, Meert 2005) | 1 interview<br>1 survey | In 1 study conducted in the UK with families that had lost a child, some parents referred to the importance of rituals:<br>*"Candles were lit all through our stay" (R16) (parent using cold* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of | Coherent | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *rooms)*<br><br>In another study conducted in the USA with parents of children who died at the PICU, parents relied on spiritual songs as a source of strength.<br>Other religious rituals described by parents included baptism and last rites. These were primarily requested to ensure the child's safe passage to afterlife. | findings | | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 5: legacies, memories** | | | | | |
| 3 studies (Foster 2009, Lundqvist 2003, Meert 2005) | 2 interviews, 1 survey | In 1 study conducted in the USA with bereaved families (parents and siblings), it was suggested that many children did things to be remembered (such as writing letters, giving special gifts or personal belongings…):<br>• A sibling said, *"She [20-year-old] gave me lots of stuff. She gave me like a bunch of old t-shirts because she loved t-shirts. Those are special."*<br>• One 3-year-old explicitly talked to her mom about giving away belongings so others could remember her: *"One thing that me and her did before she passed … we talked about her belongings. And each one of her nurses was to have a specific toy that she had. And she told me, she says, "Mommy, you have to give it to them after I am gone. And they have to know that I wanted them to have this to remember me*."<br>• Another sibling reported: *"Yeah, he [16-year-old] did [made] a cement stone, like a stepping stone. And he put sign language "I love you" and he put like "sis" on the bottom … He did [one] for my mom, my dad, my grandma, and good friends of his."*<br><br>Few ill children explicitly said their intent was to be | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | remembered, yet their actions implied that this was their wish:<br>• *One mother said: "We never had like the one moment to talk about that. But so, she [13-year-old] made these crafts … flowers out of paper. We have that as a token of her. (Interviewer: So even though she didn't do it purposely to be remembered by …) No, no, no. But she just made it there [in the hospital] and she goes, "Okay, mommy, I thought you'd like one." Or when her aunts would come or her cousins, she actually made some for them, too. (Interviewer: So she wasn't aware that she was passing, but she made things for everybody?) Yeah. Yeah."*<br><br>Other family members perceived that their child with cancer did not need to do or say anything to be remembered:<br>• A father said, *"I think she [14-year-old] was well aware of how deeply loved she was. So she didn't need to leave anything behind."*<br>• A mother shared, *"I asked her [17-year-old] actually if there was anything that she wanted me to relay to anybody, and she said, 'nope' cause everybody knew it from her that she loved them … She never wanted to be famous or anything, but she wanted to be remembered."*<br>• One sibling shared how his 19-year-old brother living with advanced cancer realised that he had already left behind a legacy: *"Before he died, he told me and his girlfriend and mom. He goes, "Before I die, I want to carry out a legacy or do something that nobody else has ever done." Then, 2 weeks later he goes, "You know, I have carried out a legacy. I've been like a dad to (sibling), and I've treated him like one more than the real dad did." And he goes, "I've already done what I needed to do."*<br><br>Similarly, in 1 study conducted in the USA with parents of | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of theme or finding | Criteria | Rating | Overall |
| | | children who died at the PICU, parents described that memories of specific events during hospitalisation that approximated usual child-rearing experiences were especially comforting to them:<br>• *"They were feeding J___ through tubes. You know it's hard to see your child with tubes through his nose. They took him out of the crib and they let me hold J___ and my husband held the syringe that feeds. And they said, 'You want to feed your baby?' You know that's what we have now are those memories." (Mother)*<br>• *"You know, I just always think, she's in my heart, and I'm walking with her in my heart, you know." (Mother)*<br>• *"They wouldn't let us take anything. I wanted her gown because it was the last thing she wore. I wanted the sheet from the bed, I wanted her bracelet from the hospital. They said they couldn't give us anything." (Mother)*<br>• *"The chaplain took pictures of her and cut a lock of her hair and gave it to me. It was supportive, you know, she really cared." (Mother)*<br>• *"I didn't even want to forget the pain because I feel like if I lose it, then I'm kind of forgetting her." (Father)*<br><br>In another study conducted in Sweden with Muslim women, some mothers said that mementoes were forbidden in their religion:<br>• *"Photos is okay before the baby is death. When the baby is dead, no. I don't want it. Other mementos than pictures of the living baby is too hard to look at. We are not doing that. All that reminds us of the baby is given to someone else in order to forget" (Woman3)*<br>• *"Mementos do not support [me]. You will have the baby in your heart"* | | | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | | | | |
| **Sub-theme 6: memorials** | | | | | |
| 1 study (Meert 2005) | 1 interview | In 1 study conducted in the USA with parents of children who died at the PICU, parents found memorials useful:<br>• *"So one of my needs that was meet spiritually was coming back to the hospital for the memorial service that they offer. That meant so much to me coming here again because, I guess, this is where I left him".* | Limitation of evidence | Minor limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Not applicable | |
| | | | Sufficiency or saturation | Not saturated | |

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2 **Table 52: Summary of clinical evidence (adapted GRADE-CERQual): Theme 5 – Perceived benefits**

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: social support** | | | | | |
| 2 studies (Hexem 2011, Meert 2005) | 2 interviews | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, parents mentioned that participating in a particular religious community resulted in parents receiving support from a large number of fellow congregants, the pastor and God. Support from others ranged from phone calls, e-cards and cooking meals to people praying for the child and family:<br>• *"People we don't even know [are] praying for this little guy."*<br><br>Parents benefited from prayer groups, and saw the church as providing *"a network"* and a source of *"unconditional support and love."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | • Pastors were occasionally referred to as *"good friends."*<br>• Parents also felt supported by God:<br>• *"Casting all your care to Him gives you the feeling that you're not alone."*<br><br>In another study conducted in the USA with parents of children who died at the PICU, parents also felt that spiritual support was received from others. These included spouses, parents and other family members, friends, neighbours, co-workers, clergy, health professionals, and parents of other PICU patients:<br>• *"And if somebody's there by theyself [sic], please try to get somebody there to be with them. I think that's more important than anything 'cause nobody should have to go through that alone." (Parent)*<br>• *"I used to surf [the Internet] and I'd meet people online, talk to parents who have children with the same problem and who lost their kids and stuff. 'Cause talking to someone with the same problems, whose child died with the same hypoplastic left heart as mine, exchanging stories and stuff was good. That helped a lot. It was encouragement." (Mother)*<br>• *"Their prayers, their hugs, just being there, just knowing they cared. I remember when S___ was 12 and he had open heart surgery. He was in the sixth grade and the outpouring of cards and letters and pictures and it just meant so much to us. I think that's what always helped me was to know that people cared and that they would be there to help." (Mother)* | | | |
| **Sub-theme 2: faith seen as helpful** | | | | | |
| 1 study (Robinson 2006) | 1 survey | In 1 study conducted in the USA with parents whose child had died in the ICU, Parents identified their faith in God as most | Limitation of evidence | Minor limitations | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | helpful to them at the end of their child's life and they would suggest it to other parents who were facing similar situations:<br>• *"My faith and knowing that my child had the same faith."*<br>• *"My faith and trust in God who was in charge of Jessie. Knowing she would not suffer no more when she went home to be with the Lord."*<br>• *"The people God provided for us along the journey, friends, family, doctors, nurses, clergy."*<br>• *"Put your faith in God."*<br>• *"Trust in God."* | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: peace and comfort** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, parents reported that feelings of trust in God resulted in feelings of peace and comfort:<br>• *"It comforts us as parents spiritually to think that hopefully, when she passes, she'll have an opportunity [in Heaven] to do [normal] things and it's just a happy place."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: guidance** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, some parents associated their religion with trying to be good:<br>• *"I am supposed to be taking care of my child, and therefore going home and being lazy ... that would be wrong."*<br><br>Parents sometimes contrasted their religious, spirituality and life philosophy beliefs with their beliefs in the medical profession. Sometimes a pastor was seen as being able to | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | mediate between the parents and the doctors:<br>• *"[Our pastor] can understand a lot of the things that the doctors need him to process [for] us on our belief level." (parent)* | | | |
| **Sub-theme 5: help in decision-making** | | | | | |
| 4 studies (Hexem 2011, Lundqvist 2003, Meyer 2006, Robinson 2006) | 2 interview, 2 surveys | There were 4 studies conducted in Australia, Sweden and the USA with parents of children receiving palliative care which reported that spiritual and religious beliefs were helpful in the decision-making process.<br><br>Several parents advised others to honour and be guided by their own values as a way to approach difficult end of life decision-making:<br>• *"Based on your own values and decisions, make the best choice you can."*<br>• *"Do what you feel is emotionally right for you, your family, and your child."*<br>• *"Know when to say enough is enough."*<br>• *"Ask yourself, would I want my child to have a poor quality of life if he/she survives?"*<br><br>Some parents sought the formal guidelines of their religion; as 1 parent said:<br>• *"I want to know what the church teaches on extraordinary measures as to ordinary measures, to give you comfort about DNRs and how far do we go, and just something to really be at peace about." (parent)*<br><br>For some parents the decisions were less difficult when they felt as if they knew or accepted God's will: | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | • *"Knowing that there is a God, that gives me peace, and it helps me to deal with the difficult decisions." (parent)*<br>• *"I believe in God, that it is God who has given me this ill baby and it is His will that I shall take care of the baby. God has given me the medicine too, but I will not take part in any discussion" (Woman2)*<br>• *"No, I don't want to participate in a conversation about it [withdrawing], I think it is God who makes the decision, I am being very distressed, it is too difficult to talk about it" (W8)* | | | |
| **Sub-theme 6: hope** | | | | | |
| 4 studies (Boss 2008, Keene-Reder 2009, Robinson 2006, Zelcer 2010) | 1 interview, 1 survey, 2 focus groups, | Four studies conducted in the UK and the USA with parents of children receiving palliative care or bereaved parents found hope to be a recurrent theme.<br><br>Regardless of medical information, parents maintained hope that everything would be fine, and this guided most parents' decision-making. They were told by friends and family members to pray for miracles, and to trust that a miracle will happen. Some parents felt that they did not have to make a decision regarding resuscitation in the delivery room, they wanted physicians to do everything they could, and the rest was in God's hands. The families also described the need to hold to 2 beliefs: the realisms of the poor diagnosis, and the search for a miracle:<br>• *"You always have that hope that this is going to be the one that solves everything; you don't want to give that up" (FG2)*<br>• *"I could not be the one to decide if God chooses to take the baby away at this time or just let it run its course" (mother of an infant diagnosed prenatally as having trisomy 18)*<br>• *"When they told me they thought she was not going to survive, I put it in God's hands. God had made her into a* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *baby, and if I had made it that far [with the pregnancy], it was up to him"*<br>• *"You know everyone told me don't worry about what [the doctors] say, she will make it, she's a miracle. And so that's pretty much I heard"*<br>• *"There was a lady who said 'you know this child has all these problems, why are you going to bring him into the world? Are you looking for God to step in?' I said 'Well, as a matter of fact I am' If you think God is going to come in and perform a miracle, you have a right to do that."*<br><br>For some parents, hope was also related to acceptance:<br>• *"For me, I believe that you have to have some type of spirituality first to get through any situation in life, but as far as being hopeful, it's like, okay, this is bad, but we want to be able to make a good day, just life each day" (parent)*<br>• *"I accept hope as acceptance that no matter what happens, it's going to be okay and this kind of spells out everything" (parent)*<br>• *"I think hope from a family standpoint is driven by love that we can't even conceptualize personally in that situation as healthcare providers. I think it's the love for that child that drives that hope. I think that's maybe an element of the parent–child bond" (nurse)* | | | |
| **Sub-theme 7: making meaning of the situation** | | | | | |
| 3 studies (Meert 2005, Meyer 2006, Talbot 1996) | 2 interviews, 1 survey | Three studies conducted in the USA with parents who had lost their child reflected that religious beliefs help parents to make meaning of the situation.<br><br>Some mothers have learned from bereavement, and integrated this into a new identity: | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | • *"After Bobby's death I found compassion for other people that I did not know existed in my personality. I can walk in a room sometimes now and I can zero in on the person that's in the room that is hurting terribly for whatever reason. It's like a homing device. It has – Bobby's death has made me a much better person. It's made me aware that everyone out there in the entire world belongs to a family. And everybody loves; everybody grieves; everybody hurts; everybody has joy. It's another lesson that God is teaching me in this journey that I'm on to survive the death of my son" (Irene)*<br><br>Parents also showed thankfulness for their life and their children's lives<br>• *"I just look at the blessing part of it. In spite of her dilemma, I got an actual chance to experience her, and she got a chance to experience daddy and momma. So I feel thankful for that." (Father)*<br>• *"To the day I die, I will find some meaning in what happened to my daughter, whatever it takes. I refuse to believe that she lived on this earth for 14 months and had no impact on anybody or anything. I am not going to allow that to happen." (Father)*<br>• *"He was put here for a reason, and them 9 years, he had a good life. He brought a lot of joy in people's lives. He knew people in the church and he knew people in the streets. I've seen him melt hearts of people that were ice cold. Maybe that's why he was put here, you know." (Parent)*<br><br>However other parents showed a loss of self-purpose after their child's death:<br>• *"And at the funeral, when I closed the casket, part of me went in that casket." (Parent)*<br>• *"She's just like the centre focal point of our marriage and our* | Sufficiency or saturation | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *lives now. You know, without her, I just don't know if I could do it." (Mother)* | | | |
| **Sub-theme 8: coping** | | | | | |
| 2 studies (Hexem 2011, Meyer 2006) | 1 interview | In 2 studies conducted in Australia and the USA with parents of children receiving paediatric palliative care, parents reported that feelings of trust in God helped them coping with the situation and with their anger.<br>• *One mother remarked on the need to keep her "Christian cool" when communicating with a doctor.*<br>• *Another said, "Every time I'm mad or upset, I start writing to Him."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 9: locus of control and patience** | | | | | |
| 1 study (Hexem 2011) | 1 interview | Parents talked about what aspects of their child's medical situations they could and could not control.<br><br>One parent contrasted *"wanting to plan things, to control things" with her religion's teachings, which she said helped give her patience and gave her the ability to "think things through." (parent)* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 10: dignifying child's existence** | | | | | |
| 2 studies (Hexem 2011, Meert 2005) | 2 interviews | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, and another study conducted in the USA with parents of children who died at the PICU, many parents found their religious beliefs helpful in dignifying their child's existence and specialness:<br>• *"where [our child] fits in God's plan and why children like her may be born and, actually, their very special significance."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | • *"It would be wrong for me to just say that, well, her life isn't really important. You know, she is not as important as some of the other kids because she's disabled and she would be much happier in a different place. That's not true because, in my Bible, anyway, every person is important to God, equally important."*<br>• *"And my oldest son had said, 'Regardless of her cleft lip Momma, she's so beautiful, you can't even see that. She is so pretty.' I said, 'Isn't she.'" (Mother)*<br><br>Other parents saw their children as having a role on earth to help bring people together spiritually:<br>• *"[Our child is] like Mother Teresa; she would walk into a room, and everybody would be around her, you know? And so I say that she's brought down here to bring all these people together and to show [them] something."*<br><br>Parents also found that their child's death dignified their existence too:<br>• *"God allows me to see life grow right in front of me, and how beautiful life is. And I don't get so caught up in frustration like I used to. We get so caught up in our own daily life that we forget what life is really all about. God allows us to see in our kids, life itself. And we forget sometimes, with all the other things we go through. Between M___ and J___'s deaths, I try not to forget that. And when a baby is born, how good they smell. We forget that sometimes. How beautiful life is." (Mother)* | saturation | | |
| **Sub-theme 11: belief in afterlife** | | | | | |
| 3 studies | 2 interviews, 1 survey | Three studies conducted in Australia and the USA with parents of children receiving palliative care and bereaved families | Limitation of evidence | Minor limitations | MODERATE |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| (Foster 2009, Hexem 2011, Robinson 2006) | | described parents' beliefs regarding an afterlife and a relationship that endures beyond death.<br><br>Parents used many different words to describe life for their children after their deaths, including: ''afterlife,'' ''a life after this life,'' ''golden gate,'' ''a better place,'' ''a happy place,'' and ''heaven.''<br>• As 1 parent mentioned: '*The peace is there, knowing that, in the end, ultimately, while we won't have immediate perfection, we'll have complete perfection in heaven.''*<br><br>A large number of participants recounted deceased children's beliefs about an afterlife. Many talked about children believing they would go to Heaven or be with Jesus after they died:<br>• *''Nine days before she died, she told me that she was going to go be with Jesus soon … 'God's put peace in my heart'.'' (mother)*<br>• *''She [3-year-old] told me not to worry about it that she was going to make it all right with her friend. She went to Care-a-Lot Heaven. And knew she was going and told me that I would be there to meet her at the spot when it was my time to go. So she knew she was going. And I'll never forget that. Being able to know that she would not forget me. Because she would be waiting for me at the spot. Knowing that, I knew she was going to be fine.'' (mother)*<br><br>This belief in an afterlife was found to be *''reassuring,'' providing ''peace'' and ''acceptance,'' and helped parents to be ''not afraid'' of their children's deaths and ''trust in God to take care of [our child].''*<br><br>Some parents said: | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | • "*He [16-year-old] said, 'I'm gonna go now, okay … I'm gonna go to Heaven.' … he said he was gonna be okay." (Father)*<br>• *"I knew that she was really gone. She gave me a very big smile, so I know that wherever she is, she is okay and she was telling me that "Mom, it's okay." That's why I'm not worried. I know she's okay and I know she wanted to be okay with whatever or however." (mother)*<br>• *"If I don't come home, don't feel sorry for me, be envious of me." (mother)*<br><br>Some parents offered heartfelt, emotionally charged advice to other parents, emphasising the undeniable love and transcendent nature of the parent–child relationship that never dies but rather continues beyond death:<br>• *"Keep talking to your child – let your child know that you are OK. That it is OK for them to go on. I held my daughter and never stopped talking to her, reassuring her. It helped me to tell her that she would always be with me, so strong in my heart."*<br>• *"To know that [you] will never forget your child."*<br>• *"Just remember that they lived a good life and you did everything possible for your children and also believe they are in no pain anymore and that their [sic] up in heaven happy and always watching over you like you watched over them and never forget how special they were."* | | | |
| **Sub-theme 12: God seen as protector, comforter, loving** | | | | | |
| 1 study (Ebmeier 1991) | 1 storytelling | In 1 study conducted in the USA with 28 hospitalised children, the attributes assigned to God were, as a whole, positive. God was seen as helper-protector, comforter, counsellor and judge. God would help the child feel better, go home, or *"get through* | Limitation of evidence<br>Coherence of findings | Major limitations<br>Coherent | VERY LOW |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | *this".*<br>• *"Oh, thank you, you know what, nurse, I think God helped me get through this. I think if God was never here – I don't think I could – I think I'd cry and scream and stuff".*<br>• *"God's powerful" (9 year-old child)*<br><br>God was also seen as reassuring the child, and this was reflected in sayings like this:<br>• *"You'll be fine"; "You're going to be all right"; "nothing's gonna happen to you"*<br>• *God either told the child not to be afraid, gave the child a reason for the procedure, or reassured the child it would not hurt:*<br>• *"God's saying it won't hurt. It'll just feel like a little pinch. Don't worry, don't worry, the shot won't hurt".*<br>• *God's love and concern was also raised by the children:*<br>• *"He loves him, so he'll make the shot not hurt so bad"*<br>• *"He cares for him. He loves him and he's taking good care of him"* | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

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2 **Table 53: Summary of clinical evidence (adapted GRADE-CERQual): Theme 6 – Perceived difficulties**

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: questioning** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, many parents reported | Limitation of evidence | Minor limitations | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | questioning their faith, experiencing feelings of anger and blame toward God, and rejecting of specific religious beliefs and communities:<br>• *"No matter what, it's hard. There is pain. You don't want to let go."*<br>• *"I believe I will have a spiritual connection with my daughter when she passes, but just right now, I have a lot of questions that are unanswered, so I fluctuate back and forth."* | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 2: anger** | | | | | |
| 3 studies (Hexem 2011, Talbot 1996) | 2 interviews, 1 survey | In 2 studies conducted in Australia and the UK with parents of children receiving paediatric palliative care, parents expressed their anger at God and their faith, although some said that their anger was not incompatible with their faith.<br>• "I do believe in God, but I'm kind of angry at him right now."<br>• *"I have the question in my mind, why, why us? What did we do wrong? What did she do wrong?"*<br>• *"Just when I needed my faith, I hated it, for deceiving both my child and myself!"*<br><br>In another study conducted in the USA with mothers who have lost their only child, some mothers showed ambivalent feelings about living, remaining angry at God and/ or their church and were unable to incorporate their child's death into a beneficial belief system:<br>• *"Don was my life. He's what I looked forward to in getting old and him getting married and having a life and making me grandmother and havin' my house filled with little kids runnin' around, and there's nothin' now – absolutely nothin' – and it has – it's made me so angry and it made me so angry at God that this happened… Everything that I had, that I looked forward to in getting old, was taken and it's like my mind just* | Limitation of evidence | Minor limitations | MODERATE |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Saturated | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | *stops right there. I can't see any further than that. I can't imagine what else there would be. I want someone to tell me what I'm supposed to be doin'." (Parent)* | | | |
| **Sub-theme 3: rejection** | | | | | |
| 1 study (Hexem 2011) | 1 interview | In 1 study conducted in Australia with parents of children receiving paediatric palliative care, some parents moved away from their faith as a result of a child being seriously ill:<br>• *"I used to be a lot more religious, and I've had a really hard time with it."*<br>• *"I'm not going to sit and pray and hope that [my child] gets better. We're going to bring her to the hospital."* | Limitation of evidence | Minor limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: blame** | | | | | |
| 1 study (Meert 2005) | 1 interview | In 1 study conducted in the USA with parents of children who died at the PICU, parents felt the need to attribute the child's death to a specific person, place, circumstance or God.<br>• *"But, as far as I'm concerned, God did the worst thing possible He could have done to me and my wife. I mean, take the only thing in the world that meant anything to us." (Father)* | Limitation of evidence | Moderate limitations | LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |

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2 **Table 54: Summary of clinical evidence (adapted GRADE-CERQual): Theme 7 – Care of the body**

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| **Sub-theme 1: recognition of spiritual presence** | | | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| 1 study (Foster 2014) | 1 interview | In 1 study conducted in Australia with bereaved parents and healthcare professionals, nurses acknowledged that for some parents it is important to recognised the spiritual presence of the child:<br>• *"I think it all depends on what you believe, I mean, some people think that, you know, once you're gone, you're gone. But I think mum was a lot happier with the idea that even though [child's] body was there, you know, his spirit was still there and it wasn't so much the body that I was talking to. It was the spirit or how she felt about it" (Nurse4)* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Applicable | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 2: continuity of care** | | | | | |
| 1 study (Foster 2014) | 1 interview | In 1 study conducted in Australia with bereaved parents and healthcare professionals, nurses said that it is important to treat a deceased body as 1 would treat a family member who had died:<br>• *"But yes, I think just personally treat the person like they're still there basically, or how I would want to be treated or how the parents want their child to be treated" (Nurse4)*<br><br>They also raised the importance of performing bodily care as if the child could still feel:<br>• *"I think just a bit of respect for the family and for him. Like, it was only half an hour ago that he was still with us and now he's gone and I don't know. I guess we don't know where they're gone" (Nurse6)* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 3: special tradition** | | | | | |
| 1 study (Lundqvist 2003) | 1 interview | In 1 study conducted in Sweden with Muslim women, some participants said that their religion prescribes some ceremonies in the way the body should be wrapped and washed: | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of | Coherent | |

| Study information | | Description of theme or finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | • *"It is the religion, it is a special person that has to wash the baby. It is not I. If it is a women, a woman does it. If it is a man, a man does it. But with my baby it doesn't matter, but the mother and the family don't have to do it. It's because of that they know the baby and it is too hard for them. Not the staff, it is a special washing" (Woman6)* | findings | | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 4: autopsy** | | | | | |
| 1 study (Lundqvist 2003) | 1 interview | In 1 study conducted in Sweden with Muslim women, they reflected that when asked regarding the autopsy (cause of death not clear or need for further investigation), and many parents found this frightening. They said a dead infant is still living in a sense, but in another shape, and an autopsy would impede this, and the death infant feels the pain:<br>• *"I was very astonished when I came here [to Sweden] and heard about this [autopsy]. We think that the day the person is dead, he is not living, but he's still living [in a sense]. The dead person is crying and saying why shall I die" (Woman5)*<br>• *"It's Allah who decides if a baby shall live. Allah does not mean that all babies shall go on with their life. He can stop it. I don't want to know anything about genetics, a subsequent baby will come" (Woman6)* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |
| **Sub-theme 5: being with the child after death** | | | | | |
| 2 studies (Lundqvist 2003, Meert 2005) | 2 interviews | In 1 study conducted in the USA with parents of children who died at the PICU, parents described the need of maintaining connection with the child. Parents felt that during the last hospitalisation and during the time of death, they needed unlimited access to the child:<br>• *"You know, nobody don't want to leave their child in ICU by themselves and not know what's going on. As long as your child is there, you gonna want to be close to your child where* | Limitation of evidence | Major limitations | VERY LOW |
| | | | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or | Not saturated | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | *you can go back and forth. Cause my child never go through nothing without me being there." (Mother)*<br>• *"I don't know if this is spiritual or not, um, after she passed away one thing that helped us to say our good-byes was that we were able to hold her, you know, to hold her as long as we wanted to. We were able to rock her in our arms and feel her little body. We knew she was gone but just to have that closeness with her one more time." (Mother)*<br><br>However, in 1 study conducted in Sweden with Muslim women, they mentioned that although it is practice (in Sweden) to offer parents the possibility to see and be with the dead infant for some days after death, to help in dealing with grief, most women they did not want to be with the infant after death. Also their religion prescribes that the deceased should be buried within 24 hours.<br>• *"I have heard a woman whose baby died and was buried the same day. Women don't use to be present at the burial, and the mother was not there. After two days the mother doubted that the baby really was dead. She was desperate and asked them to take the dead baby to her. After many discussions the baby was taken from the grave and the mother saw her baby. But, the mother had to suffer so much, they said that she did not trust in God and that she was not one of us" (Woman2)*<br>• *"When one is dead, one is dead. The mother will be vulnerable. I would indeed not like this [being with the baby after death]" (Woman5)* | saturation | | |
| **Sub-theme 6: bereavement support** | | | | | |
| 1 study (Meert 2005) | 1 interview | In another study conducted in the USA with parents of children who died at the PICU, they expressed the need for contact with | Limitation of evidence | Minor limitations | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of theme or finding** | **Criteria** | **Rating** | **Overall** |
| | | the health professionals who had cared for their child:<br>• *"And we did call and wanted to talk to the doctor, just to ask them a few questions. And so later they called and right away, they accommodated us. They made time to meet us and Dr ___ talked to us. I would recommend that as time goes by and it's right for people, to be offered, to come in and just vent their feelings." (Parent)*<br>• *"And I feel there should be a support system that follows up. We had friends and we had family but we had no professional access..." (Parent)* | Coherence of findings | Coherent | |
| | | | Applicability of evidence | Unclear | |
| | | | Sufficiency or saturation | Not saturated | |

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### 8.3.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis

### 8.3.7 Evidence statements

#### 8.3.7.1 Quantitative review

No evidence was found which met the inclusion criteria for this part of the review.

#### 8.3.7.2 Qualitative review

**Attitude towards religion and spirituality**

Very low to low quality evidence from 1 qualitative study with parents of children receiving paediatric palliative care and 1 survey study conducted with parents whose children had died in the ICU looked at the attitudes towards religious and spiritual beliefs and support. Participants' responses were divided in four categories: having a formal religion, having spirituality, but without a formal religion, having no beliefs, and not wanting to discuss their beliefs. It was also raised that each person's personal views should be respected.

**Spiritual and religious needs**

Very low to low quality evidence from 3 qualitative studies with parents who had lost a child, and another qualitative study with social workers working in paediatric palliative care reflected on the importance of acknowledging spiritual and religious needs. Some aspects that were raised were the role of professionals in identifying when spiritual care might be necessary, as well as acknowledging when support is not needed; facilitating the access to religious support (such as the hospital chaplain or the chapel); and taking into account spiritual aspect when managing symptoms (such as pain).

**Aphorisms**

Low quality evidence from 1 qualitative study with parents of children receiving paediatric palliative care identified a number of aphorisms that could be categorised in the following: overall outlook, goodness, human capacity and the belief that there is a reason for everything.

**Practices and rituals**

Very low to moderate quality evidence from 7 qualitative studies with parents of children receiving palliative care, bereaved families and social workers and 1 qualitative study with hospitalised children reported on the various practices and rituals used. The most common practice mentioned by both children and parents was praying and talking to God. Parents also mentioned reading the sacred texts, using candles, listening to spiritual music and celebrating. The use of memories and legacies was also discussed. Whist most children wanted to be remembered, others preferred not to leave anything behind. Most parents found memories (such pictures or clothing) comforting, whist some mothers raised that some practices may be forbidden according to certain religious or cultural rules.

**Perceived benefits**

Very low to moderate quality evidence from 9 qualitative studies with parents of children receiving palliative care and bereaved parents and 1 qualitative study with hospitalised

children looked at the perceived benefits of spiritual and religious support and beliefs. Many parents found their religious beliefs were helpful in the decision-making process. They said that their beliefs gave them peace and comfort, helped them to cope with the situation and to make meaning of their child's illness and their loss. Their beliefs regarding an afterlife were also comforting and reassuring for parents. Some parents also reflected on the social and practical support received as a result of being part of a religious community. Children described God as protector and comforter, who helped them go through the situation or deal with painful procedures.

**Perceived difficulties**

Low to moderate quality evidence from 3 qualitative studies and 1 survey conducted with parents of children receiving palliative care and bereaved parents looked at the perceived difficulties in relation to religious beliefs. Parents discussed questioning and even rejecting their faith, and they described feeling of anger at God and the church, and some also blamed God for their child's death.

**Care after death**

Very low to low quality evidence from 3 qualitative studies with healthcare professionals, parents of children with life-threatening conditions and bereaved parents reflected on the importance of the care of the body. Continuity of care was identified as an important aspect, and this refers to treat the dead child as if he/ she was still alive. Recognising the spiritual presence of the child was also found to be important. Mothers mentioned that cultural and religious beliefs were to be respected, such as washing and wrapping of the body, burial times, and being with the child after death. The autopsy was identified as threatening by some parents, as this practise conflicted with their religious beliefs. Parent's also expressed the need for bereavement support after the child's death.

## 8.3.8 Linking evidence to recommendations

### 8.3.8.1 Relative value placed on the outcomes or themes considered

**Quantitative review**

For the quantitative part of the review, the guideline Committee decided that children and young people's quality of life, family functioning and children and young people/ family or carer's satisfaction would be critical for decision-making; whereas children and young people's well-being, children and young people's physical symptoms, children and young people's/ family or carers' coping, parents or carer's quality of life and children and young people's health service use would be important outcomes.

**Qualitative review**

For the qualitative part of the review, the Committee indicated many themes that could be important in the context of religious and spiritual support during end of life and after the child's death. These included: hope, meaning and purpose in life, taboos, religious artefacts, practices and rituals, and spiritual struggle related to the death of a child or young person.

### 8.3.8.2 Consideration of clinical benefits and harms

In spite of the limitations of the evidence, especially in terms of indirectness of the population, the Committee thought that the themes and sub-themes identified in the literature were useful and relevant.

The Committee acknowledged that the views of children and young people and parents and carers, religious, cultural or otherwise should be taken into account. The Committee noted that people who do not hold spiritual or religious beliefs may have strong spiritual, religious or cultural values that inform their thinking and values and that these beliefs and values also need to be taken into account.

The Committee agreed that for many children and parents, their beliefs can be a source of strength and comfort. These help them find meaning to their situation, and increase the sense of connection with their child. It was also acknowledged, that for some parents having a child with a life-limiting condition can generate feelings of anger or blame. Belief systems may be questioned or undermined.

Continuity of care was seen as a key theme arising from the literature. It is important to offer parents/carers and family members the possibility to be with their child after death, and to facilitate their wishes where possible.

Memory making was raised as an important consideration in the literature, and the Committee agreed that it is important to be mindful of different views and values in relation to commemorations and mementos, as some practices might not be acceptable for some (for example, photos of the child may be valued by some but unacceptable to others). In this regard, they agreed it is important to find a balance between informing parents about what is available to them, and understanding their preferences and wishes.

#### 8.3.8.3 Economic considerations

The Committee's recommendations stemming from this review question largely focus on being aware of the various sensitivities that surround this issue and therefore do not in themselves carry an opportunity cost. The discussions themselves involve some staff time but this can be considered to be a part of the overall nursing, medical and pastoral care that is routinely provided as current good practice. The implementation of the guideline recommendations are likely to have a negligible cost impact.

#### 8.3.8.4 Quality of evidence

Moderate to very low quality evidence was presented in this review. The main reasons leading to downgrading the evidence included:

The indirectness of the population. A number of studies included indirect populations, such as parents of children dying due to an acute medical condition and parents of children with a chronic condition, but not approaching the end of life. Some studies excluded people that hold no religious or spiritual beliefs. Most studies were conducted in the United States, and this limits the generalisability of the findings to the UK setting. Another study only included people with specific religious beliefs (for example Muslim women).

Biases in data collection were also reasons why the Committee had less confidence in the evidence. Many studies did not provide a detailed description of the methods used to collect the data or the analysis was poor or not clearly reported. Some of the studies reported data in a descriptive fashion only, when thematic analysis would have been more appropriate and informative. Among those studies where thematic analysis was done, the authors did not always report in detail how findings/themes were derived or emerged from the data in their research.

Another reason was the lack of the critical review of the researcher's role in sample recruitment, data collection or data analysis. Few studies clearly reported the relationship between researchers, interviewers and the respondents, whether the researchers had a pre-understanding about the topic or the possible influence of that in data collection and analytical process. Lack of verification of findings was not reported either in any of the studies.

Furthermore, the majority of the studies did not report whether saturation was achieved in terms of data collection or data analysis. It was difficult to ascertain from the information reported in those studies whether all possible views had been explored. When considering the evidence as a whole, it was not very saturated, as many themes were just raised in 1 study and there were few quotes to support them.

#### 8.3.8.5 Other considerations

Based on their experience, the Committee agreed that it is important to discuss with the child or young person, and the parents or carers their views and to re-explore them on a regular basis, as their beliefs and values may change over time. The Committee discussed the importance of recording these conversations in the Advance Care Plan. It was also raised, however, that some people may not want to discuss their beliefs or values with healthcare professionals, as these are seen as very intimate, and this should also be respected.

The Committee agreed that beyond providing information, it is important to explore the family's preferences and wishes. When discussing this with families, it is good practice to explore how their beliefs may influence care decisions, and not to make assumptions. It was highlighted that it is important not only to take into account the religious background of a person or a family, but the extent to which they adhere to their practices and norms and in which situations these may be particularly important to them.

The role of chaplains and multi-faith chaplaincy services was discussed. The Committee agreed that differences in beliefs and values might sometimes arise that are relevant to their care plan. The aim should be to try to achieve a mutually acceptable plan, if necessary, involving a person from a chaplaincy service or other facilitator. They emphasised, however, that this facilitator has to be acceptable to both the family and the healthcare professionals. It was also discussed that access to this services should be offered regardless of beliefs or circumstances.

Likewise, they agreed on the importance of people being able to access a multi-faith or quiet room to allow families space to practice their faith, reflect or meditate in hospital and hospice care settings. This was also mentioned in the evidence found in the Communication review, but the Committee agreed it also was important to stress its importance in this particular review.

Special emphasis was placed on the importance of acting in the best interest of the child. Although this is an overarching recommendation throughout the guideline, the Committee felt that it was important to mention it in the context of this review question. This is because respecting parents' cultural or religious beliefs may not always be the best interest of the child. A chaplain or another person of reference can help to mediate in this situation (for example in relation to blood transfusion or post-mortem exams). However, it was also acknowledged that in some situations, "amicable" solutions are not possible, and legal advice on intervention might sometimes be required.

The Committee felt that it was important to discuss a family's beliefs and values in the context of developing a child or young person's Advance Care Plan. The Committee heard that the term "blended faith" is sometimes used to describe a specific range of situations where family members are attempting to reconcile different faith traditions and for these families multi-faith chaplaincy services may be able to offer a supportive role which would hopefully avoid conflict. Sometimes family members who hold different beliefs and values find it hard to agree among themselves or with the child or young person and this could have an impact when attempting to make care decisions.

The Committee noted that if a child or young person with a life-limiting condition can be legally considered competent, their beliefs and values should be taken into account in relation to their care. The Committee noted that case law from the English Court suggests that a parent's right to religious freedom (Article 9 ECHR) will not be allowed to take

precedence over a child or young person's best interests. However, it should be noted that children and young people's own religious freedom is one of the many wide ranging welfare issues which should be weighed in the balance when deciding a child's best interests. (Wyatt & Another -v- Portsmouth Hospital NHS & Another [2005] EWCA Civ 1181, [2005] 1WLR 3995).

The Committee agreed that there were still important gaps in the evidence, particularly related to the generalisability of the evidence that was identified and discussed whether a research recommendation should be made. They concluded that future research should explore the attitudes of children and young peoples as well as parents or carers (in a UK NHS context) on spiritual, religious and cultural support with the aim to find better ways to address these needs.

#### 8.3.8.6 Key conclusions

The Committee concluded that healthcare professionals should take account of the child or young person's and parent or carers' spiritual, religious and cultural beliefs and values. Access to a multi-faith chaplaincy service should be offered to all families.

### 8.3.9 Recommendations

**84. In all discussions with children and young people and their parents or carers explore with them whether, based on their beliefs and values, there are any aspects of care about which they have particular views or feelings.**

**85. Ask children and young people with life-limiting conditions and their parents or carers if they want to discuss the beliefs and values (for example religious, spiritual or cultural) that are important to them, and how these should influence their care. Be aware that they may need to discuss their beliefs and values more than once.**

**86. Take account of the beliefs and values of children and young people and of their parents and carers in all discussions with them and when making decisions about their care.**

**87. Be aware that:**

- some children and young people and their parents or carers find discussions about their beliefs and values difficult or upsetting
- others find these discussions reassuring and helpful.

**88. Be aware that children and young people may feel differently to their parents, carers, or healthcare professionals about how their beliefs and values should influence their care. If there is disagreement, try to make a mutually acceptable care plan, and if necessary involve the chaplaincy service or another facilitator.**

**89. When thinking about the possibility of treatment withdrawal for a child or young person who is approaching the end of life, take into account their beliefs and values and those of their parents or carers.**

**90. Take account of the beliefs and values of children and young people and their parents or carers when thinking about funeral arrangements and the care of the child or young person's body after death.**

**91. When a child or young person is approaching the end of life, discuss with their parents or carers what would help them, for example:**

- important rituals
- recording or preserving memories (for example with photographs, hair locks or hand prints).

### 8.3.10 Research recommendations

**5. What are children's, young people's and their families' perceptions and attitudes about chaplaincy in paediatric end of life care and when would they like to access religious and spiritual support?**

| Research question | What are children's, young people's and their families' perceptions and attitudes about chaplaincy in paediatric end of life care and when would they like to access religious and spiritual support? |
|---|---|
| Why this is needed | |
| Importance to the person receiving care or the population | A 'good death' for children or young people receiving end of life care and their families means that religious and spiritual needs are identified and addressed in ways which enhance respect and dignity. There are faith specific needs around end of life and care of the body which may cause additional distress or spiritual struggle if they are not met and patient and or family choice in such matters is important to them. We need to understand how getting chaplaincy involved at the preferred time in the multidisciplinary team working with the patient and their family may be helpful for patients and families with specific religious and spiritual needs. This may help to inform the Advance Care Plan. |
| Relevance to NICE guidance | • Medium importance<br>Evidence was identified mostly from the perspective of parents. Only 1 study included children but it was classified as indirect evidence (not all children had a life-limiting condition). It would therefore be important for future updates of the guideline to assess the needs for chaplaincy particularly for children within an NHS setting. |
| Relevance to the NHS | There is consistent negative publicity about the lack of a good death for some patients. The Royal College of Nursing have expressed concern at the lack of time nurses report having available to address end of life concerns. Together for Short Lives guidelines emphasise the importance of taking note of religious values at end of life. Seeking to ensure a good death reduces the likelihood of complicated grief and the care necessary for that condition. Chaplaincy involvement may also facilitate discussion of treatment choices and organ donation where there are significant religious issues for some. |
| National priorities | Advance Care Plans in paediatric palliative care: Standards framework for children's palliative care (2011). Together for Short Lives.<br>Putting Patients First Business Plan 2013-14 2015-16 – Satisfied Patients number one priority and providing appropriate religious and spiritual support is integral to that. |
| Current evidence base | Limited mainly low-quality qualitative studies that largely focus on parents. Many studies were also indirect because they involved mixed populations of parents of children not necessarily suffering from a life-limiting condition. |
| Equality | Relevant issues are set out in the document Religion and Belief: a practical guide for the NHS (2009). |
| Feasibility | A study should include hospital and hospice contexts and a spread of ages and faiths over the paediatric population. It would be feasible to identify the benefits over a relatively short timescale and small number of institutions using questionnaires and focus groups with chaplains and other members of the team. The main ethical issue would be any potential of causing additional distress through the research if families were included. |

| Research question | What are children's, young people's and their families' perceptions and attitudes about chaplaincy in paediatric end of life care and when would they like to access religious and spiritual support? |
|---|---|
| Other comments | |

# 9 Managing distressing symptoms

## 9.1 Introduction

The recognition and management of symptoms in children and young people approaching the end of life can be difficult for even experienced paediatric palliative care practitioners due to their wide variety of clinical presentation. Despite there being paediatric symptom control manuals, specialist drug formularies and texts available, there is great variation in clinical practice. This chapter of the guideline focuses on the effectiveness of both pharmacological and non-pharmacological interventions regarding the management of pain, seizures, respiratory distress and agitation in children and young people with life-limiting conditions approaching the end of life. These symptoms are common at the end of life and can be very distressing to the child or young person as well as to their family or carers.

There are no existing tools that accurately recognise when or if a child or young person is approaching the end of life. Often, it can be subtle changes in their condition that suggest it. There are also times when children and young people improve once their symptoms are under control and it is not an uncommon occurrence for children and young people, their families/carers and involved healthcare professionals to prepare for end of life on multiple occasions. The aim of end of life care is effective symptom control and an appreciation of the importance of any improvements or deteriorations in the condition of the child or young person being communicated to the families/carers and, where appropriate, to the child or young person.

Managing difficult symptoms involves making the time to take a thorough history and perform an examination. It is important when managing symptoms to listen to both the child or young person and their families/carers to understand not only what is causing the symptoms, but also what their goals for management are. The positive and negative effects of any interventions must be considered and discussed openly with the child or young person and their families/carers. It is also important to listen to the healthcare professionals involved in the day-to-day care of the child or young person and their families/carers as they can add valuable information.

Methods of medication administration should be considered with regard to the negative effects it may cause to the child or young person and their families/carers. The route used may affect where care can be provided for the child or young person and this warrants discussion so that an informed choice can be made.

Pharmacological interventions are important in symptom control but must be incorporated into a multidisciplinary individualised management approach. It can be helpful for children and young people and their families/carers to be reassured that there are many options of management available including non-pharmacological ones, should the initial treatment of choice not completely alleviate the symptom or symptoms.

# 9.2 Managing Pain

## 9.2.1 Review question

**What pharmacological and non-pharmacological (excluding psychological) interventions are effective for the management of pain in children and young people with a life-limiting condition?**

## 9.2.2 Description of clinical evidence

The aim of this review was to assess the clinical effectiveness, the safety and the cost-effectiveness of pharmacological and non-pharmacological treatments for the management of pain in children and young people with a life-limiting condition.

The aim was to include systematic reviews of randomised controlled trials (RCTs), RCTs, cohort studies and uncontrolled studies.

Nine Cochrane reviews were identified in the search, but none of them met the inclusion criteria stated in our protocol:

- One Cochrane review (Beecham 2015) was excluded as it only identified children with cerebral palsy and *osteogenesis imperfecta*. While these children had a life-limiting condition, they were not receiving end of life care, and therefore the pain management strategies differed considerably. The management of pain in cerebral palsy will also be addressed in a specific NICE guideline (which is currently in development). Similarly, another Cochrane review (Stanton 2013) was excluded as it addressed complex regional pain syndrome.
- Seven Cochrane reviews (Bauer 2011, Bradt 2010, Fellowes 2004, McQuay 1999, Schmidt-Hansen 2015, Stevens 2015, Wiffen 2013) were excluded as the authors did not find any studies that included children. The references of the included and excluded studies were checked for potential inclusion in our review. Where the study had not been identified in our search, the titles, abstracts or full copies of the papers were retrieved for assessment.

Another systematic review was identified (Quigley 2003) that included 3 studies with children; however, these children were treated for acute pain and were not receiving end of life care and so this study was excluded.

There were 4 observational studies included in this review (Anghelescu 2005, Hunt 2001, Ruggiero 2007, Schiessl 2008). All of them used an uncontrolled study design to compare outcomes before and after the intervention was implemented. .

One study was conducted in the UK (Hunt 2001), 1 in Germany (Schiessl 2008), 1 in Italy (Ruggiero 2007) and 1 in the USA (Anghelescu 2005).

With regard to the population, all the studies included children and young people with pain due to cancer or other life-limiting conditions. One study included an indirect population, as some of the people included were up to 20 years old (Anghelescu 2005).

With regard to the intervention and comparators included, 1 study (Hunt 2001) compared the efficacy and safety of transdermal fentanyl in children who were not able to tolerate oral morphine. The other 3 studies compared different methods of administration. Two studies compared patient controlled analgesia (PCA) with the usual mode of administration (Ruggiero 2007, Schiessl 2008) and 1 study compared standard PCA with PCA by proxy (Anghelescu 2005).

Of the outcomes listed in the protocol and agreed by the Committee:

- 3 studies reported on pain (Hunt 2001, Ruggiero 2007, Schiessl 2008)
- 1 study reported on control of other symptoms (Hunt 2001)
- 1 study reported on parents or caregivers' quality of life (Hunt 2001)
- 3 studies reported on adverse events (Anghelescu 2005, Hunt 2001, Ruggiero 2007)

No results were found for children and young people's and parents or caregivers' levels of distress, and the proportion of children taken home/ readmissions to hospital or hospice.

A summary of the included studies is presented in Table 55.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix and in the GRADE profiles below and in Appendix J. Summary of included studies

### 9.2.3 Summary of included studies

**Table 55: Summary of included studies**

| Study | Intervention/ Comparison | Population | Outcomes | Comments |
|---|---|---|---|---|
| Anghelescu 2005<br>Uncontrolled study | • **Intervention: PCA by proxy.**<br>• The study did not describe the identity of the proxy (parent or nurse).<br>• PCA was administered using a CADD-Prizm® Infusion pump.<br>• The opioids used included: morphine, fentanyl and hydromorphone<br>• Comparison: Standard PCA | N=1,011 participants<br>4,972 24-hour periods<br>• PCA by proxy: n=576 24-hour periods<br>• Standard PCA: n=4,396 24-hour periods<br>Characteristics<br>• Age: up to 20 years<br>• Condition: patients with cancer, including solid tumour, brain tumour and leukaemia<br>• Every patient who had received PCA in the previous 24-hour was identified from the pharmacy records | • Adverse events<br>o Neurological complications<br>o Respiratory complications | • Before-after design<br>• Retrospective study<br>• Indirect population (the population includes up to 20 year olds) |
| Hunt 2001<br>Uncontrolled study | **Intervention**<br>**Transdermal fentanyl,** 15-day phase<br>• n=34 patch size 25 µg/h;<br>• n=5 at 50 µg/h; | N=41 children<br>n=26 completed the 15-day treatment phase, reasons for withdrawal:<br>• 7 children died | • Pain well controlled<br>• Control of other distressing symptoms<br>o Sleeping | • Prospective data collection<br>• Before-after design<br>• Potential conflict of interest |

| Study | Intervention/ Comparison | Population | Outcomes | Comments |
|---|---|---|---|---|
| | • n=1 at 75 µg/h;<br>• n=1 at 150 µg/h<br>**Comparison**<br>**Oral morphine**<br>• Median dose of oral morphine at entrance: 60 mg (range: 0 to 520)<br>• Note: Reasons for transfer to transdermal fentanyl included: difficulty with or reluctance in swallowing oral medication and occurrence of unacceptable morphine side-effects | due to disease progression<br>• 8 children were withdrawn due to inadequate response (n=5); change to parenteral opioids (n=1); adverse events (n=2)<br>Characteristics<br>• Median age: 10.5 years (range: 2.6 to 18.8)<br>• Diagnosis:<br>• haematological malignancy: n=4<br>• brain tumour: n=5<br>• other solid tumour: n=27<br>• neuro-muscular disease: n=5 | well<br>• ICYP quality of life<br>○ Convenient for the child<br>• Able to follow usual activities Parents/ carers QoL<br>○ Convenient for the parent<br>• Adverse events<br>○ Minor events (drowsy, constipation, dry mouth, nausea & vomiting, itchy skin)<br>○ Central nervous system symptoms<br>○ Serious adverse events – deaths due to treatment | • Loss to follow-up<br>• Results are extracted from a bar graph, so percentages might not be accurate |
| Ruggiero 2007<br>Uncontrolled study | **Intervention**<br>**PCA pump** (PCA VYGON freedom 5) programmed to deliver a booster dose of fentanyl when required. Fentanyl was delivered IV for at least 48 h.<br><br>**Comparison**<br>Usual care | N=18<br>Characteristics<br>• Median age: 10 years (range: 6 to 15)<br>• (median 10 years) Moderate to severe cancer pain<br>• Treated with opioids<br>• All patients had a central or peripheral IV catheter<br>• Condition:<br>○ primary bone tumour: n=10<br>○ metastatic disease: n=3<br>○ Medulloblastomas: n=3<br>○ metastatic Wilm´s tumour: n=1 | • Pain<br>• AFS score<br>• VAS score<br>• Adverse events<br>○ Minor adverse events<br>○ constipation<br>○ major adverse events | • Prospective<br>• Before-after study<br>• Small population<br>• Children with pain due to cancer only |

| Study | Intervention/ Comparison | Population | Outcomes | Comments |
|---|---|---|---|---|
| | | ○ metastatic neuroblastoma: n=1 | | |
| Schiessl 2008<br>Uncontrolled study | **Intervention**<br>**IV PCA with a strong opioid.**<br>• Morphine was the most used opioid, except in those cases where the child had a history of side effects.<br>• Median duration of treatment: 9 days (range: 1 to 50)<br>Note: Depending on the child's age, the boluses were activated by the child, the parents or the nurses. | N=8<br>Characteristics<br>median age: 8.5 years (range: 3 to 17)<br>Children who were treated with IV PCA (Graseby® 3300, Smiths medical) in the last 7 days of their life<br>Diagnosis<br>Leukaemia: n=3<br>Brain tumour: n=3<br>Solid tumour: n=2 | • Pain | • Retrospective<br>• Small sample size<br>• unclear which pain scale was used |

1

### 9.2.4 Clinical evidence

The clinical evidence profiles for this review question are presented in Table 56, Table 57, Table 58 and Table 59.

**Table 56: Summary clinical evidence profile: IV fentanyl versus oral morphine**

| Opioids: IV fentanyl compared with oral morphine for end of life care | | | | | | |
|---|---|---|---|---|---|---|
| **Outcomes** | **Illustrative comparative risks* (95% CI)** | | **Relative effect (95% CI)** | **No of Participants (studies)** | **Quality of the evidence (GRADE)** | **Comments** |
| | **Assumed risk** | **Corresponding risk** | | | | |
| | **Oral morphine** | **Opioids: IV fentanyl** | | | | |
| Pain well controlled<br>Own scale; nominal scale, categories not reported<br>Follow-up: mean 15 days | 615 per 1000 | 732 per 1000 (498 to 1000) | RR 1.19 (0.81 to 1.74) | 26 (Hunt 2001)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low1,2 | |
| Other distressing symptoms: sleeping well<br>Own scale; nominal scale, categories not reported<br>Follow-up: mean 15 days | 615 per 1000 | 652 per 1000 (431 to 991) | RR 1.06 (0.7 to 1.61) | 26 (Hunt 2001)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low1,3 | |
| Quality of life – proxy: convenient for the child<br>Own scale; range of scores not reported<br>Follow-up: mean 15 days | 538 per 1000 | 883 per 1000 (603 to 1000) | RR 1.64 (1.12 to 2.41) | 26 (Hunt 2001)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low1,2 | |
| Quality of life – proxy: convenient for the parents<br>Own scale; nominal scale, categories not reported<br>Follow-up: mean 15 days | 462 per 1000 | 498 per 1000 (286 to 882) | RR 1.08 (0.62 to 1.91) | 26 (Hunt 2001)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low1,3 | |
| Quality of life – proxy: child able to follow usual activities<br>Play Performance Scale; | 577 per 1000 | 923 per 1000 (652 to 1000) | RR 1.6 (1.13 to 2.26) | 26 (Hunt 2001)<br>Uncontrolled study | ⊕⊖⊖⊖<br>very low1 | |

| Opioids: IV fentanyl compared with oral morphine for end of life care | | | | | | |
|---|---|---|---|---|---|---|
| nominal scale, categories not reported<br>Follow-up: mean 15 days | | | | | | |
| Minor adverse: drowsiness<br>Follow-up: mean 15 days | 538 per 1000 | 463 per 1000<br>(269 to 797) | RR 0.86<br>(0.5 to 1.48) | 26<br>(Hunt 2001) | ⊕⊖⊖⊖<br>very low1,3 | |
| Minor adverse events: constipation<br>Follow-up: mean 15 days | 654 per 1000 | 536 per 1000<br>(340 to 843) | RR 0.82<br>(0.52 to 1.29) | 26<br>(Hunt 2001) | ⊕⊖⊖⊖<br>very low1,3 | |
| Minor adverse events: dry mouth<br>Follow-up: median 15 days | 577 per 1000 | 306 per 1000<br>(156 to 600) | RR 0.53<br>(0.27 to 1.04) | 26<br>(Hunt 2001) | ⊕⊖⊖⊖<br>very low1,2 | |
| Minor adverse events: nausea/ vomiting<br>Follow-up: mean 15 days | 769 per 1000 | 462 per 1000<br>(292 to 738) | RR 0.6<br>(0.38 to 0.96) | 26<br>(Hunt 2001) | ⊕⊖⊖⊖<br>very low1,2 | |
| Minor adverse events itchy skin<br>Follow-up: mean 15 days | 538 per 1000 | 269 per 1000<br>(129 to 555) | RR 0.5<br>(0.24 to 1.03) | 26<br>(Hunt 2001) | ⊕⊖⊖⊖<br>very low1,2 | |
| Adverse events: central nervous system symptoms<br>Follow-up: mean 15 days | The number of children experiencing serious adverse events before the intervention:<br>13 | Not reported | Not estimable | 26<br>(Hunt 2001) | ⊕⊖⊖⊖<br>very low1<br>(See comment) | The relative and absolute effect are not calculable.<br>Imprecision is not calculable. |
| Adverse events: serious adverse events (admissions to hospital or deaths)<br>Follow-up: mean 15 days | The number of children experiencing serious adverse events before the intervention: | The number of children experiencing serious adverse events after the intervention was:<br>0 | Not estimable | 26<br>(Hunt 2001) | ⊕⊖⊖⊖<br>very low1<br>(See comment) | The relative and absolute effect are not calculable.<br>Imprecision is not calculable. |

| Opioids: IV fentanyl compared with oral morphine for end of life care | | | | | | |
|---|---|---|---|---|---|---|
| | 0 | | | | | |

*The basis for the assumed risk (for example, the median control group risk across studies) is provided in footnotes. The corresponding risk (and its 95% confidence interval) is based on the assumed risk in the comparison group and the relative effect of the intervention (and its 95% CI).

CI: Confidence interval; RR: Risk ratio

[1] *This is an observational study and the quality of the evidence was further downgraded by 2 due to the study design high risk of selection, performance bias and detection bias*

[2] *The quality of the evidence was downgraded by 1, as the CI crosses 1 default MID*

[3] *The quality of the evidence was downgraded by 2, as the CI crosses 2 default MIDs*

**Table 57: Summary clinical evidence profile: opioids (morphine) – Patient controlled analgesia (PCA) by patient or proxy versus usual care**

| Opioids, morphine: PCA compared with usual end of life care | | | | | | |
|---|---|---|---|---|---|---|
| **Outcomes** | **Illustrative comparative risks* (95% CI)** | | **Relative effect (95% CI)** | **No of Participants (studies)** | **Quality of the evidence (GRADE)** | **Comments** |
| | **Assumed risk** | **Corresponding risk** | | | | |
| | **Usual care** | **Opioids, morphine: PCA** | | | | |
| Pain<br>Own scale; range of scores 0 to 10 (better indicated by higher values)<br>Follow-up: median 9 days | The median (range) pain before the intervention was:<br>3.7 (0 to 6) | The median range pain after the intervention was:<br>0 to 3 | Not estimable | 8<br>(Schiess 2008) | ⨁⊖⊖⊖<br>very low[1]<br>(See comment) | Median was not reported for PCA group.<br>Imprecision is not calculable. |

*The basis for the assumed risk (for example, the median control group risk across studies) is provided in footnotes. The corresponding risk (and its 95% confidence interval) is based on the assumed risk in the comparison group and the relative effect of the intervention (and its 95% CI).

CI: Confidence interval

[1] **This is an observational study and the quality of the evidence was further downgraded by 2 due to high risk of selection, performance bias, reporting bias and detection Table 58: Summary clinical evidence profile: opioids (fentanyl) – patient controlled analgesia (PCA) versus usual care**

| **Opioids, fentanyl: PCA compared with usual care for end of life care** | | | | | | |
|---|---|---|---|---|---|---|
| **Outcomes** | **Illustrative comparative risks* (95% CI)** | | **Relative effect (95% CI)** | **No of Participants (studies)** | **Quality of the evidence (GRADE)** | **Comments** |
| | **Assumed risk** | **Corresponding risk** | | | | |
| | **Usual care** | **Opioids, fentanyl: PCA** | | | | |
| Pain<br>AFS; range of scores 0 to 9 (better indicated by lower scores)<br>Follow-up: mean 48 hours | The mean pain in the control group was:<br>6.5 | The mean pain in the intervention group was:<br>4.18 | p<0.01 | 18<br>(Ruggiero 2007) | ⊕⊖⊖⊖<br>very low[1]<br>See comment | The relative and absolute effect are not calculable.<br>Imprecision is not calculable.<br>AFS: affective facial score |
| Pain<br>VAS; range of scores 0 to 90 (better indicated by lower scores)<br>Follow-up: mean 48 hours | The mean pain in the control group was:<br>68.5 | The mean pain in the control group was:<br>40 | Not p<0.01 | 18<br>(Ruggiero 2007) | ⊕⊖⊖⊖<br>very low[1]<br>See comment | The relative and absolute effect are not calculable.<br>Imprecision is not calculable.<br>VAS: Visual Analogue Scale |
| Minor adverse events (itchiness, vomiting, rashes, constipation)<br>Follow-up: mean 48 hours | Not reported | The % of children experiencing minor adverse events in the intervention group was:<br>38.9% | Not estimable | 18<br>(Ruggiero 2007) | ⊕⊖⊖⊖<br>very low[1]<br>See comment | The relative and absolute effect are not calculable.<br>Imprecision is not calculable. |
| *The basis for the assumed risk (for example, the median control group risk across studies) is provided in footnotes. The corresponding risk (and its 95% confidence interval) is based on the assumed risk in the comparison group and the relative effect of the intervention (and its 95% CI).<br><br>CI: Confidence interval; | | | | | | |

[1] *1 This is an observational study and the quality of the evidence was further downgraded by 2 due to high risk of selection, performance bias, reporting bias and detection bias*

**Table 59: Summary clinical evidence profile: Opioids – patient controlled analgesia by proxy (PCA by proxy) versus patient controlled analgesia (PCA)**

| Opioids: PCA by proxy compared with standard PCA for end of life care | | | | | | |
|---|---|---|---|---|---|---|
| **Outcomes** | **Illustrative comparative risks* (95% CI)** | | **Relative effect (95% CI)** | **No of Participants (studies)** | **Quality of the evidence (GRADE)** | **Comments** |
| | **Assumed risk** | **Corresponding risk** | | | | |
| | **Standard PCA** | **Opioids: PCA by proxy** | | | | |
| Adverse events<br>Follow-up: mean 24 hours | 7 per 1000 | 3 per 1000<br>(1 to 10) | RR 0.52<br>(0.19 to 1.42) | 4972 24-hour periods of PCA usage in 1011 children and young people (Anghelescu 2005) | ⊕⊖⊖⊖<br>very low[1,2,3] | |
| Adverse events – neurological complications<br>Follow-up: mean 24 hours | 8 per 1000 | 3 per 1000<br>(1 to 14) | RR 0.46<br>(0.11 to 1.92) | 4972 24-hour periods of PCA usage in 1011 children and young people<br>(Anghelescu 2005) | ⊕⊖⊖⊖<br>very low[1,2,3] | |
| Adverse events – respiratory complications<br>Follow-up: mean 24 hours | 6 per 1000 | 3 per 1000<br>(1 to 15) | RR 0.59<br>(0.14 to 2.47) | 4972 24-hour periods of PCA usage in 1011 children and young people<br>(Anghelescu 2005) | ⊕⊖⊖⊖<br>very low[1,2,3] | |
| *The basis for the assumed risk (for example, the median control group risk across studies) is provided in the footnotes. The corresponding risk (and its 95% confidence interval) is based on the assumed risk in the comparison group and the relative effect of the intervention (and its 95% CI).<br><br>CI: Confidence interval; RR: Risk ratio; | | | | | | |

a) *1 This is an observational study and the quality of the evidence was further downgraded by 2 due to high risk of selection, performance bias and detection bias*

b) *2 The quality of the evidence was downgraded by 1 as part of the population included in the study was over 18 years (indirect population)*

c) *3 The quality of the evidence was downgraded by 21 as the CI crosses 21 default MIDs*

### 9.2.5 Economic evidence

This review question was prioritised for economic analysis.

A systematic review did not identify any relevant economic literature relating to pharmacological and non-pharmacological interventions (excluding psychological) for the management of pain in children and young people with a life-limiting condition who are approaching the end of life.

As no clinical evidence was identified, de novo analysis was not undertaken but the cost of various pharmacological and non-pharmacological interventions are presented below.

#### 9.2.5.1 Pharmacological interventions

In addition to the drug costs there are other costs involved in the provision of pharmacological interventions, the most important of which relate to staff time, which will vary according to the route of administration. For example, in the NICE guideline on Bacterial meningitis and meningococcal septicaemia in children (CG102 – https://www.nice.org.uk/guidance/cg102/evidence/full-guideline-134564941), it was estimated that giving an intravenous drug would take 10 minutes of a Band 5/6 nurses time, which would include getting the drug and equipment to draw and make it up, checking the prescription and the patient; and delivery which takes 3–5 minutes. In addition it was estimated that cannula placement by a specialty registrar would take 5-10 minutes. The unit costs for health care professionals typically involved in the administration of intravenous drugs is given in Table 60.

**Table 60: Staff unit costs**

| Staff | Unit cost | Source |
|---|---|---|
| Band 5 nurse | £105 | PSSRU 2015 |
| Band 6 nurse | £125 | PSSRU 2015 |
| Specialty registrar | £72 | PSSRU 2015 |

*Source/Note:*

*Based on per hour of patient contact and including qualification costs*

*Based on a 40-hour week and including qualification costs*

##### *9.2.5.1.1 Paracetamol*

**Table 61: Paracetamol acquisition costs gives the acquisition costs for various formulations of paracetamol which can be used for mild to moderate pain.**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 500mg | 100 | £2.56 |
| Effervescent tablet [a] | 500mg | 100 | £8.33 |
| Soluble tablet [b] | 120mg | 16 | £0.97 |
| Orodispersible tablet [b] | 250mg | 24 | £3.59 |
| Capsule a | 500mg | 100 | £3.47 |
| • Oral suspension [a] | 120mg/5ml | 500ml | £3.14 |
| Oral solution [b] | 120mg/5ml | 500ml | £2.86 |
| Solution for infusion [b] | 1g/ml | 10 vial | £12.00 |
| Suppository [a] | 120mg | 10 | £11.26 |
| Suppository [a] | 125mg | 10 | £13.80 |
| Suppository [a] | 240mg | 10 | £22.01 |

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Suppository [a] | 250mg | 10 | £27.60 |

(a) http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320671/Part VIIIA products P (accessed 09/05/2016)
(b) BNF for children – https://www.medicinescomplete.com/mc/bnfc/current/PHP2632-paracetamol.htm?q=paracetamol&t=search&ss=text&tot=338&p=1#_hit (accessed 09/05/2016)

#### *9.2.5.1.2 Ibuprofen*

**Table 62: Ibuprofen acquisition costs illustrates the acquisition costs for ibuprofen, for use in mild to moderate pain**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 200mg | 84 | £3.19 |
| Tablet [a] | 400mg | 84 | £3.61 |
| Tablet [a] | 600mg | 84 | £4.79 |
| Modified-release tablet [a] | 800mg | 56 | £7.74 |
| Capsules [a] | 200mg | 30 | £4.40 |
| Effervescent granules [a] | 600mg | 20 sachet | £6.80 |
| Oral suspension [a] | 100mg/5ml | 500ml | £8.88 |
| Solution for infusion [b] | 10mg/2ml | 4 ampoule | £288.00 |

(a) http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320389/Part VIIIA products I (accessed 09/05/2016)
(b) BNF for children – https://www.medicinescomplete.com/mc/bnfc/current/PHP2632-paracetamol.htm?q=paracetamol&t=search&ss=text&tot=338&p=1#_hit (accessed 09/05/2016)

#### *9.2.5.1.3 Diamorphine*

Acquisition costs for diamorphine, used for moderate to severe pain, are shown in Table 63.

**Table 63: Diamorphine acquisition costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 10mg | 100 | £27.67 |
| Powder for solution for injection [a] | 5mg | 5 ampoule | £11.36 |
| Powder for solution for injection [a] | 10mg | 5 ampoule | £15.10 |
| Powder for solution for injection [a] | 30mg | 5 ampoule | £14.79 |
| Powder for solution for injection [a] | 100mg | 5 ampoule | £42.39 |
| Powder for solution for injection [a] | 500mg | 5 ampoule | £187.70 |
| Powder for solution for injection [a] | 500mg | 5 vials | £209.00 |

(a) http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320783/Part VIIIA products D

For a dose of 600mcg/kg/per day, the daily drug costs for a 20kg child would be calculated as follows:

**20kg child:**

- Administration: Subcutaneous injection
- Preparation: Diamorphine 30mg powder for solution for injection ampoules

- Cost: £14.79 ampoules (5 pack) Drug Tariff (Part VIIIA Category A)
- Cost/ampoule: £2.95
- Weight: 20kg
- Dose: 600mcg/kg = 0.6 x 20 = 12mg
- Ampoules/injection: 1
- Daily injections: 1
- Cost per day: £2.95

#### *9.2.5.1.4 Morphine sulphate*

Acquisition costs for morphine sulphate, for moderate to severe pain are given in Table 64.

**Table 64: Morphine sulphate acquisition costs**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 10mg | 56 | £5.31 |
| Modified-release tablet [a] | 10mg | 60 | £5.20 |
| Modified-release tablet [a] | 100mg | 60 | £38.50 |
| Modified-release tablet [a] | 200mg | 60 | £81.34 |
| Modified-release capsules [a] | 10mg | 60 | £3.47 |
| Modified-release capsules [a] | 200mg | 60 | £43.60 |
| Modified-release granules [b] | 20mg | 30 sachet | £24.58 |
| Suppository [a] | 30mg | 12 | £18.60 |
| Oral solution [a] | 10mg/5ml | 300ml | £5.45 |
| Solution for injection [a] | 30mg/ml | 10 ampoule | £8.84 |

*(a) NHS Drugs Tariff: http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320751/Part VIIIA products M*
*(b) BNFc: https://www.medicinescomplete.com/mc/bnfc/current/PHP2740-morphine.htm?q=morphine&t=search&ss=text&tot=92&p=2#_hit*

For a dose of 50mcg/kg/hour the daily drug costs for a 20kg child would be calculated as follows:

**20kg child:**

- Administration: Subcutaneous infusion
- Preparation: Morphine sulfate 30mg/1ml solution for injection ampoules
- Cost: £8.84 (pack of 5 ampoules) Drug Tariff (Part VIIIA Category A)
- Cost/ampoule: £1.77
- Weight: 20kg
- Dose: 50mcg/kg/hour = 0.05 x 20 x 24 = 24mg
- Cost per day: £1.77

#### *9.2.5.1.5 Oxycodone*

**Table 65: Oxycodone acquisition costs**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Modified-release tablet [a] | 5mg | 28 | £12.52 |
| Modified-release tablet [a] | 120mg | 56 | £305.02 |
| Capsules [a] | 5mg | 56 | £11.43 |
| Capsules [a] | 20mg | 56 | £45.71 |
| Oral solution [a] | 5mg/5ml | 250ml | £9.71 |

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Oral solution [a] | 10mg/ml | 120ml | £46.63 |
| Solution for injection [a] | 10mg/ml | 5 ampoules | £8.00 |
| Solution for injection [a] | 50mg/ml | 5 ampoules | £70.10 |

*(a) http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320904/Part VIIIA products O (accessed 10/05/16)*

#### *9.2.5.1.6 Fentanyl*

**Table 66: Fentanyl acquisition costs**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Buccal tablet [a] | 100mcg | 28 | £139.72 |
| Buccal tablet [a] | 800mcg | 28 | £139.72 |
| Patch [a] | 12mcg/hour | 5 | £12.59 |
| Patch [a] | 25mcg/hour | 5 | £17.99 |
| Patch [a] | 50mcg/hour | 5 | £33.66 |
| Patch [a] | 75mcg/hour | 5 | £46.99 |
| Patch [a] | 100mcg/hour | 5 | £57.86 |

*(a) http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320568/Part VIIIA products F (accessed 10/05/16)*

### 9.2.5.2 Non-pharmacological interventions

A range of non-pharmacological interventions are also available for the management of pain. A selection of frequently used non-pharmacological interventions as suggested by the Committee and their typical costs are listed in Table 67.

A range of non-pharmacological interventions are also available for the management of pain. A selection of non-pharmacological interventions and typical costs are listed in Table 67.

**Table 67: Costs of non-pharmacological interventions**

| Intervention | Cost |
|---|---|
| • Music therapy [a,b] | £40 per hour |
| • Massage [c] | £20-£60 per hour |
| • Acupuncture [d] | £127 |
| • Physiotherapy [e] | £46 |
| • Reflexology [f] | £25-£50 per hour |

*(a) http://www.cancerresearchuk.org/about-cancer/cancers-in-general/treatment/complementary-alternative/therapies/music-therapy accessed (10/05/2016)*
*(b) G group session are cheaper (£21 per person) http://www.richmondmusictrust.org.uk/musictherapy accessed (10/05/2016)*
*(c) http://www.nhs.uk/ipgmedia/National/Penny%20Brohn%20Cancer%20Care/assets/Meditationandmindfulness(PBCC).pdf accessed (10/05/2016)*
*(d) NHS Reference Costs (2014/15) – Acupuncture for pain management; Currency Code AB23Z Service Description: Pain management*
*(e) NHS Reference Costs (2014-15) – Service code: 150*
*(f) http://www.nhs.uk/ipgmedia/National/Penny%20Brohn%20Cancer%20Care/assets/Reflexology(PBCC).pdf*

## 9.2.6 Evidence statements

### 9.2.6.1 Pharmacological interventions

#### Non-opioids

No evidence was found.

#### Opioids

***Within-class comparison***

Very low quality evidence from 1 uncontrolled study with 26 children with pain associated with cancer and other life-limiting conditions, showed no clinically significant improvement in the reported pain (as measured with study own scale) when children were transferred to transdermal fentanyl compared with previous treatment with oral morphine at 15 days follow-up. There was uncertainty around this estimate effect.

Very low quality evidence from 1 uncontrolled study with 26 children with pain associated with cancer and other life-limiting conditions, showed no clinically significant improvement in the reported quality of sleep (as measured with study own scale) when children were transferred to transdermal fentanyl compared with previous treatment with oral morphine at 15 days follow-up. There was considerable uncertainty around this estimate effect.

Very low quality evidence from 1 uncontrolled study with 26 children with pain associated with cancer and other life-limiting conditions, showed that a clinically significant higher number of children found transdermal fentanyl more convenient than oral morphine at 15 days follow-up. There was uncertainty around that estimate effect. However, no differences were reported by the parents in this regard. There was considerable uncertainty around this estimate effect.

Very low quality evidence from 1 uncontrolled study with 26 children with pain associated with cancer and other life-limiting condition, showed a clinically significant improvement in the quality of life (measured as the children being more able to follow usual activities) when they were treated with transdermal Fentanyl than when they were treated with oral morphine at 15 days follow-up. There was uncertainty around this estimate effect.

Very low quality evidence from 1 uncontrolled study with 26 children with pain associated with cancer and other life-limiting condition, showed that there were no clinically significant differences in the reported minor adverse events (including drowsiness, constipation, dry mouth, nausea/ vomiting and itchy skin) between transdermal fentanyl and oral morphine at 15 days follow-up. There was considerable uncertainty around these estimate effects.

Very low quality evidence from 1 uncontrolled study with 26 children with pain associated with cancer and other life-limiting conditions reported that 50% of the children reported central nervous system symptoms when they received transdermal fentanyl at 15 days follow-up. The clinical significance of this outcome could not be calculated with the data reported.

Very low quality evidence from 1 uncontrolled study with 26 children with pain associated with cancer and other life-limiting conditions showed that there were no differences in the number of serious adverse events (including admissions to hospitals and deaths) between transdermal fentanyl and oral morphine at 15 days follow-up. The clinical significance of this outcome could not be calculated with the data reported.

***Delivery system: Patient controlled analgesia (PCA)***

Very low quality evidence from 1 uncontrolled with 8 children with pain due to cancer showed that the reported pain (as measured with study own scale) was milder when the children were receiving a strong opioid using PCA (activated by the child, the parents or the nurses) at 9 days follow-up. The clinical significance of this outcome could not be calculated with the data reported.

Very low quality evidence from 1 uncontrolled study with 18 children with pain due to cancer showed that the reported pain as assessed by a validated scale (as measured with the Affective Facial Score or Visual Analogue Scale) was lower when the children were receiving fentanyl using PCA at 48 hours follow-up. The clinical significance of this outcome could not be calculated with the data reported.

Very low quality evidence from 1 uncontrolled study with 18 children with due to cancer showed that 38.9% of the children reported minor adverse events (including itchiness, vomiting, rashes and constipation) when they received fentanyl using PCA at 48 hours follow-up. The clinical significance of this outcome could not be calculated with the data reported.

Very low quality evidence from 1 uncontrolled study with 1,011 children and young people with cancer evaluating 4972 24-hour periods of PCA showed a clinically significant lower occurrence of adverse events and neurological and respiratory complications when the children were receiving standard PCA than when they were receiving PCA by proxy at 24 hours follow-up. There was considerable uncertainty around these estimate effects.

**Local anaesthetics**

No evidence was found

**Adjuvants**

No evidence was found

**Palliative chemotherapy**

No evidence was found

**Palliative radiotherapy**

No evidence was found

**Steroids**

No evidence was found

**Medical formulations of cannabis**

No evidence was found

**Chronic pain rehabilitation strategies**

No evidence was found for chronic rehabilitation strategies, that includes both pharmacological and non-pharmacological interventions.

#### 9.2.6.2 Non-pharmacological interventions

No evidence was found for any of the non-pharmacological interventions listed in the protocol.

### 9.2.7 Linking evidence to recommendations

#### 9.2.7.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were levels of pain, adverse effects from pharmacological treatments and child or young person and their parents or carers' quality of life; whereas control of other distressing symptoms, children or young person and their parents or carers' levels of distress and proportion of children re-admitted to hospital/ hospice were considered as important outcomes.

#### 9.2.7.2 Consideration of clinical benefits and harms

Whereas symptom management of respiratory distress, agitation and seizures were considered for children and young people who approach the end of life, pain was considered over a longer timeframe. The following types of pain were considered: nociceptive pain, bone pain (due to cancer), headache (related to raised intracranial pressure), neuropathic pain, and visceral pain (for example, bowel or bladder). The literature regarding the management of pain in children and young people with a life-limiting condition was scarce. The Committee acknowledged that carrying out research in this population is difficult. Most published papers are case series or case reports, and they therefore did not meet the criteria for inclusion in this review. In providing effective pain management, a clear assessment of the type or mechanism of pain as well as possible contributing factors is critical to find an approach that alleviates symptoms while minimises side effects (such as unwanted levels of sedation).

Only 1 study that compared 2 different opioids (oral morphine versus transdermal fentanyl) was identified. However, the Committee agreed that the evidence was too limited to directly base recommendations on. The Committee agreed that non-pharmacological management of pain should be considered whenever applicable because there are numerous adverse events associated with pharmacological pain management. Opioids can cause nausea and vomiting as well as high level of sedation which could be distressing to the child or young person and their family.

Given the lack of evidence, the recommendations were based mostly on the Committee's discussion.

#### 9.2.7.3 Economic considerations

It was difficult for the Guideline Committee to make specific recommendations on pain management derived from economic considerations as the population within this guideline is diverse and pain management is often very individualised. Nevertheless, the recommendations generally encourage a stepwise approach to pharmacological pain management which will tend to lead to cheaper drugs, doses and administration route being used unless adequate pain relief is not achieved. The Committee noted that there would be occasions when a more expensive drug or formulation could be optimal in order to provide more flexible dosing and/or safer administration.

Non-pharmacological treatments vary in cost and can sometimes be provided more cheaply in a group setting. The strength of the recommendations with respect to non-pharmacological interventions reflects the lack of evidence on effectiveness and cost-effectiveness for these interventions in this population but the Committee nevertheless thought they offered valuable benefits as part of an overall pain management strategy.

#### 9.2.7.4 Quality of evidence

The quality of each study was assessed using the NICE manual methodology checklists and is reported in the study evidence tables and the quality of the evidence for an outcome (that is, across studies) was assessed using GRADE.

The overall evidence was of very low quality. This was due to the methodological flaws inherent to uncontrolled studies and the fact that data was collected retrospectively in several studies. In addition, further concerns were raised about population indirectness, as all studies included children and young people with a life-expectancy beyond 2 months.

The recommendations were therefore mainly based on consensus within the Committee rather than on the available evidence.

#### 9.2.7.5 Other considerations

The Committee concluded that due to the limited evidence found, recommendations were mainly based on Committee members' clinical experience, and consensus on good clinical practice.

In their discussion, the Committee agreed that in order to effectively manage pain in children and young people living with a life-limiting condition, the main steps should be assessment, development of a management plan, considering non-pharmacological and pharmacological interventions accordingly and reassessment.

With regard to assessment of pain, the Committee emphasised that the children and young people's pain could have single or multiple causes as well as contributing factors (for example, emotional, environmental or physical), and therefore the assessment should take all of these into consideration. They also highlighted the importance of listening to the views of the child or young person, the parents or carers and of other healthcare professionals involved in the child or young person's care. In relation to this, it is important to establish the most effective mode of communication appropriate for the age and developmental stage of the patient.

The Committee agreed on the importance of taking a pain history, and also on the need to repeat the assessment regularly as pain can recur or worsen unexpectedly. It is important if possible to determine the underlying causes of pain as well as possible triggers. Examples of reversible causes of pain may include musculoskeletal problems, or constipation. Common factors related to the causes and the severity of pain that were identified by the Committee included anxiety, the comfort of the environment and social, emotional and religious or spiritual considerations.

The Committee agreed that it is important to evaluate the severity of pain. The cause or causes of the pain are important because this has implications for the drug choice.

The Committee agreed that the identification of an underlying cause or contributing factor could be critical in planning the optimal management of pain.

With regard to the management of pain, the Committee agreed that an overarching recommendation was needed to ensure a comprehensive approach that incorporates both non-pharmacological and pharmacological interventions. They agreed that it is important to discuss with the child or young person and the parents or carers the benefits and harms, of any potential intervention. While reducing suffering is paramount, the control of pain may sometimes lead to adverse events, such as unwanted sedation or constipation with opioid analgesia.

For pharmacological treatment, the Committee was aware of the existence of World Health Organisation (WHO 2012) guidance on pain management (not specifically related to palliative care), and given the absence of evidence, they agreed on adopting/ adapting some

of the principles from that document, as they are consistent with their current clinical practice. Minor changes were made to emphasise the role of breakthrough analgesia, and in relation to the dosages.

The Committee adopted and adapted the 3 key WHO principles: by the clock, by the mouth and by the individual, based on their experience. Drugs should be given at regular intervals, whether there is pain or not, and special emphasis was placed on the use of additional doses, if necessary, for breakthrough pain. The idea is to prevent pain from occurring and to treat any breakthrough pain rapidly. The least invasive and non-painful route of administration should be used, favouring the use of oral drugs. It is important to avoid injections wherever possible, as they are painful, and some children may underreport pain to avoid them. The Committee suggested considering the transmucosal route.

There was consensus in recommending the WHO 2-step approach. Simple analgesia, such as paracetamol or ibuprofen, should be considered for mild pain (step 1). For moderate to severe pain, or pain that does not respond to simple analgesia (step 1), an opioid should be considered. There was also agreement that morphine should be the first choice treatment, as indicated by the WHO guidelines.

Treatment should be initiated with the lowest recommended dose and then be titrated to the individual's needs. The Committee also discussed the need for the management of pain at predictable times. They discussed the issue of possible overdosing and decided to add a statement in the recommendations highlighting to clinicians not to include anticipatory doses when calculating the required daily background dose of analgesia. The Committee agreed that this statement is meant to prevent professionals from increasing the background dose for the next 24 hours. Having this statement, they agreed, would essentially protects against overdosing. The Committee pointed out that it is important to take into account that the drug dosages should be based on the child's weight, rather than their age, as a significant proportion of children are underweight in this population (up to 30%).

The role of patient-controlled analgesia was discussed. The Committee decided not to make a particular recommendation with regard to this. It was recognised that it is important to give the children and the parents' adequate control over the use of analgesia, but to leave it to the relevant healthcare professional to decide how this is best implemented. The Committee agreed that non-pharmacological interventions are also important. Some simple non-pharmacological approaches could be useful, such as the use of heat and cold pads or distraction / calming techniques.

They also discussed a number of interventions that were included in the review protocol and are in use managing pain, such as acupuncture, massage therapy, music therapy, physiotherapy, TENS and play therapy. However, due to the lack of evidence found, the Committee did not make recommendations on these interventions. However, based on the Committee's consensus and expertise it was decided that measures that increase relaxation could also lead to pain reduction. They therefore agreed that environmental changes (for example reduction in noise) and other methods to promote relaxation such as playing music, touch, holding and massage, should be considered.

The Committee also discussed the need for research recommendations, given the lack of evidence in this area. They acknowledged that conducting research in this population is quite challenging but agreed that this topic was so important to improve the wellbeing of the child and reduce the distress of their parents or carers that a research recommendation should be made.

#### 9.2.7.6 Key conclusions

The Committee recognised that pain management is a core component in the care of some children and young people with a life-limiting condition. Pain assessment should be an ongoing process, as the pain can develop unexpectedly and can vary in severity. In

assessing pain, it is important to look at the intensity and quality of pain, the potential underlying causes, as well as triggers that may cause, contribute or exacerbate it.

When treating pain, a comprehensive approach that incorporates both non-pharmacological and pharmacological interventions is needed. The main objective is to prevent pain from occurring and to treat pain rapidly, and therefore the use of regular analgesia with additional doses for breakthrough pain is recommended. The Committee agreed with the core principles of the WHO guidance that simple analgesia is recommended for mild pain, and opioids are recommended for moderate to severe pain. Treatment should be titrated to the individual's needs, and the use of oral, transmucosal or transdermal formulations should be favoured where possible, as they are less painful.

### 9.2.8 Recommendations

**92. When assessing and managing pain, be aware that various factors can contribute to it, including:**

- causative factors, for example musculoskeletal disorders or constipation
- environmental factors, such as an uncomfortable or noisy care setting
- psychological factors, such as anxiety and depression
- social, emotional, religious, spiritual or cultural considerations.

**93. When assessing pain in children and young people:**

- use an age-appropriate approach that takes account of their stage of development and ability to communicate
- try to identify what is causing or contributing to their pain, and be aware that this may not relate to the life-limiting condition
- take into account the following causes of pain and distress that might have been overlooked, particularly in children and young people who cannot communicate:
  - neuropathic pain (which can be associated with cancer)
  - gastrointestinal pain (which can be associated with diarrhoea or constipation)
  - bladder pain (which can be caused by urinary retention)
  - bone pain (which can be associated with metabolic diseases)
  - pressure ulcers
  - headache (which can be caused by raised intracranial pressure)
  - musculoskeletal pain (particularly if they have neurological disabilities)
  - dental pain.

**94. Be aware that pain, discomfort and distress may be caused by a combination of factors, which will need an individualised management approach.**

**95. For children and young people who have pain or have had it before, regularly reassess for its presence and severity even if they are not having treatment for it.**

**96. Think about non-pharmacological interventions for pain management, such as:**

- Changes that may help them to relax, for example:
  - environmental adjustments (reducing noise)
  - music

- physical contact such as touch, holding or massage
- local hot or cold applications to the site of pain
- comfort measures, such as sucrose for neonates.

**97. When tailoring pain treatment for an individual child or young person, take into account their views and those of their parents or carers on:**

- the benefits of pain treatment
- the following possible side effects of analgesia for moderate to severe pain (such as opioids):
  - unwanted sedation
  - reduced mobility
  - constipation.

**98. Consider using a stepwise approach to analgesia in children and young people, based on pain severity and persistence:**

- For mild pain, consider paracetamol or ibuprofen sequentially, and then in combination if needed.
- For moderate to severe pain, consider one of the following options:
  - paracetamol or ibuprofen sequentially, and then in combination if needed **or**
  - low-dose oral opioids (such as morphine), **or**
  - transmucosal opioids **or**
  - subcutaneous opioids **or**
  - intravenously infused opioids (if a central venous catheter is in place).

**99. If treatment with a specific opioid does not give adequate pain relief or if it causes unacceptable side effects, think about trying an alternative opioid preparation.**

**100. When using opioids, titrate treatment to find the minimal effective dose that will relieve and prevent pain.**

**101. Titrate treatment to provide continuous background analgesia, and prescribe additional doses for breakthrough pain if this occurs.**

**102. In addition to background analgesia, consider giving anticipatory doses of analgesia for children and young people who have pain at predictable times (for example when changing dressings, or when moving and handling). Do not include anticipatory doses when calculating the required daily background dose of analgesia.**

**103. Calculate opioid dosages for children and young people who are approaching the end of life using weight rather than age, because they may be underweight for their age.**

**104. If you suspect neuropathic pain and standard analgesia is not helping, consider a trial with other medicines, such as:**

- gabapentin **or**
- a low-dose tricyclic antidepressant (for example amitriptyline ) **or**

- an anti-NMDA agent (for example ketamine or methadone), used under guidance from a specialist.

### 9.2.9 Research recommendations

**6. What is the acceptability, safety, and effectiveness of different types of opioid analgesia for breakthrough pain in children and young people with life-limiting conditions who are having end of life care in the community?**

| Research question | What is the acceptability, safety, and effectiveness of different types of opioid analgesia for breakthrough pain in children and young people with life-limiting conditions who are having end of life care in the community? |
|---|---|
| Why this is needed | |
| Importance to 'patients' or the population | Children and their families may receive end of life care in a variety of settings. They consistently report that good symptom management, particularly with respect to 'being free from pain', may influence their choice of setting. Being able to offer robust, rapid onset, needle-free analgesia for breakthrough pain, in community settings would enable children and their families to feel confident in choosing their place of care without as much anxiety about uncontrolled pain. |
| Relevance to NICE guidance | • High priority:<br>No studies were identified that directly examined the safety or effectiveness of different management strategies for treating breakthrough pain in children receiving end of life care. Future NICE guidance would greatly benefit from the identification of appropriate strategies to administer opioid analgesia in a flexible way to children being cared for outside of an acute hospital setting. |
| Relevance to the NHS | While any medication provided will carry a finite cost, this would need to be offset against the cost of current treatments. There is likely to be a cost saving to the NHS if more children are empowered to receive end of life care outside of acute paediatric hospital beds, |
| National priorities | The Medicines for Children Research Network (NIHR) Pain & Palliative Care Clinical Studies Group have conducted a research priorities setting exercise. The outcome included a number of topics related to breakthrough analgesia for children. |
| Current evidence base | There is currently no robust evidence about which breakthrough analgesia strategies are patient acceptable, safe and effective for children receiving out of hospital end of life care. Many preparations are licensed for use in adults or older children only. |
| Equality | Children in need of end of life care are relative therapeutic orphans. While the numbers of children involved are relatively small, they have an equal right to safe analgesic medication. |
| Feasibility | There are always ethical issues in conducting studies in vulnerable populations, and there are additional considerations relating to pain relief interventions. These would require careful consideration, but could be overcome. The numbers of children affected are also (fortunately) small, however a well conducted multicentre study would be likely to be adequately powered. |
| Other comments | It has traditionally been difficult to get funding for studies looking at existing drugs used in particular populations within paediatric palliative care. There is no financial incentive for drug companies, and larger funding bodies have not always considered research in small groups of patients to be a high priority. |

# 9.3 Managing agitation

## 9.3.1 Review question

**What pharmacological and non-pharmacological interventions (excluding psychological) are effective for the management of agitation in children and young people with a life-limiting condition who are approaching the end of life?**

## 9.3.2 Description of clinical evidence

The aim of this review was to assess the clinical effectiveness, the safety and the cost-effectiveness of pharmacological and non-pharmacological treatments for the management of agitation in children and young people with a life-limiting condition who are approaching the end of life.

Systematic reviews of randomised controlled trials (RCTs), RCTs, cohort studies and uncontrolled studies were looked for, but no relevant studies were identified in the search.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

## 9.3.3 Summary of included studies

No evidence was found which met the inclusion criteria for this review.

## 9.3.4 Clinical evidence

No evidence was found which met the inclusion criteria for this review.

## 9.3.5 Economic evidence

This review question was prioritised for economic analysis.

A systematic review did not identify any relevant economic literature relating to pharmacological and non-pharmacological interventions (excluding psychological) for the management of agitation in children and young people with a life-limiting condition who are approaching the end of life.

As no clinical evidence was identified, de novo analysis was not undertaken but costings of various treatment alternatives are presented below.

### 9.3.5.1 Pharmacological interventions

In addition to the drug costs there are other costs involved in the provision of pharmacological interventions, the most important of which relate to staff time, which will vary according to the route of administration. For example, in the NICE guideline on Bacterial meningitis and meningococcal septicaemia in children (CG102), it was estimated that giving an intravenous drug would take 10 minutes of a Band 5/6 nurses time, which would include getting the drug and equipment to draw and make it up, checking the prescription and the patient; and delivery which takes 3–5 minutes. In addition it was estimated that cannula placement by a specialty registrar would take 5–10 minutes.

The Guideline Committee also reported that drugs are often double checked in paediatric palliative care due to the small doses and/or local policy. Table 68 shows the unit costs of health care professionals typically involved in the administration of intravenous drugs.

**Table 68: Staff unit costs**

| Staff | Unit Cost | Source |
|---|---|---|
| Band 5 nurse | £105 | PSSRU 2015 |
| Band 6 nurse | £125 | PSSRU 2015 |
| Specialty registrar | £72 | PSSRU 2015 |

*Based on per hour of patient contact and including qualification costs*
*Based on a 40-hour week and including qualification costs*

#### *9.3.5.1.1 Midazolam*

**Table 69: Midazolam acquisition costs gives acquisition for midazolam solution for injection**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Solution for injection [a] | 10mg/5ml | 10 ampoules | £6.38 |
| Solution for injection [a] | 10mg/2ml | 10 ampoules | £6.25 |
| Solution for injection [a] | 50mg/10ml | 10 ampoules | £25.00 |
| Oromucosal solution [b] | 10mg/2ml | 4 unit dose | £91.50 |
| Oromucosal solution [b] | 2.5mg/0.5ml | 4 unit dose | £82.00 |
| Oromucosal solution [b] | 5mg/ml | 4 unit dose | £85.50 |
| Oromucosal solution [b] | 7.5mg/1.5ml | 4 unit dose | £89.00 |

*(a) BNFc NHS indicative price https://www.medicinescomplete.com/mc/bnfc/current/PHP3037-midazolam.htm?q=midazolam&t=search&ss=text&tot=56&p=1#PHP77320-solution-for-injection (accessed 12/05/2016)*
*(b) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320751/Part VIIIA products M*

For a suggested a dose of 0.7mg/kg/day, the daily cost of treatment for a child of 20kg would be calculated as follows:

| | |
|---|---|
| Administration: | Injection |
| Preparation: | Midazolam 10mg/2ml solution for injection ampoules |
| Cost: | £6.25 ampoules (pack of 10) NHS indicative price |
| Cost/ampoule: | £0.63 |
| Weight: | 20kg |
| Dose: | 0.7mg/kg/day = 0.7 x 20 = 14mg |
| Ampoules/day: | 2 |
| Cost per day: | 2 x £0.63 = £1.26 |

#### *9.3.5.1.2 Levomepromazine*

Acquisition costs for levomepromazine are illustrated in Table 70.

**Table 70: Levomepromazine acquisition costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Solution for injection [a] | 25mg/ml | 10 ampoules | £20.13 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320722/Part VIIIA products L (accessed 12/05/2016)*

The dose is variable but using an example of 3mg/kg over 24 hours, the daily cost of treatment for a child of 20 kg would be calculated as shown below:

| | |
|---|---|
| Administration: | Injection |
| Preparation: | Levomepromazine 25mg/1ml solution for injection ampoules |
| Cost: | £20.13 ampoules (pack of 10) Drug Tariff (Part VIIIA Category C) |
| Cost/ampoule: | £2.01 |
| Weight: | 20kg |
| Dose: | 3mg/kg/day = 3 x 20 = 60mg |
| Ampoules/day: | 3 |
| Cost per day: | 3 x £2.01 = £6.03 |

#### *9.3.5.1.3 Haloperidol*

The acquisition costs for haloperidol are given in Table 71

**Table 71: Haloperidol acquisition costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet a | 1.5mg | 28 | £2.35 |
| Tablet a | 5mg | 28 | £3.39 |
| Tablet a | 10mg | 28 | £12.85 |
| Tablet a | 20mg | 28 | £21.97 |
| Capsules a | 500mcg | 30 | £1.18 |
| Oral solution a | 5mg/5ml | 100ml | £6.44 |

*(a) NHS Drugs Tariff*
*(b) http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320587/Part VIIIA products H (accessed 12/05/2016)*

#### *9.3.5.1.4 Diazepam*

Table 72 shows the acquisition costs for rectal diazepam and diazepam solution for injection.

**Table 72: Diazepam injection costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Solution for injection a | 10mg/2ml | 10 | £5.50 |
| Rectal solution tube a | 5mg/2.5ml | 5 | £5.85 |

*(a) NHS Drugs Tariff*
*(b) http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320783/Part VIIIA products D (accessed 12/05/2016)*

Based on a dose of 400mcg/kg the daily cost of treatment for a child of 20 kg would be calculated as shown below:

| | |
|---|---|
| Administration: | Injection |
| Preparation: | Diazepam 10mg/2ml solution for injection ampoules |
| Cost: | £5.50 ampoules (pack of 10) Drug Tariff (Part VIIIA Category C) |
| Cost/ampoule: | £0.55 |
| Weight: | 20kg |

Dose: 400mcg/kg = 0.4 x 20 = 8mg

Ampoules/day: 1

Cost per day: 1 x £0.55 = £0.55

#### 9.3.5.2 Non-pharmacological interventions

Several non-pharmacological interventions were included in the review protocol and approximate costs for some of these interventions are given in Table 73. However, other non-pharmacological interventions such as a soothing voice have a negligible and difficult to quantify cost but would usually be provided as part of on-going nursing care.

**Table 73: Costs of non-pharmacological interventions for**

| Intervention | Cost |
|---|---|
| Music therapy [a,b] | £40 per hour |
| Massage [c] | £20-£60 per hour |
| Play [d] | £50 per session |

*(a) http://www.cancerresearchuk.org/about-cancer/cancers-in-general/treatment/complementary-alternative/therapies/music-therapy accessed (10/05/2016)*
*(b) G group session are cheaper (£21 per person) http://www.richmondmusictrust.org.uk/musictherapy accessed (10/05/2016)*
*(c) http://www.pennybrohn.org.uk/wp-content/uploads/2014/08/Massage-EBISv4.1_without-refs.pdf (accessed 12/05/2016)*
*(d) http://www.playtherapybase.co.uk/?page_id=81 (accessed 12/05/2016) Group sessions for 3-6 children are £70 per session*

### 9.3.6 Evidence statements

No studies were included in the review.

### 9.3.7 Linking evidence to recommendations

#### 9.3.7.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were reduction of agitation, child or young person's levels of distress and their quality of life; whereas parents or carers' levels of distress and quality of life, child or young person and their parents or carers satisfaction and adverse events were rated as important outcomes. No evidence was identified.

#### 9.3.7.2 Consideration of clinical benefits and harms

The Committee considered it important to highlight that symptoms such as agitation can be common when the child or young person is approaching the end of life. However, the causes of these symptoms can vary widely and the possible benefits of treatments should be weighed against the side effects of some of the drugs that are considered in the protocol which could, for instance cause unwanted levels of sedation. The distress and burden caused by symptoms of agitation at the end of life can not only affect the child or young person, but also have a detrimental effect on family members and caregivers. Good communication about possible causes and treatments are an essential component of symptom management. Due to the possible adverse effects from the pharmacological management of agitation (for instance nausea and vomiting or over-sedation amongst others), the Committee considered it important that non-pharmacological treatment options are considered as a first line option.

#### 9.3.7.3 Economic considerations

Although agitation can be a common symptom in children and young people approaching the end of life, the overall population is small. The review did not identify any clinical evidence and treatment is highly specific to the circumstances of the individual which meant that recommendations were not explicitly derived from considerations of cost-effectiveness.

However, non-pharmacological interventions are recommended as first-line and most of these have negligible cost – calm speaking, reassurance and changes to the environment to make it more comfortable, for example. Where medicine is used for agitation the drugs given are cheap. Therefore, the Committee thought that the recommendations made would not have a large cost impact and that they represented a cost-effective use of NHS resources.

#### 9.3.7.4 Quality of evidence

Not applicable.

#### 9.3.7.5 Other considerations

The Committee concluded that due to the lack of evidence, recommendations were mainly based on Committee members' clinical experience, expert opinion and existing guidance.

In their discussion the Committee members agreed that in order to effectively manage agitation in children or young people living with life-limiting conditions who are approaching the end of life, the main steps should be identification of agitation, assessment of underlying causes, treatment of any reversible causes and consideration of non-pharmacological and pharmacological interventions accordingly.

For the identification and establishment of terminal agitation, the Committee noted that terminal agitation could occur in some children or young people before the end of life, and manifest as restlessness, irritability, aggressive behaviour, or as well as being distressed for example inconsolable crying. Establishing the possible causes of agitation will help guide the treatment plan. The manifestation could be both internal and external, and some children and young people may appear to be aggressive in behaviour and distressed as well. The Committee also noted that sometimes delirium could be confused with terminal agitation, however agitation is only a part of delirium. A child or young person with delirium could show signs of confused thinking, disrupted attention, disordered speech and hallucinations in addition to agitation. The Committee thought it important to stress this to healthcare professionals providing care.

The Committee noted that children and young people with neurodisability may present in different ways with manifestation such as with seizures or dystonia. They recommended that healthcare professionals should be aware of this and not to confuse them with terminal agitation when providing care to this group of children and young people. It is important to understand what is normal for each individual patient.

With regard to assessing or determining the cause or causes of agitation, the Committee noted that it was important to look for any untreated symptoms such as pain or urinary retention which may be causing agitation in children and young people. These untreated symptoms should be considered by healthcare professionals when providing care and treatment for agitation. As for the other underlying causes of agitation, there were a variety of them including hypoxia, anaemia, dehydration, constipation, fear, anxiety or depression. These can be grouped into 3 categories, namely, medical disorders, psychological factors and adverse effects from medication. The decision as to whether any underlying causes are treated should be assessed and the risks versus benefits of treatment considered.

With regard to treatment, the Committee agreed that before treating presumed primary agitation, healthcare professionals should identify and treat any potential underlying causes for agitation first.

For the treatment of agitation, the Committee agreed that non-pharmacological management should be the first-line approach, including providing environmental and/or psychological support to the children and young people and their family/carers. They noted that providing support to parents or carers is important alongside the management of the child or young person. However parents' and carers' distress should not be confused with the children and young people's distress. In order to provide support to family/carers and the children and young people properly, their spiritual and cultural needs and expectations also needed to be considered.

The Committee also discussed the physical restraint of the child and young person in danger of self-harm due to excessive agitation. They thought it is important to keep the child and young person safe and provide them with comfort. Due to the issue of personal liberty that is involved and other possible impacts that physical restraint may have on the child/young person and their family/carers, this should be approached with caution, in full communication with family/carers and always in the best interest of the child and young person.

For pharmacological treatment, the Committee discussed and recommended 2 classes of drugs for the treatment of agitation: neuroleptics, such as haloperidol or levomepromazine, and benzodiazepines, such as midazolam, diazepam or lorazepam. They did not recommend specific dosages because these vary between age groups, but did recommend that treatment should start with the lowest clinically effective dose and be titrated until optimum symptom relief is achieved for each individual child. Special emphasis was made on the issue of sedation, and the Committee agreed that the primary treatment goal should be managing agitation and avoiding sedation wherever possible so that sedation is not the primary aim of treatment).

Finally the Committee discussed whether a research recommendation should be drafted for this topic. They concluded that research would be very difficult to conduct, because of the variety of possible causes of agitation in the last days of life.

#### 9.3.7.6 Key conclusions

The Committee concluded that when treating agitation in children and young people approaching the end of life, it is important to be aware that agitation may manifest in different ways and the underlying causes for agitation should be assessed. The identified underlying causes should be addressed and treated if appropriate and investigations should be undertaken to assess their effectiveness. When treating agitation, non-pharmacological management should be considered as the first-line approach. When needed, pharmacological interventions such as neuroleptics and benzodiazepines could be considered and treatment should start from the lowest recommended dose and be titrated according to response. It may also be necessary to ensure the children and young person's safety in states of excessive agitation. Healthcare professionals should be aware of the risk of unnecessary over-sedation when managing agitation.

### 9.3.8 Recommendations

**105. Be aware that as children and young people with life-limiting conditions approach the end of life they may:**

- become agitated, shown by restlessness, irritability, aggressive behaviour, crying or other distress
- show signs of delirium, such as confusion, disrupted attention, disordered speech, hallucinations and agitation.

**106. If a child or young person who is approaching the end of life becomes agitated or delirious, make sure that they are safe from physical injury.**

**107. If a child or young person becomes agitated as they are approaching the end of life, look for causes and factors that may be contributing to this, including:**

- medical disorders and conditions such as pain, hypoxia, anaemia, dehydration, urinary retention or constipation
- psychological factors such as fear, anxiety or depression
- adverse effects from medication.

**108. For children and young people with a neurological disability who are approaching the end of life, be aware that the symptoms and signs of agitation or delirium can be mistaken for the signs and symptoms of seizures or dystonia.**

**109. If a child or young person who is approaching the end of life needs treatment for agitation:**

- identify and if possible treat any medical or psychological conditions that may be contributing to it
- think about non-pharmacological interventions, such as:
  - calm speaking, reassurance, distraction, and physical contact such as holding and touch
  - changes to the environment to make it more comfortable, calm and reassuring, to reduce noise and lighting, to maintain a comfortable room temperature, and to provide familiar objects and people and relaxing music
  - religious and spiritual support if this is wanted and helpful
- think about pharmacological interventions (beginning with low doses and increasing if necessary). Drugs to think about using include:
  - benzodiazepines, such as midazolam, diazepam or lorazepam
  - neuroleptics, such as haloperidol or levomepromazine.

# 9.4 Managing respiratory distress

## 9.4.1 Review question

**What pharmacological and non-pharmacological interventions (excluding psychological) are effective for the management of respiratory distress in children or young people with a life-limiting condition who are approaching the end of life?**

## 9.4.2 Description of clinical evidence

The aim of this review was to assess the clinical effectiveness, the safety and the cost-effectiveness of pharmacological and non-pharmacological treatments for the management of respiratory distress in a child or young person with a life-limiting condition.

The aim was to include systematic reviews of randomised controlled trials (RCTs), RCTs, cohort studies and uncontrolled studies, but no evidence was found which met the inclusion criteria for this review.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

## 9.4.3 Summary of included studies

No evidence was found which met the inclusion criteria for this review.

## 9.4.4 Clinical evidence

No evidence was found which met the inclusion criteria for this review.

## 9.4.5 Economic evidence

This review question was prioritised for economic analysis.

A systematic review did not identify any relevant economic literature relating to pharmacological and non-pharmacological interventions (excluding psychological) for the management of respiratory distress in children and young people with a life-limiting condition who are approaching the end of life.

As no clinical evidence was identified de Novo analysis was not undertaken but costings of the various alternatives are presented below.

### 9.4.5.1 Pharmacological interventions

In addition to the drug costs there are other costs involved in the provision of pharmacological interventions, the most important of which relate to staff time, which will vary according to the route of administration. For example, in the NICE guideline on Bacterial meningitis and meningococcal septicaemia in children (CG102 – https://www.nice.org.uk/guidance/cg102/evidence/full-guideline-134564941), it was estimated that giving an intravenous drug would take 10 minutes of a Band 5/6 nurses time, which would include getting the drug and equipment to draw and make it up, checking the prescription and the patient; and delivery which takes 3–5 minutes. In addition it was estimated that cannula placement by a specialty registrar would take 5-10 minutes.

The Guideline Committee noted that drugs are often double checked in paediatric palliative care due to the small doses and/or local policy. Controlled drugs such as morphine and

midazolam legally have to be checked by 2 nurses. The unit costs for health care professionals typically involved in the administration of intravenous drugs is given in Table 74.

**Table 74: Staff unit costs**

| Staff | Unit cost | Source |
|---|---|---|
| Band 5 nurse | £105 | PSSRU 2015 |
| Band 6 nurse | £125 | PSSRU 2015 |
| Specialty registrar | £72 | PSSRU 2015 |

*Source/Note:*

*Based on per hour of patient contact and including qualification costs*

*Based on a 40-hour week and including qualification costs*

### Glycopyrronium bromide

The acquisition costs for various formulations of glycopyrronium bromide are listed in Table 75.

**Table 75: Glycopyrronium bromide acquisition costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 1mg | 30 | £178.50 |
| Tablet [a] | 2mg | 30 | £201 |
| Solution for injection [a] | 200mcg/ml | 10 ampoule | £8.28 |
| Solution for injection [b] | 600mcg/ml | 10 ampoule | £11.50 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289785/Part VIIIA products G (accessed 13/05/2016)*

*(b) BNFc indicative price https://www.medicinescomplete.com/mc/bnfc/current/PHP8204-glycopyrronium-bromide.htm?q=Glycopyrronium&t=search&ss=text&tot=31&p=2#_hit (accessed 12/05/2016)*

The Guideline Committee considered a typical dose for subcutaneous injection was 0.6-1.2mg/day. The daily cost of a dose of 0.6mg per day is calculated as follows:

| | |
|---|---|
| Administration: | Injection |
| Preparation: | Glycopyrronium bromide 600mcg/ml solution for injection ampoules |
| Cost: | £11.50 ampoules (pack of 10) Drug Tariff (Part VIIIA Category A) price |
| Cost/ampoule: | £1.15 |
| Dose: | 0.6mg |
| Ampoules/day: | 1 |
| Cost per day: | 1 x £1.15 = £1.15 |

### Nebulised salbutamol

The acquisition costs for various formulation of nebulised salbutamol are listed in Table 76.

**Table 76: Acquisition costs for nebulised salbutamol**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Nebuliser liquid [a] | 2.5mg/2.5ml | 20 unit dose | £1.91 |
| Nebuliser liquid [a] | 5mg/2.5ml | 20 unit dose | £3.82 |
| Nebuliser liquid [a] | 5mg/ml | 20 ml | £2.18 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289716/Part VIIIA products S (accessed 05/01/2016)*

### Nebulised ipratroprium

**Table 77: Acquisition costs for nebulised ipratropium**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Nebuliser liquid [a] | 250mcg/ml | 20 unit dose | £4.39 |
| Nebuliser liquid [a] | 500mcg/2ml | 20 unit dose | £2.88 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289251/Part VIIIA products I (accessed 05/01/2016)*

### Dexamethasone

The acquisition costs for various formulation of dexamethasone are given in Table 78.

**Table 78: Acquisition costs for dexamethasone**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 500mcg | 28 | £54.26 |
| Tablet [a] | 2mg | 50 | £49.22 |
| Oral solution [a] | 10mg/5ml | 150ml | £94.45 |
| Oral solution [a] | 2mg/5ml | 150ml | £42.30 |
| Solution for injection [a] | 3.8mg/ml | 10 vial | £19.99 |
| Solution for injection [b] | 6.6mg/2ml | 5 vial | £24.00 |
| Solution for injection [b] | 6.6mg/2ml | 5 ampoule | £11.00 |
| Solution for injection [b] | 3.3mg/ml | 10 ampoule | £12.00 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289643/Part VIIIA products D (Accessed 05/01/2016)*
*(b) BNFc NHS indicative price (accessed 05/01/2016)*

### Oral diazepam

The acquisition costs for oral diazepam are given in Table 79.

**Table 79: Acquisition costs for oral diazepam**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 2mg | 28 | £0.86 |
| Tablet [a] | 5mg | 28 | £0.90 |
| Tablet [a] | 10mg | 28 | £1.01 |
| Oral solution [a] | 2mg/5ml | 100ml | £31.75 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289643/Part VIIIA products D (accessed 07/01/2016)*

### Lorazepam

The acquisition costs for various formulations of Lorazepam are listed in Table 80.

**Table 80: Acquisition costs for lorazepam**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 1mg | 28 | £2.36 |
| Tablet [a] | 2.5mg | 28 | £3.23 |
| Oral solution [b] | 4mg/ml | 10 ampoule | £3.54 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289582/Part VIIIA products L (accessed 07/01/2016)*
*(b) BNFc NHS indicative price (accessed 07/01/2016)*

The Guideline Committee considered a typical dose for lorazepam for respiratory distress would be 1mg taken 3 times daily. The daily cost of this dose is shown below:

Administration: Tablet

Preparation: Lorazepam 1mg tablets

Cost: £2.36 (28 tablets) Drug Tariff (Part VIIIA Category A) price

Cost/tablet: £0.08

Dose: 3mg

Tablets/day: 3

Cost per day: 3 x £0.08 = £0.24

**Midazolam**

The acquisition costs for various formulations of midazolam are shown in Table 81

**Table 81: Acquisition costs for midazolam**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Oromucosal solution [a] | 10mg/2ml | 4 unit dose | £91.50 |
| Oromucosal solution [a] | 2.5mg/0.5ml | 4 unit dose | £82.00 |
| Oromucosal solution [a] | 5mg/ml | 4 unit dose | £85.50 |
| Oromucosal solution [a] | 7.5mg/1.5ml | 4 unit dose | £89.00 |
| Solution for injection [b] | 5mg/5ml | 10 ampoule | £6.00 |
| Solution for injection [b] | 10mg/5ml | 10 ampoule | £6.38 |
| Solution for injection [b] | 10mg/2ml | 10 ampoule | £6.25 |
| Solution for injection [b] | 50mg/10ml | 10 ampoule | £25.00 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289611/Part VIIIA products M (accessed 07/01/2016)*
*(b) BNFc indicative price (accessed 07/01/2016) https://www.medicinescomplete.com/mc/bnfc/current/PHP3037-midazolam.htm?q=midazolam&t=search&ss=text&tot=56&p=1#PHP77320-solution-for-injection (accessed 12/05/2016)*

The Guideline Committee suggested that 5mg would be a typical dose and the daily cost for this is described below.

Administration: Oromucosal solution

Preparation: Midazolam 5mg/1ml oromucosal solution, pre-filled oral syringes

Cost: £85.50 (4 unit dose) Drug Tariff (Part VIIIA Category A) price

Cost/unit: £21.38

Dose: 5mg

Units/day: 1

Cost per day: 1 x £21.38 = £21.38

## Morphine

**Table 82: Acquisition costs for morphine**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 10mg | 56 | £5.31 |
| Tablet [a] | 20mg | 56 | £10.61 |
| Tablet [a] | 50mg | 56 | £28.02 |
| Modified-release tablet [a] | 5mg | 60 | £3.29 |
| Modified-release tablet [a] | 10mg | 60 | £3.47 |
| Modified-release tablet [a] | 15mg | 60 | £9.10 |
| Modified-release tablet [a] | 30mg | 60 | £12.47 |
| Modified-release tablet [a] | 60mg | 60 | £24.32 |
| Modified-release tablet [a] | 100mg | 60 | £38.50 |
| Modified-release tablet [a] | 200mg | 60 | £81.34 |
| Modified-release capsule [a] | 10mg | 60 | £3.47 |
| Modified-release capsule [a] | 30mg | 60 | £8.30 |
| Modified-release capsule [a] | 60mg | 60 | £16.20 |
| Modified-release capsule [b] | 90mg | 28 | £22.04 |
| Modified-release capsule [b] | 100mg | 60 | £21.80 |
| Modified-release capsule [b] | 120mg | 28 | £29.15 |
| Modified-release capsule [b] | 150mg | 28 | £36.43 |
| Modified-release capsule [a] | 200mg | 60 | £43.60 |
| Modified-release granules [b] | 20mg | 30 sachet | £24.58 |
| Modified-release granules [b] | 30mg | 30 sachet | £25.54 |
| Modified-release granules [b] | 60mg | 30 sachet | £51.09 |
| Modified-release granules [b] | 100mg | 30 sachet | £85.15 |
| Modified-release granules [b] | 200mg | 30 sachet | £170.30 |
| Oral solution [a] | 10mg/5ml | 300ml | £5.45 |
| Oral solution [a] | 20mg/ml | 120ml | £19.50 |
| Solution for injection [b] | 10mg/10ml | 10 ampoule | £34.90 |
| Solution for injection [b] | 1mg/ml | 10 ampoule | £22.90 |
| Solution for injection [b] | 5mg/5ml | 10 ampoule | £32.20 |
| Solution for injection [a] | 10mg/ml | 10 ampoule | £9.36 |
| Solution for injection [a] | 15mg/ml | 10 ampoule | £8.95 |
| Solution for injection [b] | 20mg/ml | 10 ampoule | £46.99 |
| Solution for injection [a] | 30mg/ml | 10 ampoule | £8.84 |
| Solution for injection [b] | 60mg/2ml | 5 ampoule | £10.07 |
| Solution for infusion [b] | 50mg/50ml | 10 vial | £24.80 |
| Solution for infusion [b] | 100mg/50ml | 10 vial | £41.70 |
| Suppository [b] | 10mg | 12 suppository | £18.34 |
| Suppository [a] | 15mg | 12 suppository | £16.48 |
| Suppository [a] | 30mg | 12 suppository | £18.60 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289611/Part VIIIA products M (accessed 07/01/2016)*

*(b) BNFc indicative price (accessed 07/01/2016)*

## Diamorphine

**Table 83: Acquisition costs for diamorphine for various formulations**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 10mg | 100 | £25.29 |
| Powder for solution for injection [a] | 5mg | 5 ampoule | £11.36 |
| Powder for solution for injection [b] | 5mg | 5 vial | £11.89 |
| Powder for solution for injection [a] | 10mg | 5 ampoule | £14.33 |
| Powder for solution for injection [b] | 5mg | 5 vial | £15.99 |
| Powder for solution for injection [a] | 30mg | 5 ampoule | £15.46 |
| Powder for solution for injection [b] | 30mg | 5 vial | £16.99 |
| Powder for solution for injection [a] | 100mg | 5 ampoule | £42.39 |
| Powder for solution for injection [b] | 100mg | 5 vial | £42.99 |
| Powder for solution for injection [a] | 500mg | 5 ampoule | £187.70 |
| Powder for solution for injection [a] | 500mg | 5 vial | £209.00 |
| Nasal spray [b] | 1.6mg | 160 dose | £123.75 |
| Nasal spray [b] | 720mcg | 160 dose | £112.50 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289643/Part VIIIA products D (accessed 07/01/2016)*
*(b) BNFc NHS indicative price (accessed 07/01/2016)*

## Furosemide

**Table 84: Furosemide acquisition costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 20mg | 28 | £0.81 |
| Oral solution [a] | 20mg/5ml | 150ml | £14.49 |
| Solution for injection | 20mg/2ml | 10 ampoule | £3.50 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320568/Part VIIIA products F (accessed 12/05/2016)*
*(b) BNFc NHS indicative price*
*(c) https://www.medicinescomplete.com/mc/bnfc/current/PHP815-furosemide.htm?q=furosemide&t=search&ss=text&tot=93&p=1#PHP75596-solution-for-injection (accessed 12/05/2016)*

The Guideline Committee considered a typical dose for furosemide for respiratory distress would be 20-40mg a day. The daily cost of a 20mg dose is shown below:

| | |
|---|---|
| Administration: | Tablet |
| Preparation: | Furosemide 20mg tablets |
| Cost: | £0.81 (28 tablets) Drug Tariff (Part VIIIA Category A) price |
| Cost/tablet: | £0.03 |
| Dose: | 20mg |
| Tablets/day: | 1 |
| Cost per day: | 1 x £0.03 = £0.03 |

### Hyoscine hydrobromide

**Table 85: Hyoscine hydrobromide acquisition costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 300mcg | 12 | £1.67 |
| Chewable tablet [b] | 150mcg | 12 | £1.55 |
| Transdermal patch [b] | 1.5mg | 2 | £4.52 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320587/Part VIIIA products H (accessed 12/05/2016)*
*(b) BNFc indicative price*
*(c) https://www.medicinescomplete.com/mc/bnfc/current/PHP2576-hyoscine-hydrobromide.htm?q=hyoscine&t=search&ss=text&tot=28&p=1#PHP77566-chewable-tablet (accessed 12/05/2016)*

The Guideline Committee considered a typical dose for hyoscine hydrobromide for respiratory distress would be 300mcg taken 4 times daily. The daily cost of this dose is shown below:

Administration: Tablet

Preparation: Hyoscine hydrobromide 300microgram tablets

Cost: £1.67 (12 tablets) Drug Tariff (Part VIIIA Category A) price

Cost/tablet: £0.14

Dose: 1.2mg

Tablets/day: 4

Cost per day: 4 x £0.14 = £0.56

### Prednisolone

**Table 86: Prednisolone acquisition costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 5mg | 28 | £1.45 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320671/Part VIIIA products P (accessed 16/05/2016)*

### Sodium chloride

**Table 87: Sodium chloride acquisition costs**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Nebuliser liquid[a] | 2.5ml | 20 unit dose vials | £13.50 |

*(a) BNFc indicative price https://www.medicinescomplete.com/mc/bnfc/current/PHP5143-sodium-chloride.htm?q=sodium%20chloride&t=search&ss=text&tot=408&p=1#PHP76722-nebuliser-liquid (accessed 16/05/2016)*

## 9.4.5.2 Non pharmacological interventions

A range of non-pharmacological interventions were included in the protocol:

- repositioning
- fans and opening windows
- square breathing (breathing techniques)
- chest physiotherapy

- mechanical airway suctioning
- non-invasive ventilation (BIPAP, CPAP).

Costs are not provided for repositioning, square breathing, fans and opening windows because the costs associated with these interventions are trivial and/or can be subsumed within standard care.

**Table 88: Costs relating to non-pharmacological interventions for respiratory distress**

| Item | Cost | Source |
|---|---|---|
| Chest physiotherapy | £438 | NHS Reference Costs 2014-15 [a] |
| Manual Suction Unit Emivac | £83 | Medical Suction [b] |
| Portable Suction Unit Askir230 12 BR | £283 | Medical Suction [c] |
| Portable Suction Unit Askir36 BR | £383 | Medical Suction [d] |
| Suction Unit Aspiret | £149 | Medical Suction [e] |
| Suction Unit AskirC30 FS | £458 | Medical Suction [f] |
| Suction Unit Hospivac350 FS | £833 | Medical Suction [g] |
| Suction Unit Hospivac400 FULL | £1,158 | Medical Suction [h] |
| FLOVAC® Disposable Liners (1L) x 10 | £33 | Medical Suction [i] |
| OB2012 Portable Medical Suction Unit | £678 | DS medical [j] |
| BiPAP ST C SERIES INTERNATIONAL | £2,081 | NHS Supply Chain 2015 |
| BiPAP Synchrony International | £5,309 | NHS Supply Chain 2015 |
| REMstar Pro C-Flex+ Sys One 60 Srs GB | £269 | NHS Supply Chain 2015 |
| REMstar Auto A-Flex W/HUMID SD Card INT | £551 | NHS Supply Chain 2015 |
| Disposable CPAP unit 02-max full face mask x 10 | £607 | NHS Supply Chain 2015 |
| Disposable medical suction tubing | £5.94 | SP Services [k] |

*(a) Outpatient procedure; Service description: Paediatric respiratory medicine; Currency Code DZ30Z*
*(b) Portable suction unit, includes 400ML Reusable Collection Jar with Overflow Valve, 6x10 Silicone tubing, Ø 8-9-10 mm Conical Connector, Antibacterial and Hydrophobic Filter; http://www.medicalsuction.co.uk/manual-suction-unit-emivac.html (accessed 06/01/2016)*
*(c) Includes 1L Autoclavable Collection Jar with Overflow Valve System, 6x10 Silicone Tubing (autoclavable), Ø 8-9-10 mm Conical Connector, CH20 Cannula, Antibacterial and Hydrophobic Filter, Power Cord with Schuko Plug, 12V Car Adapter http://www.medicalsuction.co.uk/portable-suction-unit-askir230-12v.html (accessed 06/01/2016)*
*(d) Includes1L Autoclavable Collection Jar with Overflow Valve System, 6x10 Silicone Tubing (autoclavable, Ø 8-9-10 mm Conical Connector, CH20 Cannula, Antibacterial and Hydrophobic Filter, Power Cord with Schuko Plug, 12V Car Adapter, AC/DC Universal Adapter http://www.medicalsuction.co.uk/portable-suction-unit-askir36.html (accessed 06/01/2016)*
*(e) Home care suction unit; includes 1L Reusable Collection Jar with Overflow Valve System, 6x10 Silicone Tubing (autoclavable), Ø 8-9-10 mm Conical Connector, CH20 Cannula, Antibacterial and Hydrophobic Filter, Power Cord with Schuko Plug http://www.medicalsuction.co.uk/suction-unit-aspiret.html (accessed 06/01/2016)*
*(f) Home care/theatre suction unit; Includes 2 Autoclavable Collection Jars with Overflow Valve System 2L, 8x14 Silicone Tubing (autoclavable), Ø 10-11-12 mm Conical Connector, CH20 Cannula, Antibacterial and Hydrophobic Filter, Power Cord with Schuko Plug, Footswitch http://www.medicalsuction.co.uk/suction-unit-askirc30-footswitch.html (accessed 06/01/2016)*
*(g) Theatre suction unit; includes 2 Autoclavable Collection Jars with Overflow Valve System 2L, 8x14 Silicone Tubing (autoclavable), Ø 10-11-12 mm Conical Connector, CH20 Cannula, Antibacterial and Hydrophobic Filter, Power Cord with Schuko Plug, Footswitch http://www.medicalsuction.co.uk/suction-unit-hospivac350-footswitch.html (accessed 06/01/2015)*
*(h) Theatre suction unit; includes 2 Autoclavable Collection Jars with Overflow Valve System 2L, 8x14 Silicone Tubing (autoclavable), Ø 10-11-12 mm Conical Connector, CH20 Cannula, Antibacterial and Hydrophobic Filter, Power Cord with Schuko Plug, Electronic Change-over System and Footswitch http://www.medicalsuction.co.uk/suction-unit-hospivac400-full.html (accessed 06/01/2016)*
*(i) http://www.medicalsuction.co.uk/suction-unit-flovac-disposable-liners1l.html (accessed 06/01/2016)*
*(j) Includes Autoclavable 1000 ml jar for secretion collection with overflow valve and filter directly integrated in the cover, Patient tube with Yankauer sucker, Cable for connection to a 12 Volts C.C. in a vehicle.*
*(k) https://www.spservices.co.uk/item/Brand_DisposableSterileSuctionTubing-3mx7mm_54_0_2832_1.html (accessed 13/05/2016)*

The cost of mechanical airway suctioning would primarily consist of the equipment costs, consumables such as disposable liners used in the collection vessels and staff time. It is estimated that a suction kit would have a lifespan of 10 years (http://blog.sscor.com/how-often-you-should-really-replace-your-medical-suction-machine). Even taking the most expensive mechanical suction unit listed in Table 88 the equipment cost over such a lifespan becomes relatively trivial. The annual equivalent cost can be calculated as follows:

$$E = (K – (S \div (1+r)n) \div A(n,r)$$

Where:

E = equivalent annual cost

K = purchase price of equipment

S = resale value

r = discount (interest rate)

n = equipment lifespan

A(n, r ) = annuity factor* (n years at interest rate r )

A(n,r) = 8.32 for an interest rate of 3.5% over 10 years.

If it was assumed that the equipment has no resale value and a 3.5% discount rate then the annual equivalent cost of a medical suction kit costing £1,158 would be £160 or £0.44 per day.

The disposable liners used for collecting secretions cost £3.30 each and the suction tubing about £6 (see Table 88) and that cost would be incurred with each use of the equipment.

### 9.4.6 Evidence statements

No studies were included in the review.

### 9.4.7 Linking evidence to recommendations

#### 9.4.7.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were reduction of respiratory distress, child or young person subjective distress alleviated, and parents or carers' distress alleviated: whereas child or young person and their parents or carers' quality of life, child or young person and their parents or carers' satisfaction and the number of different types of interventions needed to change noise intensity were considered as important outcomes. No evidence was identified.

#### 9.4.7.2 Consideration of clinical benefits and harms

When the child or young person is approaching the end of life, altered breathing (for example, increased work of breathing, increased respiratory effort and respiratory rate, noisy breathing) is common. However, the causes of these symptoms can vary and the possible benefits of treatments should be weighed against the side effects of the interventions that are considered in the protocol.

The Committee recognised from their experience, that one cause of a change in breathing can be anxiety, and this should be considered and if it is contributing it should be appropriately addressed. The Committee did not want to make a prescriptive recommendation with regard to particular sedatives to manage anxiety related respiratory

distress due to the serious side effects that they may cause (vomiting and nausea or unwanted sedation). Reassurance may be effective as first line approaches. The Committee discussed the fact that noisy breathing at the end of life was sometimes more of a concern and a source of upset to the family members or carers than for the child or young person themselves. Good communication about this is therefore essential.

#### 9.4.7.3 Economic considerations

The cost of interventions for respiratory distress are relatively inexpensive and although it is a common symptom in the population of children and young people approaching the end of life, this population itself is small.

Reassurance may often obviate the need for pharmacological intervention and the Committee thought that non-pharmacological interventions (such as repositioning) were often more effective than pharmacological ones. Mechanical ventilation, while an option, is not routinely used in the context of breathlessness in palliative care, unless there is a clearly identified reversible cause with the prospect of meaningful recovery.

In this diverse population the management of respiratory distress is highly individualised and therefore the recommendations are not too directive. The Committee considered that there was not much variation in practice in how this symptom was managed and there is unlikely to be a significant resource impact from implementing the guideline recommendations on the management of respiratory distress.

#### 9.4.7.4 Quality of evidence

No studies were found for inclusion in this review.

#### 9.4.7.5 Other considerations

The Committee concluded that due to the lack of evidence, recommendations would be mainly based on Committee members' clinical experience, expert opinion and existing guidance.

In their discussion, the Committee members agreed that in order to effectively manage respiratory distress in children and young people living with life-limiting conditions who are approaching the end of life, the main steps should be assessment of underlying causes, and where appropriate treatment of reversible causes, establishing a treatment plan, with consideration of non-pharmacological and pharmacological interventions as appropriate, and regular re-assessment of the plan.

The Committee agreed that establishing the possible underlying causes of respiratory distress would help guide the treatment plan. They noted that when assessing the causes there might be a high degree of variability, but as a general guidance, it is important to explore the following contributor factors: anxiety, physical discomfort, accumulated airway secretions, infection and other acute medical disorders, for example lower respiratory infections, pleural effusion, bronchospasm or pulmonary oedema. They noted that it is important to assess the child or young person's environment.

Regarding the treatment, they agreed that it is important to discuss with the parents the available options, considering the benefits and harms. It is also important to reassure the parents, as for them the signs of respiratory distress can be quite worrying.

The Committee advised consideration of simple non-pharmacological approaches in the first instance. Non-pharmacological management should be considered as the first-line approach for the treatment of respiratory distress. A number of strategies were discussed, including airway positioning (for example, this is frequently used in children using a wheelchair, as sometimes the position of the head may be a possible cause for airway obstruction),

improving the air quality or airflow (for example, by opening windows, using a fan). Other more complex non-pharmacological interventions were also discussed, such as the use of airway suctioning. In relation to the latter, the Committee noted that this might not always be helpful unless the difficulty related to accumulated secretions and unnecessary or frequent suctioning might itself cause distress to some.

The Committee acknowledged that there are two recent Cochrane reviews for both the use of opioids (Barnes 2016) and benzodiazepines (Simon 2010) in adult palliative care, but nothing similar exists for the paediatric population. They agreed that results from these studies could not be generalised to a population of children and young people.

For pharmacological treatment, the role of different classes of drugs was discussed, including anti-secretory agents, bronchodilators, nebulised saline, anxiolytic agents, and opioids (low-dose morphine can be an opioid of choice, as it does not have significant sedative effects). These were listed as treatments to be considered as appropriate. These treatments were ordered alphabetically, so it does not appear as an escalating list, and the choice will depend on the child's individual circumstances and medical condition. The Committee also noted that the treatment should start with the lowest dosage, and then be titrated according to the need. Patches or oral formulations should be the first choice, as injections may be painful.

The role of non-invasive ventilation in children with advanced respiratory symptoms was also discussed, but no recommendation was made regarding its use, as it was agreed that expert advice should be sought beforehand.

The use of some drugs, such as anxiolytic agents and opioids can have a sedative effect. The Committee recognised that this needed to be thought about and discussed when considering these agents.

Finally the Committee discussed whether a research recommendation should be drafted for this topic. They agreed that there was considerable uncertainty because of the lack of evidence. In light of the Cochrane reviews citing evidence in adults they thought that this type of research was at least feasible and possible and would provide important information for future guidance.

#### 9.4.7.6 Key conclusions

The Committee concluded that when treating respiratory distress in children and young people approaching the end of life, it is important to be aware that contributing factors and underlying causes should be assessed and considered. Treatments could include repositioning, changes to the environment, or the management of underlying medical conditions that impact on breathing. The identified underlying cause should be addressed and treated and regular assessment should take place to review the effectiveness of the treatment.

Non-pharmacological management should be considered as the first-line approach for the treatment of respiratory distress. The Committee made a series of recommendations with regard to the assessment and management of altered breathing.

### 9.4.8 Recommendations

**110. If a child or young person is approaching the end of life and has respiratory distress, breathlessness or noisy breathing, think about and if possible treat the likely causes or contributing factors. If it is likely to be caused by:**

- Anxiety:
  - discuss why they are anxious

- reassure them and manage the anxiety accordingly
- consider breathing techniques and guided imagery.

- Physical discomfort – think about what could be causing the discomfort (for example their position) and help them with it if possible.
- Environmental factors – think about environmental changes such as changing the temperature.
- Accumulated airway secretions – think about repositioning, airway suctioning, physiotherapy or anti-secretory drugs.
- Medical disorders (for example pneumonia, heart failure, sepsis or acidosis) – use appropriate interventions (according to their Advance Care Plan) such as:
  - anti-secretory agents
  - bronchodilators
  - nebulised saline
  - sedatives or anxiolytic agents
  - opioids
  - oxygen.

**111. For children and young people who are approaching the end of life and have respiratory distress, breathlessness or noisy breathing that needs further assessment, consider referral to an appropriate specialist (for example, a respiratory or cardiac specialist).**

**112. If a child or young person is approaching the end of life and has respiratory distress, breathlessness or noisy breathing:**

- explain to them and to their parents or carers that these symptoms are common
- discuss the likely causes or contributing factors
- discuss any treatments that may help.

### 9.4.9 Research recommendations

**7. What is the acceptability, safety and effectiveness of oral / trans-mucosal opioids or benzodiazepines in the management of acute breathlessness in the context of end of life care?**

| Research question | What are the acceptability, safety and effectiveness of oral / trans-mucosal opioids or benzodiazepines in the management of acute breathlessness in the context of end of life care? |
|---|---|
| Why this is needed | |
| Importance to 'patients' or the population | Children may experience acute breathlessness as they approach the end of their life. This can be a terrifying experience both for the child and for those caring for them. A number of non-pharmaceutical options are available, but in some cases, rapidly effective, patient acceptable, pharmacological strategies are needed. |
| Relevance to NICE guidance | • High:<br>There were no studies suitable to be included in the review of evidence for the management of breathlessness in children at the end of life. Future guidance would be much more robust if some research was conducted specifically in this population. There are recent Cochrane reviews for both the use of opioids (Barnes 2016) and benzodiazepines (Simon 2010) in adult palliative care, but nothing similar exists for the paediatric population. Current practice is |

| Research question | What are the acceptability, safety and effectiveness of oral / trans-mucosal opioids or benzodiazepines in the management of acute breathlessness in the context of end of life care? |
|---|---|
| | therefore likely to be inconsistent and may need to be studied in a pilot project. |
| Relevance to the NHS | It is likely that a number of emergency hospital admissions could be prevented if families had effective medication for breathlessness management in the community. The cost of the medications involved is relatively small, however there is currently no licensed preparation of buccal opioid in a suitable dose range available. A trial buccal opioid product would need to be developed, in order to conduct the study. |
| National priorities | In European Commissioning the products that are intended for the diagnosis, prevention or treatment of life-threatening or very serious conditions that affect no more than 5 in 10,000 people in the European Union are highlighted as a special priority category.<br>To date, the European Commission has already authorised 126 medicines for the benefit of patients suffering from rare diseases.<br>Equally important, the European Commission has designated 1311 products as these types of medicinal products for rare diseases. This financial assistance provided for such products should facilitate the development and authorisation of innovative medicines for the benefit of the patients. |
| Current evidence base | While recent Cochrane reviews have been conducted on the management of breathlessness in adults at the end of life, there are no comparable studies in children. There are significant differences in the range of conditions involved in children's palliative care, as well as significant potential differences in pharmacokinetics / pharmacodynamics. |
| Equality | Children are therapeutic orphans in this context. Because they represent a smaller percentage of the population of patients facing acute breathlessness, they have not benefitted from any specific research. |
| Feasibility | While there are always issues to consider when conducting research in vulnerable subjects, children have an equal right to good quality research conducted for their benefit. This study would be numerically feasible as a national, multicentre trial. Recruitment ought to be possible in both hospice and hospital settings. It may be possible to conduct a randomised trial of the first line use of midazolam or morphine, since based on the findings in the Cochrane reviews for adults neither seem to be clearly superior. |
| Other comments | It may be possible to consider asking the Medicines for Children Research Network (MCRN) Pain & Palliative Care Clinical Studies Group to support or adopt such a study in an advisory capacity. Expertise may be needed from the MCRN formulations group with respect to producing a trial formulation of buccal opioid in the correct dose range, if this is to be included in the study. |

# 9.5 Managing seizures

## 9.5.1 Review question

**What pharmacological and non-pharmacological (excluding psychological) interventions are effective for the management of seizures in children and young people with a life-limiting condition who are approaching the end of life?**

## 9.5.2 Description of clinical evidence

The aim of this review is to assess the clinical effectiveness, the safety and the cost-effectiveness of pharmacological and non-pharmacological treatments for the management of seizures in children and young people with a life-limiting condition who are approaching the end of life.

We aimed to include systematic reviews of randomised controlled trials (RCTs), RCTs, cohort studies and uncontrolled studies, but no studies were identified in the search. Details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

## 9.5.3 Summary of included studies

No evidence was found which met the inclusion criteria for this review.

## 9.5.4 Clinical evidence

No evidence was found which met the inclusion criteria for this review.

## 9.5.5 Economic evidence

This review question was prioritised for economic analysis.

A systematic review did not identify any relevant economic literature relating to pharmacological and non-pharmacological interventions (excluding psychological) for the management of seizures in children and young people with a life-limiting condition who are approaching the end of life.

As no clinical evidence was identified de novo analysis was not undertaken but costings of the various alternatives are presented below.

### 9.5.5.1 Pharmacological interventions

In addition to the drug costs there are other costs involved in the provision of pharmacological interventions, the most important of which relate to staff time, which will vary according to the route of administration. For example, in the NICE guideline on Bacterial meningitis and meningococcal septicaemia in children (CG102), it was estimated that giving an intravenous drug would take 10 minutes of a Band 5/6 nurses time, which would include getting the drug and equipment to draw and make it up, checking the prescription and the patient; and delivery which takes 3–5 minutes. In addition it was estimated that cannula placement by a specialty registrar would take 5–10 minutes. The Guideline Committee noted that drugs are often double checked in paediatric palliative care due to the small doses and/or local policy. Controlled drugs such as morphine and midazolam legally have to be checked by 2 nurses. Unit costs for health care typically involved in the administration of an intravenous drug are given in Table 89.

**Table 89: Staff unit costs**

| Staff | Unit Costs | Source |
|---|---|---|
| Band 5 nurse [a] | £105 | PSSRU 2015 |
| Band 6 nurse [a] | £125 | PSSRU 2015 |
| Specialty registrar [b] | £72 | PSSRU 2015 |

*(a) Based on per hour of patient contact and including qualification costs*
*(b) Based on a 40-hour week and including qualification costs*

#### *9.5.5.1.1 Phenobarbital*

The acquisition costs of various formulations of phenobarbital are given in Table 90.

**Table 90: Acquisition costs for phenobarbital**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 15mg | 28 | £24.83 |
| Tablet [a] | 30mg | 28 | £0.85 |
| Tablet [a] | 60mg | 28 | £6.21 |
| Oral solution [a] | 15mg/5ml | 500ml | £83.00 |
| Solution for injection [b] | 200mg/ml | 10 ampoule | £60.57 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289531/Part VIIIA products P (accessed 06/01/2016)*
*(b) BNFc NHS indicative price https://www.medicinescomplete.com/mc/bnfc/current/PHP2950-phenobarbital.htm?q=phenobarbital&t=search&ss=text&tot=50&p=1#_hit (accessed 13/01/2016)*

The Guideline Committee suggested a typical dose of 125mg once daily and on this basis the daily cost of treatment would be calculated as follows:

| | |
|---|---|
| Administration: | Injection |
| Preparation: | Phenobarbital 200mg/1ml solution for injection ampoules |
| Cost: | £60.57 (10 ampoules) NHS indicative price |
| Cost/ampoule: | £6.06 |
| Dose: | 125mg per day |
| Ampoules per day: | 1 |
| Cost per day: | 1 x £6.06 = £6.06 |

#### *9.5.5.1.2 Phenytoin*

Table 91 shows the acquisition costs for various formulations of phenytoin

**Table 91: Acquisition costs for phenytoin**

| Formulation | Strength | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 100mg | 28 | £30.00 |
| Chewable tablet [b] | 50mg | 200 | £13.18 |
| Capsule [a] | 25mg | 28 | £15.74 |
| Capsule [a] | 50mg | 28 | £15.98 |
| Capsule [a] | 100mg | 84 | £54.00 |
| Capsule [a] | 300mg | 28 | £57.38 |
| Oral suspension [a] | 6mg/ml | 500ml | £4.27 |
| Solution for injection [b] | 250mg/5ml | 10 ampoule | £48.79 |
| Solution for injection [b] | 250mg/5ml | 5 ampoule | £14.55 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289531/Part VIIIA products P (accessed 06/01/2016)*
*(b) BNFc NHS indicative price (accessed 06/01/2016)*

A daily dose of 100mg 3 times daily was suggested by the Guideline Committee as a typical dose. The daily cost of treatment is calculated as shown below:

| | |
|---|---|
| Administration: | Injection |
| Preparation: | Phenytoin sodium 250mg/5ml solution for injection ampoules |
| Cost: | £14.55 (5 ampoule) NHS indicative price |
| Cost/ampoule: | £2.91 |
| Dose: | 100mg x 3 |
| Ampoules per day: | 3 |
| Cost per day: | 3 x £2.91 = £8.73 |

#### *9.5.5.1.3 Midazolam*

Acquisition costs for various formulations of midazolam are listed in Table 92.

**Table 92: Acquisition costs for midazolam**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Oromucosal solution [a] | 10mg/2ml solution | 4 pre-filled oral syringes | £91.50 |
| Oromucosal solution [a] | 2.5mg/0.5ml | 4 pre-filled oral syringes | £82.00 |
| Oromucosal solution [a] | 5mg/ml | 4 pre-filled oral syringes | £85.50 |
| Oromucosal solution [a] | 7.5mg/1.5ml | 4 pre-filled oral syringes | £89.00 |
| Solution for injection [b] | 2mg/ml | 10 ampoule | £4.50 |
| Solution for injection [b] | 5mg/5ml | 10 ampoule | £6.00 |
| Solution for injection [b] | 10mg/5ml | 10 ampoule | £6.38 |
| Solution for injection [b] | 10mg/2ml | 10 ampoule | £7.11 |
| Solution for injection [b] | 50mg/10ml | 10 ampoule | £25.00 |
| Solution for infusion [b] | 50mg/50ml | 1 vial | £9.56 |
| Solution for infusion [b] | 100mg/50ml | 1 vial | £9.05 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289611/Part VIIIA products M (accessed 06/01/2016)*
*(b) BNFc NHS indicative price (accessed 06/01/2016)*
*(c) A dose 100mcg/kg per hour was suggested by the Guideline Committee as a typical dose. An illustrative single day's treatment course is shown below for a young person of 20kg:*

| | |
|---|---|
| Administration: | Injection |
| Preparation: | Midazolam 10mg/2ml solution for injection ampoules |
| Cost: | £7.11 ampoules (pack of 10) NHS indicative price (BNFc) |
| Cost/ampoule: | £0.71 |
| Weight: | 20kg |
| Dose: | 100 mcg/kg/hour = 0.1 x 20 x 24 = 48mg |
| Ampoules/day: | 5 |
| Cost per day: | 5 x £0.71 = £3.55 |

#### 19.5.5.1.4 *Lorazepam*

The Acquisition cost for lorazepam solution for injection is shown in Table 93.

**Table 93: Acquisition costs for lorazepam**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Solution for injection [a] | 4mg/ml | 10 ampoule | £3.54 |

*(a) BNFc NHS indicative price (accessed 06/01/2016)*

The Guideline Committee suggested that a typical dose would be 4mg. On that basis the cost of a day's treatment would be as shown below:

Administration: Injection

Preparation: Ativan 4mg/1ml solution for injection ampoules

Cost: £3.54 (10 ampoules) NHS indicative price (BNFc)

Cost/ampoule: £0.35

Dose: 4mg

Ampoules/day: 1

Cost per day: 1 x £0.35 = £0.35

#### 19.5.5.1.5 *Rectal diazepam*

The acquisition cost for rectal diazepam is shown in Table 94.

**Table 94: Acquisition costs for rectal diazepam**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Rectal solution [a] | 10mg/2.5ml | 5 tubes | £7.35 |
| Rectal solution [a] | 5mg/2.5ml | 5 tubes | £5.85 |
| Rectal solution [b] | 2.5mg/1.25ml | 5 tubes | £5.65 |

*(a) NHS Drugs Tariff http://www.drugtariff.nhsbsa.nhs.uk/#/00289861-DD/DD00289643/Part VIIIA products D (accessed 07/0/2016)*

*(b) BNFc indicative price (accessed 06/01/2016)*

Based on a dose of 10mg per day the daily cost of treatment is as shown below:

Administration: Rectally

Preparation: Diazepam 10mg/2.5ml rectal solution tube

Cost: £7.35 (5 tube) Drug Tariff (Part VIIIA Category A) price

Cost/tube: £1.47

Dose: 10mg

Tubes/day: 1

Cost per day: 1 x £1.47 = £1.47

#### 19.5.5.1.6 *Clobazam*

**Table 95: Clobazam acquisition costs**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 10mg | 30 | £3.09 |

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Oral suspension [a] | 5mg/5ml | 150ml | £90.00 |

(a) *http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320527/Part VIIIA products C (accessed 12/05/2016)*

The Guideline Committee suggested that a typical dose would be 5mg a day increasing to 20-50mg per day. The daily cost based on 30mg a day is illustrated below:

Administration: Oral

Preparation: Clobazam 10mg tablets

Cost: £3.09 (30 tablets) Drug Tariff (Part VIIIA Category A) price

Cost/tablet: £0.10

Dose: 30mg

Tablets/day: 3

Cost per day: 3 x £0.10 = £0.30

#### *9.5.5.1.7 Clonazepam*

**Table 96: Clonazepam acquisition costs**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Tablet [a] | 2mg | 100 | £27.26 [b] |
| Oral solution [c] | 2mg/5ml | 150ml | £108.36 |

(a) *http://www.drugtariff.nhsbsa.nhs.uk/#/00320999-DC_1/DC00320527/Part VIIIA products C (accessed 12/05/2016)*
(b) *BNFc NHS indicative price £9.19*
(c) *https://www.medicinescomplete.com/mc/bnfc/current/PHP3010-clonazepam.htm?q=clonazepam&t=search&ss=text&tot=28&p=1#_hit (accessed 12/05/2016)*

A typical daily dose of 3-6mg was suggested by the Guideline Committee. The daily cost of treatment based on a dose of 4mg per day is calculated below:

Administration: Oral

Preparation: Clonazepam 2mg tablets

Cost: £27.26 (100 tablets) Drug Tariff (Part VIIIA Category A) price

Cost/tablet: £0.27

Dose: 4mg

Tablets/day: 2

Cost per day: 2 x £0.27 = £0.54

#### *9.5.5.1.8 Paraldehyde*

**Table 97: Paraldehyde acquisition costs**

| Formulation | Dose | Pack size | Cost |
|---|---|---|---|
| Solution for injection [a] | 5mg/5ml | 1 vial | Special order |

(a) *https://www.medicinescomplete.com/mc/bnfc/current/PHP12550-paraldehyde.htm?q=Paraldehyde&t=search&ss=text&tot=4&p=1#_hit (accessed 11/05/2016)*

#### 9.5.5.2 Non-pharmacological interventions

Music therapy can be used as a trigger avoidance strategy. Some exemplar costs of organisations who provide music therapy are shown in Table 98. Although these are not NHS providers they could be commissioned by the NHS. These costs are consistent with the approximate £40 per session that Cancer Research UK suggest is the cost of a typical private session arranged through the British Association of Music Therapists.

**Table 98: Exemplar music therapy costs**

| Provider | Cost per session | Notes |
|---|---|---|
| North Yorkshire Music Therapy Centre [a] | £42 | Individual session |
| Richmond Music Trust [b] | £45-£47 | Individual session |
| Richmond Music Trust [b] | £21 per client | Group session (3-5) |

*(a) http://www.music-therapy.org.uk/FAQ.html (accessed 06/01/2015)*
*(b) http://www.richmondmusictrust.org.uk/musictherapy (accessed 06/01/2015)*

### 9.5.6 Evidence statements

No studies were included in the review.

### 9.5.7 Linking evidence to recommendations

#### 9.5.7.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were reduction of seizures, child or young person's quality of life, child or young person levels of distress and parents or carers' satisfaction; whereas child or young person's satisfaction, parents or carers' distress alleviated, parents or carers' quality of life, and adverse events were rated as important. No evidence was identified.

#### 9.5.7.2 Consideration of clinical benefits and harms

The Committee considered it important to highlight that symptoms such as seizures can be common when the child or young person is approaching the end of life, particularly if they had seizures before or have an intracranial pathology. When pharmacological management is indicated the benefits of treatment should be weighed against the side effects of some of the drugs that are considered in the protocol which could, for instance cause unwanted levels of sedation. The distress and burden caused by seizures at the end of life can not only affect the child, but also have a detrimental effect on family members and caregivers who are not always aware of what the symptoms of the seizure are. Recommendations were made to support parents and carers in recognising these events, to help them manage these particularly if they happen at home.

Although there is a lack of evidence in this review, the Committee agreed that management of symptoms such as seizures would generally benefit the child and young person although the potential adverse effects of treatment would need individual consideration.

#### 9.5.7.3 Economic considerations

Most of the pharmacological interventions used to treat seizures are relatively cheap. Buccal preparations of midazolam are more expensive but the Committee reasoned that their use was important because of their convenience and ease of administration by children, young people and their families and carers. This made it easier to support children and young people in their preferred place of care. The Committee also noted that while the costs of the buccal preparation was more expensive it had much lower costs of administration than intravenous delivery for example, as healthcare professionals were not required.

### 9.5.7.4 Quality of evidence

Not applicable.

### 9.5.7.5 Other considerations

The Committee concluded that due to the lack of evidence, recommendations were mainly based on Committee members' clinical experience, expert opinion and consensus regarding accepted good clinical practice.

In their discussion the Committee members agreed that in order to effectively manage seizures in children and young people living with life-limiting conditions who are approaching the end of life, the main steps involved should be discussions with the parents or carers including the potential likelihood of seizures; the planning of treatment; and how to position the child if a seizure occurs at home. The recognition of seizures and assessment of underlying causes, triggers and contributing factors are also important. Non-pharmacological and pharmacological interventions should both be considered. In addition to those, the Committee gave special attention to those children receiving care in community settings and the fact that seizures must be frightening and upsetting, and might be thought to have a particular significance for some people, which may be related to their belief system.

Regarding recognition of seizures towards the end of life, the Committee noted that seizures at this stage are sometimes difficult to assess because healthcare professionals cannot always be certain whether a seizure is due to the child approaching the end of life or due to another underlying cause. For those children who are thought to be at significant risk of seizures (for example, those who were not pre-disposed to seizure disorders), healthcare professionals should discuss with families/ carers about the potential risks of recurrence of seizures, how seizures could be recognised and how they could be managed, whether there is an existing plan for their management, and whether this existing plan needs to be adapted or changed. Because seizures in children and young people could appear alarming or distressing, the Committee thought that healthcare professionals should forewarn parents/carers of this and prepare them for how to manage the seizures should they occur. The Committee emphasised that when assessing or recognising whether there are seizures, healthcare professionals should be aware that disorders of movement such as dystonic spasms could sometimes be mistaken for seizures and that this should be taken into consideration.

With regard to assessing or determining the cause or precipitating factors in the assessment of seizures, the Committee noted that the child and young person's medical condition, treatment and environment routines should be considered. They noted that there were a variety of contributing factors to seizures and emphasised that healthcare professionals should also assess factors such as environmental, sensory stimulation, drug reactions, pain, fever, and lack of sleep. The Committee advised that for those receiving end of life care in the home setting, attention should be given to preparing parents/carers for the possible occurrence of seizures, and if necessary, the actions they should take to manage seizures should they occur in their child.

The Committee did not make specific recommendations on the pharmacological management of seizures with anti-convulsants because this may not always be in the child or young person's best interest and is very condition specific. They did discuss the potential value of subcutaneous administration of anti-convulsants, and made a recommendation on providing this in a home setting. They did recommend that, if appropriate (i.e. when children were already on such medication due to their history or their condition and they had been provided with this as an option), parents and carers be prepared to give home anti-convulsive therapy if seizures occurred.

It is important that healthcare professionals discuss seizure management where appropriate, and explain the aim of controlling or reducing the distress caused by them. The Committee

specifically considered the needs of those receiving end of life care in community settings. They thought it was important to make the families/carers fully aware of the impact of management choices on the ability and possibility to deliver care in those settings. When discussing those issues, the families and the child/young person's preferences about place of care and death should always be considered. They agreed to recommend the use of anti-epileptic medications for children and young people having seizures, so that those medications will be available to them if they are cared for in this setting when needed.

Finally, the Committee discussed the need for re-assessment of the presence of seizures and agreed that this -assessment should be carried out regularly so as to tailor the treatment accordingly.

#### 9.5.7.6 Key conclusions

The Committee concluded that when assessing and treating seizures in children and young people approaching the end of life, it is important to be aware that seizures at this stage may be difficult to assess and appear distressing. This should be discussed with families/carers. For children and young people at significant risk of seizures who are being cared for at home or in community settings, healthcare professionals should prepare their families/carers for the management of seizures should they occur. Realistic management and treatment goals should be set up after full discussion with families/carers and decisions made jointly. If the child or young person is receiving care in a community setting, their parents or carers, if appropriate, should be taught how to deal with the seizures should they occur (for example, giving buccal midazolam). Underlying contributing factors to seizures should be considered and assessed before any treatment is given. Healthcare professionals should discuss with families/carers the impact of management choices on the ability to deliver care in specific settings while taking into account the families/carers and the child's preferred place for care and death. For children and young people receiving end of life care in community settings, the Committee also recommend appropriate medication should be available to them to be used at home when needed.

### 9.5.8 Recommendations

**113. If a child or young person is approaching the end of life and has a seizure, look for and if possible treat or remove any potential causes, triggers or contributing factors, for example:**

- fever
- electrolyte disturbances
- drug reactions
- sleep deprivation
- pain
- excessive environmental stimulation.

**114. If a child or young person is thought to be at increased risk of seizures, include seizure management in their Advance Care Plan. Think about the benefits and drawbacks of specific seizure treatments and:**

- take into account how any decisions could affect the choices available for place of care and place of death and
- discuss this with the child or young person and their parents or carers.

**115. For children and young people who are approaching the end of life, be aware that abnormal movements (such as dystonic spasms) might be mistaken for seizures. If in doubt seek specialist advice.**

**116. If a child or young person is approaching the end of life and is thought to be at increased risk of seizures (for example because they have had seizures before or because of an existing brain disorder), explain to them and their parents or carers:**

- how likely it is that they may have a seizure
- what they might notice if a seizure happens
- that seizures can be frightening or upsetting
- what parents or carers should do if a seizure happens at home (for example, placing the child or young person in a safe position).

**117. Ensure that parents or carers who have been provided with anticonvulsive therapy (such as buccal midazolam) know how and when to use it if the child or young person has a seizure at home.**

### 9.5.9 Research recommendations

**8. What is the acceptability, safety and effectiveness of delivering different subcutaneous infusions of anti-epileptic medication during the out of hospital management of persistent seizures close to the end of life?**

| Research question | What are the acceptability, safety and effectiveness of delivering different subcutaneous infusions of anti-epileptic medication in the out of hospital management of persistent seizures close to the end of life? |
|---|---|
| Why this is needed | |
| Importance to 'patients' or the population | Children may experience persistent seizures close to the end of their life. This may be due to raised intracranial pressure in the context of inoperable brain tumours, or may be a consequence of progressive neurological disease. Without access to an adequate evidence base relating to out of hospital management, the families of such children may have no options but to accept escalation of care to an intensive care unit in order to be sedated and ventilated. In many situations, this may be at odds with agreed goals of care at the time of Advance Care Planning. |
| Relevance to NICE guidance | • High:<br>Current NICE guidance makes reference to the possibility of subcutaneous infusions of anti-seizure medication, but there was no evidence at all to guide a more detailed recommendation. Other areas of the current NICE guideline highlight the importance of allowing families to choose a preferred place for end of life care, so more research is needed to allow children with difficult seizures to be offered the same options as other patient groups. |
| Relevance to the NHS | The cost of the parental preparations of medication are relatively small. There is a cost related to the device (subcutaneous infusion pump and disposables associated), but most palliative care teams would own this equipment. Community Nursing (or similar) staff time is needed, but children with this level of vulnerability would be already likely to be in receipt of visits at home. There is a large potential saving to the NHS, as most children managed in this way would otherwise be likely to require a Paediatric Intensive Care bed. |
| National priorities | *'Better Care, Better Lives. Improving outcomes for children and young people and their families living with life limiting or life threatening conditions'* (Department of Health, 2008) places a strong emphasis on equitable, individualised care planning and family friendly treatment choices. |
| Current evidence base | While a range of anti-convulsant drugs (phenobarbitone, midazolam, clonazepam, levatiracetam) are licensed for subcutaneous infusion, there is currently no research to support how they should be best used individually or in combination, in the face of persistent seizures in the context of end of life care for children or young people. There is consequently a lack of consensus about what represents best practice. The Committee acknowledged that there |

| Research question | What are the acceptability, safety and effectiveness of delivering different subcutaneous infusions of anti-epileptic medication in the out of hospital management of persistent seizures close to the end of life? |
|---|---|
| | was evidence for the management of status epilepticus in the general paediatric population. However, this The available evidence has currently only addressed deals with intravenous therapy, but only and does not provide clear consensus about the safest most effective drug for infusion in the face of persistent seizures. |
| Equality | Children for whom persistent seizures are likely to present at the end of life, are currently disadvantage in terms of the options open to them for preferred place of end of life care. |
| Feasibility | This would not be an expensive study to conduct, as the medications involved are readily available and most community teams / hospices already have the equipment required. As the numbers involved are (fortunately) small, this would need to be a multicentre trial on a national scale. The study would need to be conducted in care settings where infusions can be delivered subcutaneously via standard pumps, |
| Other comments | It may be possible to seek support or adoption from the Medicines for Children (NIHR) Pain & Palliative Care Clinical Studies Group, for such a study, in an advisory capacity, this could also act as a lever for funding. |

1

# 10 Managing hydration and nutrition

## 10.1 Managing hydration

### 10.1.1 Review question

**What is the effectiveness of medically-assisted hydration in infants, children and young people during end of life?**

### 10.1.2 Introduction

Appropriate hydration is seen as a basic element of care in all medical contexts. At a social and cultural level, to be thirsty or dehydrated is a discomfort that anyone can relate to, and a harm that we seek to protect people from wherever possible. When a child or young person is no longer able to drink (either unaided or with the help of others) it is often unclear whether the possible benefits of medically-assisted hydration at the end of life outweigh the harms. In making decisions about this healthcare professionals need to be mindful of the strong social, cultural, and moral imperative to avoid any sense of a child or young person suffering as a result of lack of fluids.

When considering the benefits and harms of artificial hydration it is also essential to consider if and when it ceases to be in the child's best interest or, when it may even be harmful to a child or young person reaching the end of their life. While it is important to ensure that, where appropriate, hydration is provided in the most effective manner there will be situations in which it will be clinically inappropriate to either start or maintain artificial hydration.

Given that the withholding or withdrawal of hydration may play against very basic human instincts, the issue needs to be handled sensitively and the feelings of the parents, family and carers must be acknowledged. Where there are clinical signs that suggest that continued artificial hydration may not be in the child or young person's best interest anymore, it is imperative to establish the best means of keeping them comfortable, to reassure the parents or carers and family, and to help them understand the continued value of providing appropriate mouth care and other comfort measures. Careful communication will help to ensure that families are not burdened by understandable but avoidable concerns around this issue.

### 10.1.3 Description of clinical evidence

The aim of this review was to determine the effectiveness of medically-assisted hydration in children and young people with a life-limiting condition during the last days of their life care. We looked for systematic reviews, randomised controlled trials, cohort studies and uncontrolled studies that looked to assess the effect of medically-assisted hydration on the quality of life, length of survival and satisfaction with care at the end life.

No evidence was found which met the inclusion criteria for this review.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

### 10.1.4 Summary of included studies

No evidence was found to meet the inclusion criteria for this review.

### 10.1.5 Clinical evidence

No evidence was found which met the inclusion criteria for this review.

### 10.1.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis

### 10.1.7 Evidence statements

No evidence was found which met the inclusion criteria for this review.

### 10.1.8 Linking evidence to recommendations

#### 10.1.8.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were child or young person's comfort or distress and parents or carers satisfaction; whereas adverse events (including vomiting, respiratory distress and abdominal pain) were rated as important outcomes. No evidence was identified.

#### 10.1.8.2 Clinical benefits and harms

In the care of a child or young person who is dying, administration of fluids is a very important matter for consideration. Many will be able and willing to take oral fluids and continuing to do so may contribute to their comfort. It may also be of great symbolic importance to them and to their parents or carers that they continue to take or be offered fluid in this way. The offering of drinks should be seen as a basic element of care. The Committee considered it was necessary however to recognise the important distinction between continuing to offer oral fluids and the decision to continue or even to begin giving fluids by other routes (clinically assisted hydration), such as via an enteral tube (for example, a nasogastric tube) or even by intravenous administration. Enteral tube and intravenous administration entail the use of more invasive techniques, and therefore the balancing of benefits and burdens may look different when considering how to serve the child's best interests. For some children, artificial hydration and nutrition will have already been part of their daily routine for some time, indeed they may never have been able to drink in the normal way. For these children it is important to acknowledge and respect what has been normal for them, and manage any change with openness and sensitivity.

#### 10.1.8.3 Economic considerations

The provision of fluids is a very important to the health-related quality of life of children and young people who are dying. Where possible, the guideline recommendations state that the child or young person should be supported and encouraged to drink if they want to. Lip and mouth care, also recommended in the guideline, is also a low cost intervention which can be considered to be a part of standard nursing care.

More invasive and expensive methods of hydration (enteral tube or intravenous fluids) are available when the child or young person is dying and cannot drink, but the Committee recognised that these interventions were not always in the child or young person's interest. Where more invasive methods of hydration are used it may be necessary for the place of care and death to be changed.

Due to the relatively low costs associated with the interventions and the fundamental needs involved, the Committee agreed that recommendations would not primarily be influenced by

the costs of treatment. Decisions to withhold such treatment based on cost alone would not be considered acceptable.

#### 10.1.8.4 Quality of evidence

No evidence was found to meet the inclusion criteria for this review.

### 10.1.9 Other considerations

The recommendations were based on Committee's expert opinion. Given the absence of clinical evidence for this review topic, the Committee considered and discussed in detail the importance of emotional, cultural, ethical, and legal issues that were relevant to decision-making.

Offering drinks to a sick person is generally perceived as essential basic care. In light of this the Committee considered that it was normally appropriate to offer oral fluids to children and young people who were dying. Assuming no serious counter indication, children and young people should be supported in drinking for as long as they wished to do so and for as long as they were able to take some fluid by mouth. The Committee considered that oral fluids willingly taken were often a source of comfort for the child or young person. Moreover, for the child or young person and for their parents or carers, ceasing to offer fluids could often have emotional and symbolic significance that needs to be acknowledged.

The Committee also discussed the matter of fluid administration by other routes including enteral tube administration and intravenous fluids. A decision to proceed with artificial hydration should take account of the different burdens or risks associated with different routes of administration. For example, excessive intravenous fluid administration could be hazardous. The Committee recognised that there is currently also a lack of understanding or consensus of what a dying child requires in terms of fluid intake which is another factor that makes it difficult to decide whether or not to use medically assisted hydration.

Taking less food and drink at the end of life may even be to a degree a physiological adjustment. 'Forcing normal hydration' on to a person at this time may add to their burden.

With these more invasive forms of hydration it is important Committee thought that it was crucial to consider whether it is in a child or young person's interest to start and then maintain the fluids, but it is also important to consider the possibility of withholding or withdrawing hydration if and when circumstances change. Particularly if enteral tube or intravenous fluids are given, the Committee agreed that decisions should be regularly reviewed to make sure that this remains appropriate and continues to be in their best interests. The Committee concluded that if a child or young person was dying and could not drink, it would be important to think about the value of enteral tube or even intravenous fluids on an individual basis. In balancing the potential benefits and burdens it would be important to take account of the previously stated wishes of the child or young person as well as the consequences of these more invasive procedures. For example, it might significantly affect management in different settings and might have consequences in relation to the options for preferred place of death. Placement of nasogastric tubes can be somewhat unpleasant and many children and young people are seriously distressed by the process of intravenous cannula insertion. Children and young people receiving intravenous fluids might also be subjected to blood sampling to monitor the serum electrolyte concentrations, and this could be a further burden for them.

The Committee emphasised the importance of making decisions about fluid administration in partnership with the child or young person and with their parents or carers as appropriate.

The Committee also discussed the importance of providing mouth and lip care. They recommended that this care should continue to be provided when the child or young person is dying in order to ensure comfort.

The Committee recognised the complexities involved in this aspect of care. The Committee were aware of and took account of guidance and general principles on this issue published by health professional bodies such as Royal College of Paediatrics and Child Health (RCPCH) and the General Medical Council. They recognised that decisions must be made within the legal framework. Where there was significant and unresolvable disagreement between families and healthcare professionals around withholding or withdrawing medically-assisted hydration legal advice should be sought. Ordinarily however the Committee felt that with the assistance of guidance on this issue published by health professional bodies such as Royal College of Paediatrics and Child Health (RCPCH) and the General Medical Council clinicians and families could work together to ensure that a child's best interests were served and unduly burdensome interventions avoided.

The Committee also discussed whether research should be recommended for this topic. However, they agreed that withholding or changing ways of hydrating children in the last hours of life would be research that is unlikely to be conducted due to the possible distress that it may cause. This was therefore not prioritised for further research.

#### 10.1.9.1 Key conclusions

The Committee concluded that: during the end of life care for infants, children or young people, while clinically assisted hydration may not be necessarily in the best interests of the child, hydration for comfort should be provided. As long as it remained in the child's best interests, fluids intake by their other usual routes of administration such as oral, tube feeding or intravenous should be continued while special attention should be given to the latter 2 due to the extra burden it could cause to the child or young person.

### 10.1.10 Recommendations

**118. If a child or young person with a life-limiting condition is approaching the end of life or is dying, discuss how to manage their fluid needs with them and their parents or carers.**

**119. If a child or young person is dying, encourage and support them to drink if they want to and are able.**

**120. If a child or young person is dying, continue to provide them with lip and mouth care.**

**121. If a child or young person is dying and cannot drink, discuss with them (as appropriate) and their parents or carers whether starting or continuing enteral tube or intravenous fluids is in their best interests.**

**122. Be aware that enteral tube and intravenous fluids may have a significant effect on care, may be a burden for children and young people, and may mean the place of care and place of death need to be changed.**

**123. If a child or young person is given enteral or intravenous fluids, review this decision regularly to make sure it continues to be in their best interests.**

**124.**

# 10.2 Managing nutrition

## 10.2.1 Review question

**What is the effectiveness of medically-assisted nutrition in infants, children and young people during end of life care?**

## 10.2.2 Introduction

Medical decisions regarding assisted nutrition at the end of life take place within a cultural context which values and celebrates the provision of sustenance. Feeding and being fed is usually seen as a source of pleasure, a sign of love, and for some children where artificial feeding has always been necessary it will have been an important element of the caring relationship.

Given the underlying assumptions about the value of nutrition, families are understandably concerned about the possibility of harms associated with withholding feeding, and some will be further concerned about the impact of the withdrawal of feeding upon the time and manner of death.

It is necessary for this guidance to establish what evidence exists to assist clinicians in deciding whether continued feeding by artificial means will be in the interests of each particular child. It is also important for the sensitivity of the issue to be acknowledged, and for the clinical practice to be linked to wider discussions about communication, trust and shared decision-making.

Decision-making by healthcare professionals needs to be medically and ethically robust particularly where withdrawing or withholding artificial feeding is being considered. It is often uncertain whether continued feeding through artificial means is in a child's best interest. Hunger and the desire to eat diminish when a person is dying. It is important to recognise this while ensuring that the child continues to receive such comfort as can be given through continued feeding. It is important to recognise the potential for burden through continued treatment with medically-assisted nutrition and the sensitivities around decision not to feed. With adequate planning and good communication this matter can be sensitively and collaboratively managed.

## 10.2.3 Description of clinical evidence

The aim of this review was to determine the effectiveness of medically-assisted nutrition in children and young people with a life-limiting condition during the last days of their life.

We looked for systematic reviews, randomised controlled trials, cohort studies and uncontrolled studies that looked to assess the effect of medically-assisted nutrition on the quality of life, length of survival and satisfaction with care at the end of life.

A Cochrane systematic review (Good 2014) was identified, but it aimed to find studies in the adult population and therefore was not included in this review. None of the included adult studies could be included here, nor did the excluded studies involve children. No studies matching the protocol were identified in our literature search.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H.

## 10.2.4 Summary of included studies

No evidence was found to meet the inclusion criteria for this review.

### 10.2.5 Clinical evidence

No evidence was found to meet the inclusion criteria for this review.

### 10.2.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis.

### 10.2.7 Evidence statements

No studies were included in the review.

### 10.2.8 Linking evidence to recommendations

#### 10.2.8.1 Relative value placed on the outcomes considered

The critical outcomes considered by the Committee were child or young person's comfort or distress and parents or carers satisfaction; whereas adverse events (including vomiting, respiratory distress and abdominal pain) were rated as important outcomes. No evidence was identified.

#### 10.2.8.2 Consideration of clinical benefits and harms

Given the absence of clinical evidence for this review topic, the Committee agreed that the considerations regarding medical nutrition were similar to those discussed for medical hydration.

The Committee emphasised that for many children and young people as they approach the end of life, eating continues to be important to them as an enjoyable experience. It is often also of emotional and symbolic importance for them, and for their parents or carers. Offering food is seen as an important element of basic care. It may also be that taking of some nutrition orally, even if in limited amounts, may support the person's feeling of well-being and add to their quality of life. Where a child has been fed artificially for some time – perhaps even from birth – it is important to acknowledge and respect what is normal for them, and to be sensitive to the impact of any proposed changes. Even if the child or young person is receiving nutrition via an enteral tube or (more unusually) intravenous nutrition, the Committee recognised that they would sometimes also take some oral nutrition provided it was clinically appropriate and they wanted to eat or drink. Similarly, with decisions related to artificial hydration, the route of administration should be chosen to minimise risk, and IV administration would be considered unusual in this context. For children with difficulties swallowing, the benefits and burdens of being allowed to eat would be considered and discussed with them, and with their parents or carers, as appropriate. For example, some might be at risk of pulmonary aspiration if given oral nutrition as they approached the end of life; however, there might still be occasions when the comfort associated with sharing food would be considered an important factor in the decision.

#### 10.2.8.3 Economic considerations

Eating is supported and encouraged if the child wants and is able to. If the child is unable to eat then enteral tube feeding or intravenous nutrition is recommended, which has greater resource implications. However, it is recommended with the proviso that artificial feeding should only be continued as long as it is in the best interests of the child or young person. Decisions to withhold such treatment based on cost alone would be considered unacceptable and in conflict with other guidance.

### 10.2.8.4 Quality of evidence

No evidence was found for this review.

### 10.2.8.5 Other considerations

The recommendations were based on the expert opinion of the Committee.

The Committee considered that, as a child or young person approaches the end of their life, they should be encouraged and supported with taking appropriate oral nutrition whenever possible, desired and normal for them. When it taking oral nutrition is not possible, however, the Committee agreed that decisions with regard to medically-assisted nutrition, like other aspects of management, should be made following discussion with the child or young person and their parents or carers as appropriate. For children and young people who are dying and who have been receiving medically-assisted nutrition, whether by enteral tube administration or intravenous administration, this should be reviewed. Such treatment should be continued if it is thought to be in their best interests. As with medically-assisted fluid administration, the Guideline Committee recognised that there were potential burdens associated with these approaches to delivering nutrition, and decisions about the balance of burden and benefit needed consideration on an individual basis. Because circumstances change as the end of life approaches, any decisions about nutritional management would need regular review and continuing discussion and the Committee made a recommendation accordingly.

The Committee also discussed the different administration routes. It was concluded that where it was felt that medically-assisted nutrition would serve a child's best interest it should be provided using the least invasive route that is appropriate for them.

The Committee acknowledged the importance of cultural, religious, ethical, and legal issues that have to be taken into consideration in the decision-making regarding medically-assisted nutrition. It was highlighted that there may be considerable variation in the cultural and symbolic values that families place on nutrition during end of life care and that this should be fully respected, and the child's own values should inform any best interest assessment.

As with medically-assisted hydration, the Committee recognised the complexities involved in this issue and emphasised that decisions have to be made within the legal framework. While legal advice might be needed in rare cases of intractable disagreement between clinicians and families, with adequate consideration and discussion it should be possible to reach agreement on what is in the child's best interest. The Committee were aware and took account of guidance and general principles on this issue published by health professional bodies such as the Royal College of Paediatrics and Child Health (RCPCH) and the General Medical Council. They recognised that healthcare professionals would cross-refer to such guidance if need arose.

The Committee also discussed whether research should be recommended for this topic. However, they agreed that withholding or changing ways of providing nutrition for children in the last hours of life would be research that is unlikely to be conducted due to the possible distress that it may cause. This was therefore not prioritised for further research.

### 10.2.8.6 Key conclusions

The Committee concluded that during the end of life care for children or young people, while medically-assisted nutrition may not be necessarily in the best interest of the child, it was important not to withhold oral nutrition if the child is able and wishes to eat. As long as it remained in the child's best interest, intake by their other usual routes of administration such as oral, tube feeding or intravenous should be continued, always taking into account the benefits and possible burdens for them.

### 10.2.9 Recommendations

**125. If a child or young person is approaching the end of life or is dying, discuss how to manage their nutritional needs with them and their parents or carers.**

**126. If a child or young person with a life-limiting condition is dying, encourage and support them to eat if they want to and are able.**

**127. If a child or young person is dying and they are receiving enteral tube feeding or intravenous nutrition:**

- discuss with them (as appropriate) or their parents or carers whether continuing this is in their best interest and
- review this decision regularly.

# 11 Recognising that a child or young person is likely to die within hours or days

## 11.1 Review question

**What signs and symptoms, individually or in combination, help to recognise that infants, children or young people are likely to be in the last days of life and which of them are considered most informative by healthcare professionals?**

## 11.2 Introduction

Experienced clinicians often claim that there is an art to recognising when a child or young person is dying which develops with experience and requires a particular set of skills. This may be true, but it is clearly the case that a better understanding of signs and symptoms associated with the dying process will also help professionals to recognise that a child or young person may be approaching the last days of life. While it is important to recognise signs and symptoms relevant to dying, it is also important to consider that some symptoms may be reversible given proportionate intervention, and that there are some signs which need to be investigated further before attributing them to the dying process. This guidance seeks to equip clinicians with the knowledge needed to recognise, as far as possible, that a child or young person is at the end of their life but also to deal with the uncertainty around this issue.

Having identified signs and symptoms that, either alone or in combination, may suggest a child or young person is in the last days of their life, it will be important to consider how best to utilise, communicate and share this information. Some families have lived with the reality of a life-limiting illness for many years, but death can still be unexpected. For others, a devastating illness may have struck suddenly and for others an antenatal diagnosis may have been made, or extreme prematurity may have meant that life was always precarious. Clinicians need to feel supported in communicating the realities of dying in a range of always difficult situations, mindful of the varied histories and needs of individual patients and their families.

By recognising and acknowledging the dying process, care teams try to seize the opportunity to respond in a timely manner to the individual needs of the child or young person and their parents or carers at this difficult time. In practical terms it allows the MDT to place urgency on responding to current or previously stated wishes regarding for example the end of life care, place of care, wishes, types of symptom treatment, and for organ and tissue donation and so on It allows the team to identify and call in the expertise needed to support and prepare the patient and their families clinically, psychologically and spiritually; the hope being that even a short period of time during which everyone knows that dying is in process will contribute towards securing as a good a death as possible.

## 11.3 Description of clinical evidence

The aim of this review was to identify signs and symptoms that help recognise that children and young people are likely to be in their last days of life.

This is a mixed methods review which allows for the inclusion of different study designs (both quantitative and qualitative) in order to fully understand an area of concern.

We looked for prospective and retrospective cohort studies to identify prognostic or diagnostic factors, but no studies were identified for inclusion.

We looked as well for studies that collected data using qualitative methods, such as Delphi consensus surveys and representative surveys of healthcare professionals experienced in paediatric palliative care. One study was identified for inclusion (Shaw 2014). This study was conducted in the UK, and included 49 healthcare professionals that were providing end of life care for children with life-limiting conditions. The authors used a modified Delphi methodology.

A summary of the included study is presented in Table 99.

Full details of the review protocol are reported in Appendix D. The search strategy created for this review can be found in Appendix E. A flow chart of the study identification is presented in Appendix F. Full details of excluded studies can be found in Appendix H. Evidence from the included studies is summarised in the evidence tables in Appendix and in the GRADE profiles below and in Appendix J. Due to the nature of this study, evidence is summarised in a summary table within the evidence report. Therefore no separate Appendix is provided for this.

## 11.4 Summary of included studies

### 11.4.1 Quantitative review

No studies were identified.

### 11.4.2 Qualitative review

A summary of the study that was included in this review is presented in Table 99.

**Table 99: Summary of included studies**

| Study | Data collection method | Participants | Aim of the study | Comments |
|---|---|---|---|---|
| Shaw 2014<br>UK | Modified Delphi survey:<br>• Item generation were derived from integrative literature review and focus group and review group<br>• Itineration process limited to 2 rounds | N = 49<br>• A representative mix of healthcare professionals with expertise in paediatric palliative care.<br>• The panel included nurses, specialist paediatricians, community paediatricians, consultants in paediatric care and GPs.<br>• Key palliative care environments, such as hospital, community and children's hospices.<br>• Geographical diversity (not specified) | To identify signs and symptoms that indicate that a child with a life-limiting condition is moving into an end of life phase. | • Quality assessment was carried out using specific criteria for the assessment of Delphi studies (Diamond 2014): total score 2/ 4 (this is a modified Delphi survey, some items do not apply)<br>• Specific number of rounds, without a formal criterion for consensus<br>• UK-based study.<br>• Age group not specified.<br>• Medical conditions not specified. |

# 11.5 Clinical evidence

## 11.5.1 Quantitative review: clinical evidence

No evidence was found to meet the inclusion criteria for this part of the review. Qualitative review: clinical evidence

## 11.5.2 Clinical evidence profile

The clinical evidence for recognising dying is presented in Table 100, Table 101, Table 102, Table 103 and Table 104.

**Table 100: Summary of clinical evidence Physical changes**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of finding** | **Criteria** | **Rating** | **Overall** |
| **Changes to breathing pattern** | | | | | |
| 1 (Shaw 2014) | 1 Delphi study | One Delphi study asked palliative care professionals providing end of life care for children with life-limiting conditions (n=49) to identify the signs and symptoms that are most valuable in identifying children approaching the end of life.<br>**The findings show that:**<br>(key to ratings: 1=always; 2=very often; 3=often; 4=sometimes; 6=rarely; 7=never; 8=no opinion)<br>(* items that were modified by the participants)<br>The following symptoms are very often present:<br>Abnormal breathing patterns (for example apnoea, Cheyne Stokes): 2 (0.33)<br><br>**The following symptoms are often present:**<br>• Breathing that is noisy/ bubbly (where breathing was previously unaffected)*: 3 (0.35)<br>• Breathing that is laboured/ irregular (where breathing was previously unaffected)*: 3 (0.41) | Limitation of evidence | Major limitations | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of finding** | **Criteria** | **Rating** | **Overall** |
| | | **The following symptoms are sometimes present:**<br>• Persistent increased suction requirements: 4 (0.45)<br>• Previously beneficial oxygen in no longer effective: 4 (0.73)<br>• Severe chest infection: 4 (2.29)<br><br>**The following symptoms are rarely present:**<br>• Objective methods showing a decline (authors do not specify what they mean by objective methods): 6 (1.12) | | | |
| **Circulatory changes** | | | | | |
| 1 (Shaw 2014) | 1 Delphi study | One Delphi study asked palliative care professionals providing end of life care for children with life-limiting conditions (n=49) to identify the signs and symptoms that are most valuable in identifying children approaching the end of life.<br>**The findings show that:**<br>(key to ratings: 1=always; 2=very often; 3=often; 4=sometimes; 6=rarely; 7=never; 8=no opinion)<br>(* items that were modified by the participants)<br><br>**The following symptoms are very often present:**<br>• Peripheral shutdown (increased capillary refill time): 2 (0.39)<br>**The following symptoms are often present:**<br>• Grey skin pallor: 3 (0.57)<br>• Instability of vital signs (temperature, blood pressure, respiratory rate, heart rate): 3 (0.64)<br><br>**The following symptoms are sometimes present:**<br>• Pressure areas fail to heal despite optimal management: 4 (0.40) | Limitation of evidence | Major limitations | LOW |

| Study information | | Description of finding | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | | **Criteria** | **Rating** | **Overall** |
| | | • Oedema of extremities: 4 (0.60)<br>• Oedematous skin: 4 (0.69) | | | |
| **Feeding** | | | | | |
| 1 (Shaw 2014) | 1 Delphi study | One Delphi study asked palliative care professionals providing end of life care for children with life-limiting conditions (n=49) to identify the signs and symptoms that are most valuable in identifying children approaching the end of life.<br>**The findings show that:**<br>(key to ratings: 1=always; 2=very often; 3=often; 4=sometimes; 6=rarely; 7=never; 8=no opinion)<br>(* items that were modified by the participants)<br><br>**The following symptoms are often present:**<br>• Not tolerating feeds/ less well absorbed: 3 (0.47)<br>• Not wanting to drink (as opposed to eat) – if orally fed: 3 (0.38)<br>• Reduced urine output: 3 (0.49)<br>• Anorexia (if orally feed): 3 (0.63)<br>• Increasing feeding difficulties: 3 (0.61)<br><br>**The following symptoms are sometimes present:**<br>• Cachexia: 4 (0.74) | Limitation of evidence | Major limitations | LOW |

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2 **Table 101: Summary of clinical evidence Neurological changes**

| Study information | | Description of finding | Quality assessment | | |
|---|---|---|---|---|---|

| Number of studies | Design | | Criteria | Rating | Overall |
|---|---|---|---|---|---|
| **Neurological changes** | | | | | |
| 1 (Shaw 2014) | 1 Delphi study | One Delphi study asked palliative care professionals providing end of life care for children with life-limiting conditions (n=49) to identify the signs and symptoms that are most valuable in identifying children approaching the end of life.<br>**The findings show that:**<br>(key to ratings: 1=always; 2=very often; 3=often; 4=sometimes; 6=rarely; 7=never; 8=no opinion)<br>(* items that were modified by the participants)<br><br>**The following symptoms are very often present:**<br>• Reduced level of consciousness (reduced Glasgow Coma Scale): 2 (0.20)<br>• Asleep more often than awake: 2 (0.24)<br>• No longer relating/ less responsive: 2 (0.33)<br><br>**The following symptoms are often present:**<br>• Less alert: 3 (0.35)<br>• Increased confusion: 3 (0.55)<br>• Intractable seizures despite optimal management: 3 (0.57)<br>• Increased analgesia requirement/ increased pain: 3 (0.59)<br>• Too weak to swallow tablets or medicines: 3 (0.69)<br>• Unnatural tiredness: 3 (0.69)<br>• New profound weakness: 3 (0.73)<br><br>**The following symptoms are sometimes present:**<br>• Increased calmness/ severity: 4 (0.33)<br>• New or accelerating cognitive impairment: 4 (0.50)<br>• New of accelerating muscle spasms: 4 (0.60)<br>• Delirium: 4 (0.63) | Limitation of evidence | Major limitations | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of finding** | **Criteria** | **Rating** | **Overall** |
| | | • New loss ability to feed self: 4 (0.65)<br>• New loss of continence: 4 (0.65)<br>• New loss of mobility: 4 (0.67)<br>• New onset loss of distinction between day and night: 4 (0.79)<br>• Increased agitation: 4 (0.88) | | | |

1 **Table 102: Summary of clinical evidence Changes in disease trajectory**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of finding** | **Criteria** | **Rating** | **Overall** |
| **Changes in the disease trajectory** | | | | | |
| 1 (Shaw 2014) | 1 Delphi study | One Delphi study asked palliative care professionals providing end of life care for children with life-limiting conditions (n=49) to identify the signs and symptoms that are most valuable in identifying children approaching the end of life.<br>**The findings show that:**<br>(key to ratings: 1=always; 2=very often; 3=often; 4=sometimes; 6=rarely; 7=never; 8=no opinion)<br>(* items that were modified by the participants)<br><br>**The following symptoms are very often present:**<br>• Does not return to previous level of health: 2 (0.29)<br><br>**The following symptoms are often present:**<br>• Increasing debility in response to lesser illness: 3 (0.25)<br>• Not responding to treatment/ intractable symptoms: 3 (0.31)<br>• Persistent increase in care needs both day and night: 3 | Limitation of evidence | Major limitations | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of finding** | **Criteria** | **Rating** | **Overall** |
| | | (0.37)<br>• Takes longer to recover to usual level of health: 3 (0.40)<br>• Infections not responding to treatment: 3 (0.43)<br>• Change in appearance (that is, looks more unwell)*: 3 (0.46)<br>• Increased frequency of chest infections: 3 (0.49)<br>• Episode of critical care: 3 (0.50)<br>• Increased medication needs: 3 (0.53)<br>• Increasing contact with out of hours services: 3 (0.59)<br>• Agreement that the child or young person is not for ITU/ emergency care; has a DNAR: 3 (0.65)<br>• Increasing irreversible loss of function of a major organ (for example lungs): 3 (0.79)<br>• Repeated need for PICU (whether given or not): 3 (0.79)<br><br>**The following symptoms are sometimes present:**<br>• Increased frequency of intercurrent illness: 4 (0.37)<br>• Onset of significant new symptoms: 4 (0.48)<br>• Increasingly sleepless nights: 4 (0.53)<br>• Increased appropriate hospital admissions despite community team care availability (6 annually): 4 (0.54)<br>• Increased appropriate hospital admissions despite community team care availability (>10 annually): 4 (0.71)<br>• Referral to hospice: 4 (0.75)<br>• Increased appropriate hospital admissions despite community team care availability (2 annually): 4 (0.83)<br>• Increased frequency of blood stained or coffee ground aspirates from gastrostomy or nasogastric tube: 4 (0.83)<br>• Untreatable oncology/ haematology condition: 4 (0.84)<br>• Inoperable heart defect with persistent hypoxia below 70% or intractable congestive cardiac failure: 4 (0.90) | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of finding | Criteria | Rating | Overall |
| | | • Bleeding with or without platelet support: 4 (0.90)<br>• Haemoptysis/ haemotemesis: 4 (1.00)<br>• Intractable liver failure with encephalopathy: 4 (1.07)<br>• Severe/ persistent secondary pulmonary hypertension: 4 (1.44) | | | |

*ITU: intensive therapy unit; PICU: paediatric intensive care unit*

**Table 103: Summary of clinical evidence psychosocial changes**

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of finding | Criteria | Rating | Overall |
| **Behaviour or emotion state** | | | | | |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| **Number of studies** | **Design** | **Description of finding** | **Criteria** | **Rating** | **Overall** |
| 1 (Shaw 2014) | 1 Delphi study | One Delphi study asked palliative care professionals providing end of life care for children with life-limiting conditions (n=49) to identify the signs and symptoms that are most valuable in identifying children approaching the end of life.<br>**The findings show that:**<br>(key to ratings: 1=always; 2=very often; 3=often; 4=sometimes; 6=rarely; 7=never; 8=no opinion)<br>(* items that were modified by the participants)<br><br>**The following symptoms are often present:**<br>• Attitude change in carer (more hopeless, more fear, more angry, more accepting, planning ahead for death): 3 (0.42)<br><br>**The following symptoms are sometimes present:**<br>• Difficulties talking about feelings with parents/ significant others: 4 (0.43)<br>• Attitude change in CYP (more hopeless, more fear, more angry, more accepting, planning ahead for death): 4 (0.54)<br>• Increase in family stress levels/ decrease in coping abilities: 4 (0.75) | Limitation of evidence | Major limitations | LOW |
| **Social withdrawal** | | | | | |

| Study information | | Description of finding | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | | Criteria | Rating | Overall |
| 1 (Shaw 2014) | 1 Delphi study | One Delphi study asked palliative care professionals providing end of life care for children with life-limiting conditions (n=49) to identify the signs and symptoms that are most valuable in identifying children approaching the end of life. The findings show that:<br>(key to ratings: 1=always; 2=very often; 3=often; 4=sometimes; 6=rarely; 7=never; 8=no opinion)<br>(* items that were modified by the participants)<br>**The following symptoms are often present:**<br>• Reduced efforts to present self to usual standard (where CYP has some independence in self-care)*: 3 (0.59)<br>• Decreased participation in valued activities: 3 (0.65) | Limitation of evidence | Major limitations | LOW |

1 **Table 104: Summary of clinical evidence Clinical judgement**

| Study information | | Description of finding | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | | Criteria | Rating | Overall |
| **Clinical judgement** | | | | | |
| 1 (Shaw 2014) | 1 Delphi study | One Delphi study asked palliative care professionals providing end of life care for children with life-limiting conditions (n=49) to identify the signs and symptoms that are most valuable in identifying children approaching the end of life.<br>The findings show that:<br>(key to ratings: 1=always; 2=very often; 3=often; 4=sometimes; 6=rarely; 7=never; 8=no opinion)<br>(* items that were modified by the participants)<br>**The following is very often present:**<br>• Gut feeling/ intuition of health professional: 2 (0.33) | Limitation of evidence | Moderate limitations | LOW |

| Study information | | | Quality assessment | | |
|---|---|---|---|---|---|
| Number of studies | Design | Description of finding | Criteria | Rating | Overall |
| | | • **The following is often present:**<br>• Gut feeling/ intuition of carers: 3 (0.58)<br>**The following is sometimes present:**<br>• Gut feeling/ intuition of child or young person where their cognitive function allows assessment: 4 (0.65) | | | |

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## 11.6 Economic evidence

No health economic evidence was found and this question was not prioritised for health economic analysis.

## 11.7 Evidence statements

### 11.7.1 Quantitative review: evidence statements

No evidence was found.

### 11.7.2 Qualitative review: evidence statements

Low quality evidence from 1 study, which used a modified Delphi survey method with a panel of palliative care professionals (n = 49, providing end of life care for children with life-limiting conditions), reached consensus in relation to the frequency of signs and symptoms of breathing changes to indicate the last week of life. Participants agreed that abnormal breathing patterns were very often present. Breathing that is noisy/bubbly or breathing that is laboured/irregular (where breathing was previously unaffected) are also often present.

Low quality evidence from 1 study, which used a modified Delphi survey method with a panel of palliative care professionals (n = 49, providing end of life care for children with life-limiting conditions), reached consensus in relation to the frequency of signs and symptoms circulatory changes that indicate the last week of life. Participants agreed that peripheral shutdown (increased capillary refill time) is very often present. Grey skin pallor and instability of vital signs (temperature, blood pressure, respiratory rate, and heart rate) are also often present.

Low quality evidence from 1 study, which used a modified Delphi survey method with a panel of palliative care professionals (n = 49, providing end of life care for children with life-limiting conditions), reached consensus in relation to the frequency of signs and symptoms of feeding changes to indicate the last week of life. Participants agreed that not tolerating feeds and feeds that are less well absorbed, not wanting to drink (as opposed to eat) if orally fed, reduced urine output, anorexia (if orally feed) and increasing feeding difficulties are often present.

Low quality evidence from 1 study, which used a modified Delphi survey method with a panel of palliative care professionals (n = 49, providing end of life care for children with life-limiting conditions), reached consensus in relation to the frequency of signs and symptoms of neurological changes to indicate the last week of life. Participants agreed that reduced level of consciousness, being asleep more often than awake, and no longer relating and being less responsive are very often present. Being less alert, increased confusion, intractable seizures despite optimal management, increased analgesia requirement and increased pain, being too weak to swallow tablets or medicines, unnatural tiredness and new profound weakness are also often present.

Low quality evidence from 1 study, which used a modified Delphi survey method with a panel of palliative care professionals (n = 49, providing end of life care for children with life-limiting conditions), reached consensus in relation to the frequency of signs and symptoms of changes in disease trajectory to indicate the last week of life. Participants agreed that not returning to previous level of health is very often present. Other signs and symptoms which are also often present include:

- Increasing debility in response to lesser illness
- not responding to treatment/intractable symptoms

- a persistent increase in care needs both day and night
- taking longer to recover to usual level of health
- infections not responding to treatment
- change in appearance (that is, looking more unwell)
- increased frequency of chest infections
- having an episode of critical care
- increased medication needs
- increasing contact with out of hours services
- agreement that the child or young person will not receive ITU/emergency care
- having a do-not-resuscitate order
- increasing irreversible loss of function of a major organ (for example lungs)
- repeated need for PICU (whether given or not).

Low quality evidence from 1 study using a modified Delphi survey method with a panel of palliative care professionals (n = 49, providing end of life care for children with life-limiting conditions), reached consensus in relation to the frequency of signs and symptoms of psychosocial changes to indicate the last week of life. Attitude change in carers (such as more hopeless, more fear, more angry, more accepting, planning ahead for death), reduced efforts to present self to usual standard (where child or young person has some independence in self-care) and decreased participation in valued activities are often present.

Low quality evidence from 1 study using a modified Delphi survey method with a panel of palliative care professionals (n = 49, providing end of life care for children with life-limiting conditions), reached consensus in relation to healthcare professional intuition, and to a lesser extend carers intuition, to indicate the last week of life.

## 11.8 Linking evidence to recommendations

### 11.8.1 Relative value placed on the outcomes and themes considered

For the quantitative review, the critical outcome considered by the Committee was dying within the next few days. No evidence was identified. For the qualitative review, although themes were mainly identified from the literature, the Committee identified some expected themes that they thought would be important during the protocol stage. These included signs and symptoms in at least 1 of the following categories, all of these were reported in the Delphi consensus study that was included in this review:

- Deterioration in level of consciousness,
- Deterioration in cognition,
- Change in skin (for example colour or temperature),
- Loss of willingness to take oral fluids,
- Loss of willingness to eat,
- Ability to tolerate feeding
- Altered behaviour or emotional state (for example agitation or anxiety),
- Social withdrawal (for example cessation of talking),
- Loss of urine output
- Change in vital signs (heart rate / pattern and respiratory rate / pattern).

The Committee agreed that all of these signs and symptoms were important.

### 11.8.2 Consideration of clinical benefits and harms

The Committee discussed at length the importance of recognising that a child or young person was likely to die in the next few hours or days. The need and value of addressing this question in the guideline was considered, given the potential advantages as well as the limitations, because of the uncertainty around these signs and symptoms.

The Committee pointed out that knowing that a child or young person is nearing death may be very important to the child or young person and their parents, family or carers in making decisions, for example with regard to place of care. Practical issues were discussed, for example situations where parents, parents or carers need to know if they can leave the child to spend a night away from hospital or simply go home to take a shower. If the child or young person is nearing death, the parents may want to ask extended family to visit the child.

The Committee discussed the, now discontinued, Liverpool Care Pathway. This was a pathway which was developed to aid members of a multi-disciplinary team in matters relating to continuing medical treatment, discontinuation of treatment and comfort measures during the last days and hours of a patient's life. However, a review was carried out and findings suggested that this pathway was widely considered to be a ‘tick box exercise’. The Committee made reference to the findings of this review into the Liverpool Care pathway (More Care, Less Pathway, DoH 2013), where some parents, families and carers mentioned that they would have liked to have been told that their relative was approaching death. They would also have liked to have been prepared for the symptoms that may occur during this period, for example excess respiratory secretions (‘death rattle’). The Committee agreed that this would also relate to the topics of communication and information provision.

Having acknowledged how important it is to determine when a child or young person is approaching the end of life, the Committee also discussed the difficulties in identifying when this point is and the associated potential harms. The Committee agreed that the issue of uncertainty is central to end of life decision-making, and they discussed that, according to their experience, there could be as many as one-third of children and young people who have been identified as likely to die within hours or days but who may actually live for longer than that.

The Committee agreed that when assessing prognosis, it is important to be aware that the signs and symptoms may be different for each child and highly dependent on the specific life-limiting condition. In particular, it was pointed out that in children and young people with complex health needs (for example neuro-disability) the signs and symptoms may be substantially different, and therefore specialist expertise may be required to assess them. To overcome this limitation, the Committee agreed that it is important to use a baseline reference for the symptoms; that is, the difference between these late symptoms and the child or young person’s more general signs and symptoms prior to this point. Also, baselines can differ depending on the child (for example, children with a cardiac medical condition). Without knowledge of the individual child or young person and their underlying condition, a standard list of signs and symptoms can actually be more harmful than beneficial. Recommendations with regard to these signs should not be interpreted as a checklist. For example, a child or young person may have an infection that can be treated, but it might be decided not to treat it if it is interpreted as a sign that they are nearing death. Other than the progressive deviation from their normality, the Committee noted the importance of symptoms not being reversible, despite adequate treatment.

The Committee agreed that it is important to be aware that some parents may want to know about the prognosis of an imminent death, and therefore medical knowledge needs to be sensitively handled and conveyed to them. Furthermore, while having a list of signs and symptoms can be useful, there are limitations, mostly due to the high level of uncertainty, and the need to avoid an inappropriate check list approach.

A concern raised by the Committee was that no inference about levels of interventions should be made on the basis of signs and symptoms alone. A child or young person's treatment should be based on their existing Advance Care Plan, and changes to the plan should be discussed if necessary. The Committee agreed that these discussions should involve the child or young person, the parents or carers and the healthcare professionals, and where necessary should address aspects related to withdrawal or withholding inappropriate interventions. In relation to this it is important to take into account that some children may survive for longer than expected after withdrawal of treatment. Once again communication and information provision are key aspects in this regard.

### 11.8.3 Economic considerations

Many of the recommendations in relation to recognising dying do not represent a decision between competing alternative courses of action and as such do not carry a direct resource impact. So for example, recommendations on the signs that are common in the last hours or days of life provide information to healthcare professionals but do not in themselves suggest an action or change in management. Nevertheless, if it is thought that a child or young person is likely to die within days or hours then it is likely and appropriate that management would change. While, there is often considerable uncertainty in recognising imminent death, the Committee felt that their recommendations would aide recognition of dying in children or young people and thus promote more effective and cost-effective care whether that involved more timely intervention or withdrawal of treatment.

### 11.8.4 Quality of evidence

The Committee agreed that although the evidence in the study was of low quality, there are some patterns of signs and symptoms that they recognise from their own clinical experience. The Delphi consensus study showed uncertainty regarding the diagnosis of imminent death (none of the symptoms were marked as always present, and very few were identified as very often present). This supports the opinion of the Committee, who pointed out a high level of uncertainty when dealing with children and young people. Another flaw discussed, was that the initial questionnaire used in the study was drawn from literature with adults, and children substantially differ from adults, since they do not normally present with pre-existing organ failure.

In summary, the results are informative, but limited for recommendation making.

### 11.8.5 Other considerations

The signs and symptoms identified in the evidence from the Delphi consensus study were discussed. Even though considered to be low quality the Committee concluded that a guide of the most commonly reported signs and symptoms could be helpful for healthcare professional as well as the people close to the dying child or young person. A number of these were discussed in some detail with regard to the evidence from the Delphi consensus study:

Changes to usual breathing pattern: the Committee decided to include this in the recommendation, but noted that it is important to emphasize that these signs are meaningful only where breathing was previously unaffected.

Loss of interest in or ability to tolerate drinks or food: despite this not being one of the signs reaching consensus in the Delphi, the Committee agreed that deterioration in the ability to absorb feeds is a frequent sign. They also noted that the deterioration in the desire to eat and/or drink can happen whether the child is orally fed or not. The Committee also agreed that reduced urine output was regularly observed when a person is close to death and that this should therefore be highlighted in the recommendation (participants in the Delphi had classified this as a sign and symptom that is often present).

Neurological changes: the Committee decided to include most of the neurological symptoms identified as being very often or often present in the Delphi study. However, unnatural tiredness was removed, as although some members agreed that it is sometimes present, it was argued that this sign is more related to adults. Difficulty in swallowing tablets was also removed. The Committee decided that increasing pain medication and intractable seizures were also important signs and symptoms according to their own experience and the experts' opinion in the Delphi study. They were therefore also added to the bullet point list in the recommendation.

Changes in disease trajectory: the Committee discussed that the need for PICU and having a do not attempt resuscitate are more a consequence than a prognostic factor. They decided against the signs identified in the Delphi study, as they agreed that all of them are inherent to children and young people with a life-limiting condition, not just to the last days of life. The fact that a child or young person is deteriorating does not necessarily mean that the child is going to die.

Intuition of healthcare professionals: the Committee discussed the importance of the clinical judgement of experienced healthcare professionals in recognising when a child is approaching death.

Intuition of the child or young person or their parents of carers: likewise, the Committee discussed the importance of the judgement of the children or their parents or carers, and emphasised the importance of discussing this concern with them.

Finally, the Committee discussed the usefulness of a research recommendation, given the lack of evidence and the high levels of uncertainty. It was suggested that a prospective study could be done by collecting data in hospitals. It was also suggested that specific groups could be looked at separately, such as neonates and children with a neuro-disability.

### 11.8.6 Key conclusions

The guideline Committee concluded that:

The evidence is scarce and of low quality. There may be common signs and symptoms to recognise that someone is in the last days of life. However, there is a lot of uncertainty around this and the Committee highlighted that death may also occur without any particular signs or symptoms to indicate this. It was agreed that a list of frequently observed signs and symptoms could be useful, but it is important to acknowledge that this should not be used as a checklist. It is important to explain to the parents or carers, as well as junior professionals the particular circumstances of the child or young person and be honest about the uncertainty regarding the prognosis with all concerned. The Committee agreed that no inference about levels of interventions should be made on the basis of signs and symptoms.

## 11.9 Recommendations

**128. For children and young people with life-limiting conditions, be aware that:**

- there are various symptoms and signs (individually or in combination) that indicate they may be likely to die within hours or days **and**
- the wider clinical context is also relevant **and**
- there is often some uncertainty about this.

**129. When assessing whether a child or young person is likely to die within hours or days, be aware that the following signs are common in the last hours or days of life, and monitor these non-invasively as far as possible:**

- a change of breathing pattern (for example noisy, laboured or irregular breathing)
- impaired peripheral perfusion (which can be indicated by a pale or grey appearance, or a prolonged capillary refill time), including temperature instability
- loss of interest in or ability to tolerate drinks or food
- a marked and unexplained fall in urine output
- an altered level of awareness (for example reduced consciousness, alertness or responsiveness, excessive sleeping, or confusion)
- intractable seizures that keep occurring even with optimal management
- new onset of profound weakness
- increasing pain and need for analgesia.

**130. When assessing symptoms and signs to decide whether a child or young person is likely to die within hours or days, take into account the wider clinical context, including:**

- their normal clinical baseline
- past clinical events (such as previous episodes of temporary deterioration)
- the overall progression of their condition.

**131. When assessing whether a child or young person is likely to die within hours or days, take into account the clinical judgement of healthcare professionals experienced in end of life care.**

**132. If the child or young person or their parents or carers feel that they are likely to die within hours or days:**

- be aware that they may be correct
- discuss their concerns with them.

**133. When a child or young person is likely to die within hours or days:**

- be aware that they or their parents or carers may not express their feelings openly, and may:
  - have intense and varied feelings such as fear, hopelessness or anger **or**
  - become more accepting of the inevitability of death
- give them and their parents or carers opportunities to talk.

**134. When children and young people become seriously ill and are likely to die within hours or days, provide care as specified in their Advance Care Plan and review if needed.**

**135. If a child or young person may be approaching the end of life and they or their parents or carers want to be involved in making decisions about their care, discuss and review their Advance Care Plan with them.**

**136. When a child or young person is approaching the end of life, discuss with them and their parents or carers and with relevant healthcare professionals:**

- any available invasive treatments that might be in their best interest

- any interventions they are currently receiving that may no longer be in their best interest.

**137. If withdrawing a treatment for a child or young person who is dying, explain to them and to their parents or carers that it is often difficult to tell if or how this may affect them, or when they will die.**

**138. When a child or young person is likely to die within hours or days, ensure that they can have private time with their parents or carers.**

## 11.10 Research recommendations

**9. What signs and symptoms indicate that a child or young person with a life-limiting condition is likely to die within hours or days?**

| Research question | What signs and symptoms indicate that a child or young person with a life-limiting condition is likely to die within hours or days? |
|---|---|
| Why this is needed | |
| Importance to 'patients' or the population | Recent studies show that life expectancy is one of the most influential factors when assessing whether children should be referred to palliative care services, but referrals to such services often occur in the late stages of illness. For CYP and their families/carers receiving palliative care services it is important to be able to provide care based on need as resources are often limited. In order to achieve this it would be useful to know when it is likely that a child/young person (CYP) may die in the next few hours or days. Prospective cohort studies of physical and psychological symptoms experienced by CYPs in the last week of life are needed to ascertain whether there are certain signs and symptoms which are more prevalent during this time. This could be ascertained by undertaking a questionnaire-type study asking families and CYPs (if appropriate) about the incidence of symptoms that may be anticipated at the end of life and asking an open question at the end about the presence of other symptoms. Alternatively this could be achieved by asking professionals to document the symptoms a CYP was experiencing (although this may lead to reporting of the more obvious physical symptoms rather than psychological symptoms). Qualitative studies interviewing experienced palliative care professionals may also be helpful to ascertain what signs and symptoms professionals associate with imminent death. |
| Relevance to NICE guidance | This is a high research priority because there is currently only 1 Delphi consensus study is available to inform clinicians about the signs and symptoms that may help recognise that a child or young person with a life-limiting condition may be in the last days or hours of his or her life. The NIHR has recently funded a study for to compare such tools in the adult population (http://www.nets.nihr.ac.uk/projects/hta/132001). However, dying in the adult population is in the majority of cases due to old age and it could be argued that signs and symptoms of dying in CYP may be different. Given the uncertainties around prognosis, a clearer understanding of this topic would allow healthcare professionals and families to be able to plan ahead and be more informed about the signs and symptoms that they may expect. |
| Relevance to the NHS | Having a tool or set of criteria to help lower the uncertainties around the recognition of CYP who are likely to die in the next few hours or days. This could allow clinicians to avoid invasive interventions that may no longer be in the best interest of the child or young person, plan and deliver services more appropriately and provide a cue to prepare and support the family and CYP (if appropriate) for what may occur over the next few days. Cessation of futile/invasive interventions could result in a cost saving to the NHS. |

| Research question | What signs and symptoms indicate that a child or young person with a life-limiting condition is likely to die within hours or days? |
|---|---|
| | |
| National priorities | Two of the aims of the document Better care, Better lives (Department of Health, 2008) for children with life-limiting conditions were to:<br>• "ensure that all children have a choice on location of care, 24-hour access to multidisciplinary community teams and, when needed, specialist palliative care advice and services."<br>• Have "Access to specialist end-of-life care and 24-hour support and advice should be available."<br><br>This means that being able to better recognise when a child or young person is likely to die will help planning where to care for them in line with their own and their parents' / carers preferences. |
| Current evidence base | Currently there is 1 modified Delphi survey of 49 paediatric palliative care professionals asking them to identify signs and symptoms that indicate that a CYP is moving to an end of life phase. This survey was found to be of low methodological quality. There are no identified prospective or retrospective cohort studies aiming to answer this question. |
| Equality | There are currently geographical inequalities in the way that services are set out and how they support choices of places of care and death. Better recognition of the likely time a child has left to live may improve service provision and facilitate the child to be cared for and die in their preferred place. |
| Feasibility | A prospective or retrospective cohort study is feasible in this population. Involvement of CYP and their families in prospective data collection would need ethical approval, and with adequate patient and public involvement at the design stage this should be achievable. Due to small numbers of children requiring palliative care services any such study may need to be multicentred to achieve an adequate sample size. As it is impossible to know when a CYP is in the last few days/weeks of life data collection would need to begin at referral to the palliative care team or at a stage when it was felt the CYP may be deteriorating. This may mean that CYPs were enrolled on a prospective study for a long period of time which may be a burden to them and their family. There are currently no validated prognostic tools in this population so the data collection tool would need to ask about well-known end of life signs and symptoms and include an open question at the end to capture anything missed.<br><br>A qualitative interview study with experienced paediatric palliative care professionals would also be feasible and relatively straightforward to achieve. |
| Other comments | Funding for these studies may be difficult to secure as often funders look at the number of people a study will benefit. The number of children/young people requiring palliative care services are relatively small so the study would have less of an impact than one for a larger population. |

# 12 References

**Aho et al., 2011**

Aho, A. L., Tarkka, M.-T., Astedt-Kurki, P., Sorvari, L., Kaunonen, M., Evaluating a bereavement follow-up intervention for grieving fathers and their experiences of support after the death of a child-a pilot study, Death Studies, 35, 879-904, 2011

**Anghelescu et al., 2005**

Anghelescu, D. L., Burgoyne, L. L., Oakes, L. L., Wallace, D. A., The safety of patient-controlled analgesia by proxy in pediatric oncology patients, Anesthesia and Analgesia, 101, 1623-1627, 2005

**Arland et al., 2013**

Arland, L. C., Hendricks-Ferguson, V. L., Pearson, J., Foreman, N. K., Madden, J. R., Development of an in-home standardized end-of-life treatment program for pediatric patients dying of brain tumors, Journal for Specialists in Pediatric Nursing: JSPN, 18, 144-57, 2013

**ACT & RCPCH, 1997**

Association for Children with Life Threatening or Terminal Conditions and their Families and the Royal College of Paediatrics and Child Health. A guide to the development of children's palliative care services. Report of the joint working party. Bristol, UK, 1997.

**Barnes et al., 2016**

Barnes, H., McDonald, J., Smallwood, N., Manser, R., Opioids for the palliation of refractory breathlessness in adults with advanced disease and terminal illness, Cochrane Database of Systematic Reviews, 2016

**Bauer et al., 2011**

Bauer, K., Skoetz, N., Monsef, I., Engert, A., Brillant, C., Comparison of chemotherapy including escalated BEACOPP versus chemotherapy including ABVD for patients with early unfavourable or advanced stage Hodgkin lymphoma, Cochrane Database of Systematic Reviews, CD007941, 2011

**Baverstock & Finlay, 2008**

Baverstock, A., Finlay, F., What can we learn from the experiences of consultants around the time of a child's death?, Child: Care, Health & Development, 34, 732-9, 2008

**Beecham et al., 2015**

Beecham, E., Candy, B., Howard, R., McCulloch, R., Laddie, J., Rees, H., Vickerstaff, V., Bluebond-Langner, M., Jones, L., Pharmacological interventions for pain in children and adolescents with life-limiting conditions, Cochrane Database of Systematic Reviews, 3, CD010750, 2015

**Bluebond-Langner et al., 2013**

Bluebond-Langner, M., Beecham, E., Candy, B., Langner, R., Jones, L., Preferred place of death for children and young people with life-limiting and life-threatening conditions: a systematic review of the literature and recommendations for future inquiry and policy, Palliative Medicine, 27, 705-13, 2013

**Boss et al., 2008**

Boss, R. D., Hutton, N., Sulpar, L. J., West, A. M., Donohue, P. K., Values parents apply to decision-making regarding delivery room resuscitation for high-risk newborns, Pediatrics, 122, 583-589, 2008

**Bradford et al., 2013**

Bradford, N., Armfield, N. R., Young, J., Smith, A. C., The case for home based telehealth in pediatric palliative care: A systematic review, BMC Palliative Care, 12, 2013

**Bradt & Dileo, 2010**

Bradt, J., Dileo, C., Music therapy for end-of-life care, Cochrane Database of Systematic Reviews, CD007169, 2010

**Branchett & Stretton, 2012**

Branchett, K., Stretton, J., Neonatal palliative and end of life care: What parents want from professionals, Journal of Neonatal Nursing, 18, 40-44, 2012

**Brosig et al., 2007**

Brosig, C. L., Pierucci, R. L., Kupst, M. J., Leuthner, S. R., Infant end-of-life care: the parents' perspective, Journal of Perinatology, 27, 510-6, 2007

**Brown & Sourkes, 2006**

Brown, M. R., Sourkes, B., Psychotherapy in pediatric palliative care, Child & Adolescent Psychiatric Clinics of North America, 15, 585-96, viii, 2006

**Byrne et al., 2011**

Byrne, M., Tresgallo, M., Saroyan, J., Granowetter, L., Valoy, G., Schechter, W., Qualitative Analysis of Consults by a Pediatric Advanced Care Team During Its First Year of Service, American Journal of Hospice & Palliative Medicine, 28, 109-118, 2011

**Cadell et al., 2012**

Cadell, S., Kennedy, K., Hemsworth, D., Informing social work practice through research with parent caregivers of a child with a life-limiting illness, Journal Of Social Work In End-Of-Life & Palliative Care, 8, 356-81, 2012

**Caeymaex et al., 2011**

Caeymaex, L., Speranza, M., Vasilescu, C., Danan, C., Bourrat, M. M., Garel, M., Jousselme, C., Living with a crucial decision: a qualitative study of parental narratives three years after the loss of their newborn in the NICU, PLoS ONE [Electronic Resource], 6, e28633, 2011

**Candy et al., 2012**

Candy, B., Jones, L., Varagunam, M., Speck, P., Tookman, A., King, M., Spiritual and religious interventions for well-being of adults in the terminal phase of disease, Cochrane Database of Systematic Reviews, 5, CD007544, 2012

**Champagne & Mongeau, 2012**

Champagne, M., Mongeau, S., Effects of respite care services in a children's hospice: the parents' point of view, Journal of Palliative Care, 28, 245-51, 2012

**Contro & Sourkes, 2012**

Contro, N., Sourkes, B. M., Opportunities for quality improvement in bereavement care at a children's hospital: assessment of interdisciplinary staff perspectives, Journal of Palliative Care, 28, 28-35, 2012

**Contro et al., 2002**

Contro, N., Larson, J., Scofield, S., Sourkes, B., Cohen, H., Family perspectives on the quality of pediatric palliative care, Archives of Pediatrics & Adolescent Medicine, 156, 14-9, 2002

**Contro et al., 2004**

Contro, N. A., Larson, J., Scofield, S., Sourkes, B., Cohen, H. J., Hospital staff and family perspectives regarding quality of pediatric palliative care, Pediatrics, 114, 1248-52, 2004

**Curtis & Burns, 2015**

Curtis, L., Burns, A., Unit Costs of Health and Social Care 2015. PSSRU, University of Kent, 2015

**Davies & Connaughty, 2002**

Davies, B., Connaughty, S., Pediatric end-of-life care: lessons learned from parents, Journal of Nursing Administration, 32, 5-6, 2002

**Davies et al., 2003**

Davies, R., Davis, B., Sibert, J., Parents' stories of sensitive and insensitive care by paediatricians in the time leading up to and including diagnostic disclosure of a life-limiting condition in their child, Child: Care, Health and Development, 29, 77-82, 2003

**Davies et al., 2004**

Davies, B., Steele, R., Collins, J. B., Cook, K., Smith, S., The impact on families of respite care in a children's hospice program, Journal of Palliative Care, 20, 277-86, 2004

**Davies et al., 2010**

Davies, B., Contro, N., Larson, J., Widger, K., Culturally-sensitive information-sharing in pediatric palliative care, Pediatrics, 125, e859-65, 2010

**Davies, 1996**

Davies, H., Living with dying: families coping with a child who has a neurodegenerative genetic disorder, AXON, 18, 38-44, 1996

**deCinque et al., 2006**

deCinque, N., Monterosso, L., Dadd, G., Sidhu, R., Macpherson, R., Aoun, S., Bereavement support for families following the death of a child from cancer: experience of bereaved parents, Journal of Psychosocial Oncology, 24, 65-83, 2006

**deJong-Berg & Kane, 2006**

deJong-Berg, M. A., Kane, L., Bereavement care for families part 2: Evaluation of a paediatric follow-up programme, International Journal of Palliative Nursing, 12, 484-94, 2006

**Department of Health, 2008**

Department of Health. End of Life Care Strategy: promoting high quality care for adults at the end of their life. UK, 2008.

**Department of Health, 2013**

Department of Health. More care, less pathway: a review of the Liverpool Care Pathway. UK, 2013.

**de Sa Franca et al., 2013**

de Sa Franca, J. R., da Costa, S. F., Lopes, M. E., da Nobrega, M. M., de Franca, I. S., The importance of communication in pediatric oncology palliative care: Focus on humanistic nursing theory, Revista Latino-Americana de Enfermagem, 21, 780-786, 2013

**Diamond et al., 2014**

Diamond, I.R., Grant, R.C., Feldman, B.M., Pencharz, P.B., Ling, S.C., Moore, A.M., Wales, P.W., Defining consensus: a systematic review recommends methodologic criteria for reporting of Delphi studies, Journal of Clinical Epidemiology, 67, 401-9, 2014.

**Dixon-Woods et al., 2005**

Dixon-Woods, M., Agarwal, S., Jones, D., Young, B., Sutton, A., Synthesising qualitative and quantitative evidence: a review of possible methods, Journal of Health Services & Research Policy, 10, 45-53, 2005

**Dunsmore, 1996**

Dunsmore, J, Information, Support, and Decision-Making Needs and Preferences of Adolescents with Cancer: Implications for Health Professionals, Journal of Psychosocial Oncology, 13, 39-56, 1996

**Eaton, 2008**

Eaton, N., 'I don't know how we coped before': a study of respite care for children in the home and hospice, Journal of Clinical Nursing, 17, 3196-3205, 2008

**Ebmeier et al., 1991**

Ebmeier, C., Lough, M. A., Huth, M. M., Autio, L., Hospitalized school-age children express ideas, feelings, and behaviors toward God, Journal of Pediatric Nursing, 6, 337-49, 1991

**Eccleston et al., 2015**

Eccleston, C., Fisher, E., Law, E., Bartlett, J., Palermo, T., Psychological interventions for parents of children and adolescents with chronic illness, Cochrane Database of Systematic Reviews, 2015

**Einaudi et al., 2010**

Einaudi, M. A., Le Coz, P., Malzac, P., Michel, F., D'Ercole, C., Gire, C., Parental experience following perinatal death: exploring the issues to make progress, European Journal of Obstetrics, Gynecology, & Reproductive Biology, 151, 143-8, 2010

**ElSayed et al., 2013**

El Sayed, M. F., Chan, M., McAllister, M., Hellmann, J., End-of-life care in Toronto neonatal intensive care units: challenges for physician trainees, Archives of Disease in Childhood Fetal & Neonatal Edition, 98, F528-33, 2013

**Erby et al., 2006**

Erby, L. H., Rushton, C., Geller, G., "My son is still walking": stages of receptivity to discussions of advance care planning among parents of sons with Duchenne muscular dystrophy, Seminars in Pediatric Neurology, 13, 132-40, 2006

**Fellowes et al., 2004**

Fellowes, D., Barnes, K., Wilkinson, S., Aromatherapy and massage for symptom relief in patients with cancer, Cochrane Database of Systematic Reviews, CD002287, 2004

**Forbes et al., 2008**

Forbes, T., Goeman, E., Stark, Z., Hynson, J., Forrester, M., Discussing withdrawing and withholding of life-sustaining medical treatment in a tertiary paediatric hospital: a survey of clinician attitudes and practices, Journal of Paediatrics & Child Health, 44, 392-8, 2008

**Forrester, 2008**

Forrester, L., Bereaved parents' experiences of the use of 'cold bedrooms' following the death of their child, International Journal of Palliative Nursing, 14, 578-85, 2008

**Forster & Windsor, 2014**

Forster, M., Windsor, C., Speaking to the deceased child: Australian health professional perspectives in paediatric end-of-life care, International Journal of Palliative Nursing, 20, 502-509, 2014

**Foster et al., 2009**

Foster, T. L., Gilmer, M. J., Davies, B., Barrera, M., Fairclough, D., Vannatta, K., Gerhardt, C. A., Bereaved parents' and siblings' reports of legacies created by children with cancer, Journal of Pediatric Oncology Nursing, 26, 369-76, 2009

**Fraser et al., 2012**

Fraser, L. K., Miller, M., Hain, R., Norman, P., Aldridge, J., McKinney, P. A., Parslow, R. C., Rising national prevalence of life-limiting conditions in children in England, Pediatrics, 129, e923-9, 2012

**Friedrichsdorf et al., 2015**

Friedrichsdorf, S. J., Postier, A., Dreyfus, J., Osenga, K., Sencer, S., Wolfe, J., Improved quality of life at end of life related to home-based palliative care in children with cancer, Journal of Palliative Medicine, 18, 143-150, 2015

**Gaab et al., 2013**

Gaab, E. M., Glynn Owens, R., MacLeod, R. D., The voices of young new zealanders involved in pediatric palliative care, Journal of Palliative Care, 29, 186-192, 2013

**Good et al., 2014**

Good, P., Richard, R., Syrmis, W., Jenkins-Marsh, S., Stephens, J., Medically assisted nutrition for adult palliative care patients, Cochrane Database of Systematic Reviews, 4, CD006274, 2014

**Gordon et al., 2009**

Gordon, C., Barton, E., Meert, K. L., Eggly, S., Pollacks, M., Zimmerman, J., Anand, K. J., Carcillo, J., Newth, C. J., Dean, J. M., Willson, D. F., Nicholson, C., Accounting for medical communication: parents' perceptions of communicative roles and responsibilities in the pediatric intensive care unit, Communication & Medicine, 6, 177-188, 2009

**Grinyer et al., 2010**

Grinyer, A., Payne, S., Barbarachild, Z., Issues of power, control and choice in children's hospice respite care services: a qualitative study, International Journal of Palliative Nursing, 16, 505-10, 2010

**Groh et al., 2013**

Groh, G., Borasio, G. D., Nickolay, C., Bender, H. U., von Luttichau, I., Fuhrer, M., Specialized pediatric palliative home care: a prospective evaluation, Journal of Palliative Medicine, 16, 1588-94, 2013

**Hammes et al., 2005**

Hammes, B. J., Klevan, J., Kempf, M., Williams, M. S., Pediatric advance care planning, Journal of Palliative Medicine, 8, 766-73, 2005

**Hechler et al., 2008**

Hechler, T., Blankenburg, M., Friedrichsdorf, S. J., Garske, D., Hubner, B., Menke, A., Wamsler, C., Wolfe, J., Zernikow, B., Parents' perspective on symptoms, quality of life, characteristics of death and end-of-life decisions for children dying from cancer, Klinische Padiatrie, 220, 166-74, 2008

**Hendricks-Ferguson, 2007**

Hendricks-Ferguson, V. L., Parental perspectives of initial end-of-life care communication, International Journal of Palliative Nursing, 13, 522-31, 2007

**Hexem et al., 2011**

Hexem, K. R., Mollen, C. J., Carroll, K., Lanctot, D. A., Feudtner, C., How parents of children receiving pediatric palliative care use religion, spirituality, or life philosophy in tough times, Journal of Palliative Medicine, 14, 39-44, 2011

**Higgins & Green, 2008**

Higgins, J.P.T., Green, S. (editors). The Cochrane Handbook for systematic reviews of interventions. The Cochrane Collaboration & Wiley, Chichester, 2008.

**Hinds et al., 2000**

Hinds, P. S., Oakes, L., Quargnenti, A., Furman, W., Bowman, L., Gilger, E., Gattuso, J., Martinson, I., Yi, K. H., Drew, D., An international feasibility study of parental decision making in pediatric oncology, Oncology Nursing Forum, 27, 1233-43, 2000

**Hinds et al., 2005**

Hinds, P. S., Drew, D., Oakes, L. L., Fouladi, M., Spunt, S. L., Church, C., Furman, W. L., End-of-life care preferences of pediatric patients with cancer, Journal of Clinical Oncology, 23, 9146-54, 2005

**Hoover et al., 2014**

Hoover, S. M., Bratton, S. L., Roach, E., Olson, L. M., Parental experiences and recommendations in donation after circulatory determination of death, Pediatric Critical Care Medicine, 15, 105-11, 2014

**Hsiao et al., 2007**

Hsiao, J. L., Evan, E. E., Zeltzer, L. K., Parent and child perspectives on physician communication in pediatric palliative care, Palliative & Supportive Care, 5, 355-65, 2007

**Hughes-Hallet et al., 2011**

Hughes-Hallet, T., Craft, A., Davies, C., Mackay, I., Nielsson, T., Palliative care funding review: funding the right care and support for everyone. Department of Health, London, 2011.

**Hunt et al., 2001**

Hunt, A., Goldman, A., Devine, T., Phillips, M., Transdermal fentanyl for pain relief in a paediatric palliative care population, Palliative Medicine, 15, 405-412, 2001

**Hunt et al., 2013**

Hunt, A., Coad, J., West, E., Hex, N., Staniszewska, S., Hacking, S., Farman, M., Brown, E., Owens, C., Ashley, N., Kaur, J., May, K., Chandler, V., Barron, D., Wik, A., Magee, H., Lowson, K., Wright, D., Gunn, K., Kelly, K., Woodhead, S., Together for Short Lives, The Big Study for Life-limited Children and their Families – Final research report , 2013

**James & Johnson, 1997**

James, L., Johnson, B., The needs of parents of pediatric oncology patients during the palliative care phase, Journal of Pediatric Oncology Nursing, 14, 83-95, 1997

**Jennings & Nicholl, 2014**

Jennings, V., Nicholl, H., Bereavement support used by mothers in Ireland following the death of their child from a life-limiting condition, International Journal of Palliative Nursing, 20, 173-8, 2014

**Jones, 2006**

Jones, B. L., Companionship, control, and compassion: a social work perspective on the needs of children with cancer and their families at the end of life, Journal of Palliative Medicine, 9, 774-88, 2006

**Kassam et al., 2014**

Kassam, A., Skiadaresis, J., Alexander, S., Wolfe, J., Parent and clinician preferences for location of end-of-life care: home, hospital or freestanding hospice?, Pediatric Blood & Cancer, 61, 859-64, 2014

**Kavanaugh et al., 2010**

Kavanaugh, K., Moro, T. T., Savage, T. A., How nurses assist parents regarding life support decisions for extremely premature infants, JOGNN – Journal of Obstetric, Gynecologic, and Neonatal Nursing, 39, 147-158, 2010

**Konrad, 2007**

Konrad, S. C., What parents of seriously ill children value: parent-to-parent connection and mentorship, Omega – Journal of Death & Dying, 55, 117-30, 2007

**Laakso & Paunonen-Ilmonen, 2001**

Laakso, H., Paunonen-Ilmonen, M., Mothers' grief following the death of a child, Journal of Advanced Nursing, 36, 69-77, 2001

**Laakso & Paunonen-Ilmonen, 2002**

Laakso, H., Paunonen-Ilmonen, M., Mothers' experience of social support following the death of a child, Journal of Clinical Nursing, 11, 176-85, 2002

**Lewin et al., 2015**

Lewin, S., Glenton, C., Munthe-Kaas, H., Carlsen, B., Colvin, C. J., Gulmezoglu, M., Noyes, J., Booth, A., Garside, R., Rashidian, A., Using qualitative evidence in decision making for health and social interventions: an approach to assess confidence in findings from qualitative evidence syntheses (GRADE-CERQual), PLoS Medicine / Public Library of Science, 12, e1001895, 2015

**Lotz et al., 2015**

Lotz, J. D., Jox, R. J., Borasio, G. D., Fuhrer, M., Pediatric advance care planning from the perspective of healthcare professionals: A qualitative interview study, Palliative Medicine, 29, 212-222, 2015

**Lowson et al., 2007**

Lowson, K., Lowson, P., Duffy, S., Independent review of the palliative care for children and young people: economic study (final report). Department of Health Independent Review team: York Health Economics Consortium, 2007.

**Lundqvist et al., 2002**

Lundqvist, A., Nilstun, T., Dykes, A.-K., Both empowered and powerless: Mothers' experiences of professional care when their newborn dies, Birth: Issues in Perinatal Care, 29, 192-199, 2002

**Lundqvist et al., 2003**

Lundqvist, A., Nilstun, T., Dykes, A., Neonatal end-of-life care in Sweden: the views of Muslim women, Journal of Perinatal & Neonatal Nursing, 17, 77-87, 2003

**Macmillan, 2010**

Macmillan Cancer Support. Always there? The Impact of the End of Life Care Strategy on 24/7 community nursing in England. UK, 2010

**Malcolm et al., 2008**

Malcolm, C., Forbat, L., Knighting, K., Kearney, N., Exploring the experiences and perspectives of families using a children's hospice and professionals providing hospice care to identify future research priorities for children's hospice care, Palliative Medicine, 22, 921-8, 2008

**Maynard et al., 2005**

Maynard, L., Rennie, T., Shirtliffe, J., Vickers, D., Seeking and using families' views to shape children's hospice services, International Journal of Palliative Nursing, 11, 624-30, 2005

**McHaffie et al., 2001**

McHaffie, H. E., Lyon, A. J., Hume, R., Deciding on treatment limitation for neonates: the parents' perspective, European Journal of Pediatrics, 160, 339-44, 2001

**McQuay et al., 2013**

McQuay, H. J., Collins, S. L., Carroll, D., Moore, R. A., Derry, S., Radiotherapy for the palliation of painful bone metastases, Cochrane Database of Systematic Reviews, 11, CD001793, 2013

**Meert et al., 2005**

Meert, K. L., Thurston, C. S., Briller, S. H., The spiritual needs of parents at the time of their child's death in the pediatric intensive care unit and during bereavement: a qualitative study, Pediatric Critical Care Medicine, 6, 420-7, 2005

**Meert et al., 2007**

Meert, K. L., Eggly, S., Pollack, M., Anand, K. J., Zimmerman, J., Carcillo, J., Newth, C. J., Dean, J., Willson, D. F., Nicholson, C., Parents' perspectives regarding a physician-parent conference after their child's death in the pediatric intensive care unit, The Journal of Pediatrics, 151, 50-55, 2007

**Meert et al., 2008**

Meert, K. L., Eggly, S., Pollack, M., Anand, K. J., Zimmerman, J., Carcillo, J., Newth, C. J., Dean, J. M., Willson, D. F., Nicholson, C., National Institute of Child, Health, Human Development Collaborative Pediatric Critical Care Research, Network, Parents' perspectives on physician-parent communication near the time of a child's death in the pediatric intensive care unit, Pediatric Critical Care Medicine, 9, 2-7, 2008

**Meyer et al., 2006**

Meyer, E. C., Ritholz, M. D., Burns, J. P., Truog, R. D., Improving the quality of end-of-life care in the pediatric intensive care unit: parents' priorities and recommendations, Pediatrics, 117, 649-57, 2006

**Michelson et al., 2013**

Michelson, K. N., Patel, R., Haber-Barker, N., Emanuel, L., Frader, J., End-of-life care decisions in the PICU: roles professionals play, Pediatric Critical Care Medicine, 14, e34-44, 2013

**Midson & Carter, 2010**

Midson, R., Carter, B., Addressing end of life care issues in a tertiary treatment centre: lessons learned from surveying parents' experiences, Journal of Child Health Care, 14, 52-66, 2010

**Mitchell & Dale, 2015**

Mitchell, S., Dale, J., Advance Care Planning in palliative care: A qualitative investigation into the perspective of Paediatric Intensive Care Unit staff, Palliative Medicine, 29, 371-379, 2015

**Monterosso et al., 2007**

Monterosso, L., Kristjanson, L. J., Aoun, S., Phillips, M. B., Supportive and palliative care needs of families of children with life-threatening illnesses in Western Australia: evidence to guide the development of a palliative care service, Palliative Medicine, 21, 689-96, 2007

**Murray et al., 2000**

Murray, J. A., Terry, D. J., Vance, J. C., Battistutta, D., Connolly, Y., Effects of a program of intervention on parental distress following infant death, Death Studies, 24, 275-305, 2000

**NAO, 2008**

National Audit Office. End of Life Care. UK, 2008

**Nolbris & Hellström, 2005**

Nolbris, M., Hellström, A., Siblings' needs and issues when a brother or sister dies of cancer, Journal of Pediatric Oncology Nursing, 22, 227-234, 2005

**ONS, 2007**

Office for National Statistics (2007a) Mortality Statistics: childhood, infant and perinatal, England and Wales, 2005. Series DH3 no.38 and Office for National Statistics (2007d)

Review of the Registrar General on deaths in England and Wales: mortality cause, 2005. Series DH2 no.32

**Parker et al., 1999**

Parker, D., Maddocks, I., Stern, L. M., The role of palliative care in advanced muscular dystrophy and spinal muscular atrophy, Journal of Paediatrics & Child Health, 35, 245-50, 1999

**Pearson, 2013**

Pearson, H. N., "You've only got one chance to get it right": Children's cancer nurses' experiences of providing palliative care in the acute hospital setting, Issues in Comprehensive Pediatric Nursing, 36, 188-211, 2013

**Postier et al., 2014**

Postier, A., Chrastek, J., Nugent, S., Osenga, K., Friedrichsdorf, J., Exposure to Home-Based Pediatric Palliative and Hospice Care and Its Impact on Hospital and Emergency Care Charges at a Single Institution, Journal of Palliative Medicine, 17, 183-189, 2014

**Price et al., 2011**

Price, J., Jordan, J., Prior, L., Parkes, J., Living through the death of a child: a qualitative study of bereaved parents' experiences, International Journal of Nursing Studies, 48, 1384-92, 2011

**Price et al., 2013**

Price, J., Jordan, J., Prior, L., A consensus for change: parent and professional perspectives on care for children at the end-of-life, Issues in Comprehensive Pediatric Nursing, 36, 70-87, 2013

**Reder & Serwint, 2009**

Reder, E. A., Serwint, J. R., Until the last breath: exploring the concept of hope for parents and healthcare professionals during a child's serious illness, Archives of Pediatrics & Adolescent Medicine, 163, 653-7, 2009

**Redmond & Richardson, 2003**

Redmond, B., Richardson, V., Just Getting on with it: Exploring the Service Needs of Mothers Who Care for Young Children with Severe/Profound and Life-Threatening Intellectual Disability, Journal of Applied Research in Intellectual Disabilities, 16, 205-218, 2003

**Remedios et al., 2015**

Remedios, C., Willenberg, L., Zordan, R., Murphy, A., Hessel, G., Philip, J., A pre-test and post-test study of the physical and psychological effects of out-of-home respite care on caregivers of children with life-threatening conditions, Palliative Medicine, 29, 223-30, 2015

**Rini & Loriz, 2007**

Rini, A., Loriz, L., Anticipatory mourning in parents with a child who dies while hospitalized, Journal of Pediatric Nursing, 22, 272-82, 2007

**Robert et al., 2012**

Robert, R., Zhukovsky, D. S., Mauricio, R., Gilmore, K., Morrison, S., Palos, G. R., Bereaved parents' perspectives on pediatric palliative care, Journal Of Social Work In End-Of-Life & Palliative Care, 8, 316-38, 2012

**Robinson et al., 2006**

Robinson, M. R., Thiel, M. M., Backus, M. M., Meyer, E. C., Matters of spirituality at the end of life in the pediatric intensive care unit, Pediatrics, 118, e719-29, 2006

**Rosner et al., 2010**

Rosner, R., Kruse, J., Hagl, M., A meta-analysis of interventions for bereaved children and adolescents, Death Studies, 34, 99-136, 2010

**Ruggiero et al., 2007**

Ruggiero, A., Barone, G., Liotti, L., Chiaretti, A., Lazzareschi, I., Riccardi, R., Safety and efficacy of fentanyl administered by patient controlled analgesia in children with cancer pain, Supportive Care in Cancer, 15, 569-73, 2007

**Sackett et al., 1992**

Sackett, D , Hayes, R , Guyatt, G, Tugwell P, Clinical Epidemiology: A Basic Science for Clinical Medicine, 1992

**Schiessl et al., 2008**

Schiessl, C., Gravou, C., Zernikow, B., Sittl, R., Griessinger, N., Use of patient-controlled analgesia for pain control in dying children, Supportive Care in Cancer, 16, 531-6, 2008

**Schmidt-Hansen et al., 2015**

Schmidt-Hansen, M., Bromham, N., Taubert, M., Arnold, S., Hilgart, J. S., Buprenorphine for treating cancer pain, Cochrane Database of Systematic Reviews, 3, CD009596, 2015

**Shaw et al., 2014**

Shaw, K. L., Brook, L., Cuddeford, L., Fitzmaurice, N., Thomas, C., Thompson, A., Wallis, M., Prognostic indicators for children and young people at the end of life: A Delphi study, Palliative Medicine, 28, 501-512, 2014

**Sheridan et al., 2008**

Sheridan, J , Craig, F , McCulloch, R , Harrop, E , Comac, M , Boggs, T , Hemsley, J, Hitchin, N , Gilder, R, Can palliative care be done by phone? A telephone audit of a palliative care service, Archives of Disease in Childhood, 93, A63, 2008

**Simon et al., 2011**

Simon, S. T., Higginson, I. J., Booth, S., Harding, R., Bausewein, C., Benzodiazepines for the relief of breathlessness in advanced malignant and non-malignant diseases in adults, Cochrane Database of Systematic Reviews, 2011

**Spathis et al., 2012**

Spathis, A., Harrop, E., Robertshaw, C., Elverson, J., Lapwood, S., Learning from paediatric palliative care: lessons for adult practice, Palliative Medicine, 26, 777-9, 2012

**Stanton et al., 2013**

Stanton, T. R., Wand, B. M., Carr, D. B., Birklein, F., Wasner, G. L., O'Connell, N. E., Local anaesthetic sympathetic blockade for complex regional pain syndrome, Cochrane Database of Systematic Reviews, 8, CD004598, 2013

**Steele et al., 2013**

Steele, A. C., Kaal, J., Thompson, A. L., Barrera, M., Compas, B. E., Davies, B., Fairclough, D. L., Foster, T. L., Jo Gilmer, M., Hogan, N., Vannatta, K., Gerhardt, C. A., Bereaved parents and siblings offer advice to healthcare providers and researchers, Journal of Pediatric Hematology/Oncology, 35, 253-9, 2013

**Steele et al., 2008**

Steele, R., Derman, S., Cadell, S., Davies, B., Siden, H., Straatman, L., Families' transition to a Canadian paediatric hospice. Part two: results of a pilot study, International Journal of Palliative Nursing, 14, 287-95, 2008

**Stenekes et al., 2014**

Stenekes, S. J., Ens, C. D. L., Harlos, M., Chochinov, H. M., Mytopher, K., A Descriptive Study Evaluating Perinatal Healthcare Providers' Perspectives of Palliative Programming in 3 Canadian Institutions, Journal of Perinatal & Neonatal Nursing, 28, 280-290, 2014

**Stevens et al., 2015**

Stevens, R., Macbeth, F., Toy, E., Coles, B., Lester, J. F., Palliative radiotherapy regimens for patients with thoracic symptoms from non-small cell lung cancer, Cochrane Database of Systematic Reviews, 2015

**Sullivan et al., 2014**

Sullivan, J., Monagle, P., Gillam, L., What parents want from doctors in end-of-life decision-making for children, Archives of Disease in Childhood, 99, 216-20, 2014

**Talbot, 1996**

Talbot, K., Transcending a devastating loss: the life attitude of mothers who have experienced the death of their only child... co-published simultaneously in Bereavement: Client Adaptation and Hospice Services (ed: Donna Lind Infeld, and Nadine Reimer Penner), Hospice Journal, 11, 67-83, 1996

**Temel et al., 2010**

Temel, J. S., Greer, J. A., Muzikansky, A., Gallagher, E. R., Admane, S., Jackson, V. A., Dahlin, C. M., Blinderman, C. D., Jacobsen, J., Pirl, W. F., Billings, J. A., Lynch, T. J., Early palliative care for patients with metastatic non-small-cell lung cancer, New England Journal of Medicine, 363, 733-42, 2010

**Truog et al., 2006**

Truog, R. D., Meyer, E. C., Burns, J. P., Toward interventions to improve end-of-life care in the pediatric intensive care unit, Critical Care Medicine, 34, S373-9, 2006

**Vickers et al., 2007**

Vickers, J., Thompson, A., Collins, G. S., Childs, M., Hain, R., Place and provision of palliative care for children with progressive cancer: A study by the paediatric oncology nurses' forum/United Kingdom children's cancer study group palliative care working group, Journal of Clinical Oncology, 25, 4472-4476, 2007

**Weidner et al., 2011**

Weidner, N. J., Cameron, M., Lee, R. C., McBride, J., Mathias, E. J., Byczkowski, T. L., End-of-life care for the dying child: what matters most to parents, Journal of Palliative Care, 27, 279-287, 2011

**Wiffen et al., 2016**

Wiffen, P. J., Wee, B., Moore, A. R., Oral morphine for cancer pain, Cochrane Database of Systematic Reviews, 2016

**Wocial, 2000**

Wocial, L. D., Life support decisions involving imperiled infants, Journal of Perinatal & Neonatal Nursing, 14, 73-86, 2000

**Wolfe et al., 2014**

Wolfe, I., Macfarlane, A., Donkin, A., Marmot, M., Viner, R., Why children die: death in infants, children and young people in the UK. Part A. Royal College of Paediatrics and Child Health; National Children's Bureau and British Association for Child and Adolescent Public Health. UK, 2014

**Wood et al., 2010**

Wood, F., Simpson, S., Barnes, E., Hain, R., Disease trajectories and ACT/RCPCH categories in paediatric palliative care, Palliative Medicine, 24, 796-806, 2010

**Woolley et al., 1989**

Woolley, H., Stein, A., Forrest, G. C., Baum, J. D., Imparting the diagnosis of life threatening illness in children, BMJ, 298, 1623-6, 1989

**WHO, 1998**

World Health Organisation. Cancer pain relief and palliative care in children. Geneva, 1998.

**WHO, 2012**

World Health Organisation. WHO guidelines on the pharmacological treatment of persisting pain in children with medical illnesses. Geneva, 2012.

**Xafis et al., 2015**

Xafis, V., Wilkinson, D., Sullivan, J., What information do parents need when facing end-of-life decisions for their child? A meta-synthesis of parental feedback, BMC Palliative Care, 14, 19, 2015

**Yuen et al., 2012**

Yuen, W. Y., Duipmans, J. C., Jonkman, M. F., The needs of parents with children suffering from lethal epidermolysis bullosa, British Journal of Dermatology, 167, 613-8, 2012

**Zelcer et al., 2010**

Zelcer, S., Cataudella, D., Cairney, A. E., Bannister, S. L., Palliative care of children with brain tumors: a parental perspective, Archives of Pediatrics & Adolescent Medicine, 164, 225-30, 2010

**Zernikow et al., 2009**

Zernikow, B., Michel, E., Craig, F., Anderson, B. J., Pediatric palliative care: use of opioids for the management of pain, Paediatric Drugs, 11, 129-51, 2009

**Zwaanswijk et al., 2007**

Zwaanswijk, M., Tates, K., van Dulmen, S., Hoogerbrugge, P. M., Kamps, W. A., Bensing, J. M., Young patients', parents', and survivors' communication preferences in paediatric oncology: results of online focus groups, BMC Pediatrics, 7, 35, 2007

# 13 Glossary and abbreviations

## 13.1 Glossary

**Table 105: Glossary terms**

| Term | Definition |
|---|---|
| A priori | Reasoning or knowledge from theoretical deduction, as opposed to from observation or experience. |
| Abstract | Summary of a study, which may be published alone or as an introduction to a full scientific paper. |
| Acute pain | Pain of short duration, usually no more than 6 months, which disappears once the underlying cause has healed or been treated |
| Advance care plan | A record of a discussion between and individual (where possible), their care givers and those close to them and their future care plans; forming a core element of their palliative care planning |
| Analgesia | Pain relief |
| Ancillary support | This can include support such as hospice or home care, or include therapeutic services such a physical or nutrition therapy |
| Anorexia | Loss of appetite |
| Antenatal | Before birth |
| Anti-epileptic medications | Medicines to prevent seizures |
| Approaching end of life | Phase of illness following a recognised change in the underlying disease process making it likely that a person will die on a timescale measured in weeks or short months |
| Arm (of a clinical study) | Subsection of individuals within a study who receive a particular intervention, for example placebo arm. |
| Aspirates | Fluid from the lungs |
| Association | Statistical relationship between 2 or more events, characteristics or other variables. The relationship may or may not be causal. |
| Assumed risk | Known exposure to a hazard or procedure |
| Attrition bias | Systematic differences between comparison groups for withdrawal or exclusion of participants from a study. |
| Autopsy | Post mortem examination of the body to understand the cause of death |
| Available case analysis (ACA) | Analysis of data that is available for participants at the end of follow-up. |
| Baseline | The initial set of measurements at the beginning of a study (after run-in period where applicable) with which subsequent results are compared. |
| Before-and-after study | A study that investigates the effects of an intervention by measuring particular characteristics of a population both before and after taking the intervention, and assessing any change that occurs. |
| Benzodiazepines | A medicine used to control seizures |
| Bias | Influences on a study that can make the results look better or worse than they really are. Bias can occur by chance, deliberately or as a result of systematic errors |

| Term | Definition |
|---|---|
| | in the design and execution of a study. It can also occur at different stages in the research process, for example during the collection, analysis, interpretation, publication or review of research data. For examples see Confounding factor, Performance bias, Publication bias Selection bias. |
| Bilious aspirates | Fluid from stomach which is either vomited or aspirated from a feeding tube |
| Blood sampling | Taking blood to do tests |
| Cachexia | Loss of weight |
| Cannula | A tube inserted into a vein to give drugs and or fluids |
| Carer (caregiver) | Someone who looks after family, partners or friends in need of help because they are ill, frail or have a disability. |
| Case series | Report of a number of cases of a given disease, usually covering the course of the disease and the response to treatment. There is no comparison (control) group of patients. |
| Case-control study | A study to find out the cause(s) of a disease or condition. This is done by comparing a group of patients who have the disease or condition (cases) with a group of people who do not have it (controls) but who are otherwise as similar as possible (in characteristics thought to be unrelated to the causes of the disease or condition). This means the researcher can look for aspects of their lives that differ to see if they may cause the condition. Such studies are retrospective because they look back in time from the outcome to the possible causes of a disease or condition. |
| Cheyne stokes | A pattern of breathing often seen near death |
| Children | Under the age of 18 |
| Chronic illness | An illness that does not have cure, and will therefore go on for a the rest of the patients life |
| Clinical audit | A systematic process for setting and monitoring standards of clinical care. Whereas ‘guidelines’ define what the best clinical practice should be, ‘audit’ investigates whether best practice is being carried out. Clinical audit can be described as a cycle or spiral. Within the cycle there are stages that follow a systematic process of establishing best practice, measuring care against specific criteria, taking action to improve care and monitoring to sustain improvement. The spiral suggests that as the process continues, each cycle aspires to a higher level of quality. |
| Clinical effectiveness | How well a specific test or treatment works when used in the 'real world' (for example when used by a doctor with a patient at home), rather than in a carefully controlled clinical trial. Trials that assess clinical effectiveness are sometimes called management trials. Clinical effectiveness is not the same as efficacy. |
| Clinical efficacy | The extent to which an intervention is active when studied under controlled research conditions. |
| Clinical vignette | Patient-related cases and scenarios that have educational value |

| Term | Definition |
|---|---|
| Clinically assisted hydration | Fluid offered other that the patient asking for it |
| Clinician | A healthcare professional who provides patient care. For example a doctor, nurse or physiotherapist. |
| Close ended question | A question which can be answered with a simple ‘yes’ or ‘no’ or with a specific piece of information |
| Cochrane review | The Cochrane Library consists of a regularly updated collection of evidence-based medicine databases including the Cochrane Database of Systematic Reviews (reviews of rcts prepared by the Cochrane Collaboration). |
| Coherence of findings | Logical or consistent findings |
| Cohort study | A study with 2 or more groups of people – cohorts – with similar characteristics. One group receives a treatment, is exposed to a risk factor or has a particular symptom and the other group does not. The study follows their progress over time and records what happens. |
| Comorbidity | A disease or condition that someone has in addition to the health problem being studied or treated. |
| Comparative group | The group in the study who do not receive the treatment/procedure or receive the treatment which is the norm. This group is used to measure against the treatment/procedure being investigated. |
| Concealment of allocation | The process used to ensure that the person deciding to enter a participant into an RCT does not know the comparison group into which that individual will be allocated. This is distinct from blinding and is aimed at preventing selection bias. Some attempts at concealing allocation are more prone to manipulation than others and the method of allocation concealment is used as an assessment of the quality of a trial. |
| Confidence interval (CI) | There is always some uncertainty in research. This is because a small group of patients is studied to predict the effects of a treatment on the wider population. The confidence interval is a way of expressing how certain we are about the findings from a study, using statistics. It gives a range of results that is likely to include the 'true' value for the population.<br>The CI is usually stated as '95% CI', which means that the range of values has a 95 in 100 chance of including the 'true' value. For example, a study may state that “based on our sample findings, we are 95% certain that the 'true' population blood pressure is not higher than 150 and not lower than 110”. In such a case the 95% CI would be 110 to 150.<br>A wide confidence interval indicates a lack of certainty about the true effect of the test or treatment – often because a small group of patients has been studied. A narrow confidence interval indicates a more precise estimate (for example if a large number of patients have been studied). |
| Confounding factor | Something that influences a study and can result in misleading findings if it is not understood or appropriately dealt with. For example, a study of heart disease may look at a group of people who exercise regularly and a group who do not exercise. If the ages |

| Term | Definition |
|---|---|
| | of the people in the 2 groups are different, then any difference in heart disease rates between the 2 groups could be because of age rather than exercise. Therefore age is a confounding factor. |
| Continuous outcome | Data with a potentially infinite number of possible values within a given range. Height, weight and blood pressure are examples of continuous variables. |
| Control group | A group of people in a study who do not receive the treatment or test being studied. Instead, they may receive the standard treatment (sometimes called 'usual care') or a dummy treatment (placebo). The results for the control group are compared with those for a group receiving the treatment being tested. The aim is to check for any differences. Ideally, the people in the control group should be as similar as possible to those in the treatment group, to make it as easy as possible to detect any effects due to the treatment. |
| Corresponding risk | The risk of an outcome occurring in the group receiving the intervention in the study |
| Cost–benefit analysis (CBA) | A type of economic evaluation where both costs and benefits of healthcare treatment are measured in the same monetary units. If benefits exceed costs, the evaluation would recommend providing the treatment. |
| Cost–consequence analysis (CCA) | Cost-consequence analysis is a type of economic evaluation. This compares the costs (such as treatment and hospital care) and the consequences (such as health outcomes) of a test or treatment with a suitable alternative. Unlike cost-benefit analysis or cost-effectiveness analysis, it does not attempt to summarise outcomes in a single measure (like the quality-adjusted life year) or in financial terms. Instead, outcomes are shown in their natural units (some of which may be monetary) and it is left to decision-makers to determine whether, overall, the treatment is worth carrying out. |
| Cost-effectiveness analysis (CEA) | A type of economic evaluation comparing the costs and the effects on health of different treatments. Health effects are measured in 'health-related units', Cost-effectiveness analysis is 1 of the tools used to carry out an economic evaluation. The benefits are expressed in non-monetary terms related to<br>Health, such as symptom-free days, heart attacks avoided, deaths avoided<br>Or life years gained (that is, the number of years by which life is extended as a result of the intervention). |
| Cost-effectiveness model | An explicit mathematical framework, which is used to represent clinical decision problems and incorporate evidence from a variety of sources in order to estimate the costs and health outcomes. An explicit mathematical framework, which is used to represent clinical decision problems and incorporate evidence from a variety of sources in order to estimate the costs and health outcomes. |
| Cost-minimisation analysis (CMA) | Cost-minimisation analysis is a type of economic evaluation which can be used when the alternatives being compared have equivalent clinical effectiveness. The costs of alternatives are compared in order to |

| Term | Definition |
|---|---|
| | determine which is the cheapest. |
| Cost–utility analysis (CUA) | Cost-utility analysis is a type of economic evaluation where health effects are measured in quality-adjusted life years. A treatment is assessed in terms of its ability to both extend life and to improve the quality of life. |
| COX proportional hazard model | In survival analysis, a statistical model that asserts that the effect of the study factors (for example the intervention of interest) on the hazard rate (the risk of occurrence of an event) in the study population is multiplicative and does not change over time. |
| Credible interval (cri) | The Bayesian equivalent of a confidence interval. |
| Data saturation/sufficiency | The phase in which the researcher has continued sampling and analyzing data until no new data appear and all concepts in the theory are well-developed. It is thought to be the gold standard and is frequently reported in qualitative research. |
| Day and night care | 24/7 care |
| Decision analysis | An explicit quantitative approach to decision-making under uncertainty based on evidence from research. This evidence is translated into probabilities, and then into diagrams or decision trees which direct the clinician through a succession of possible scenarios, actions and outcomes. An explicit quantitative approach to decision-making under uncertainty based on evidence from research. This evidence is translated into probabilities, and then into diagrams or decision trees which direct the clinician through a succession of possible scenarios, actions and outcomes. |
| Delirium | A state of acute confusion |
| Delphi consensus surveys | A method for consensus-building by<br>Using a series of questionnaires to collect data from a panel of selected people |
| Descriptive survey | A survey used to describe characteristics of a population or idea being studied |
| Determinants | Something that controls or influences what happens |
| Dichotomous outcomes | Outcome that can take 1 of 2 possible values, such as dead/alive, smoker/non-smoker, present/not present (also called binary data). |
| Discounting | Costs and perhaps benefits incurred today have a higher value than costs and benefits occurring in the future. Discounting health benefits reflects individual preference for benefits to be experienced in the present rather than the future. Discounting costs reflects individual preference for costs to be experienced in the future rather than the present. Costs and perhaps benefits incurred today have a higher value than costs and benefits occurring in the future. Discounting health benefits reflects individual preference for benefits to be experienced in the present rather than the future. Discounting costs reflects individual preference for costs to be experienced in the future rather than the present. |
| Disease trajectory | The course of an illness over time |
| Dominance | A term used in health economics describing when an option for treatment is both less clinically effective and |

| Term | Definition |
|---|---|
| | more costly than an alternative option. The less effective and more costly option is said to be 'dominated'. A health economics term. When comparing tests or treatments, an option that is both less effective and costs more is said to be 'dominated' by the<br>Alternative |
| Drop-out | A participant who withdraws from a trial before the end. |
| Dying | Actively approaching death on a timescale likely to be measured in days or short weeks |
| Dyspnoea | Breathlessness |
| Dystonia | Movement disorders that cause muscle spasms and contractions |
| Dystonic spasms | As above |
| Economic evaluation | An economic evaluation is used to assess the cost effectiveness of healthcare interventions (that is, to compare the costs and benefits of a healthcare intervention to assess whether it is worth doing). The aim of an economic evaluation is to maximise the level of benefits – health effects – relative to the resources available. It should be used to inform and support<br>The decision-making process; it is not supposed to replace the judgement of healthcare professionals.<br>There are several types of economic evaluation: cost-benefit analysis, cost consequence analysis, cost-effectiveness analysis, cost-minimisation analysis and cost–utility analysis. They use similar methods to define and evaluate costs, but differ in the way they estimate the benefits of a particular drug, programme or intervention. An economic evaluation is used to assess the cost effectiveness of<br>Healthcare interventions (that is, to compare the costs and benefits of a healthcare intervention to assess whether it is worth doing). The aim of an<br>Economic evaluation is to maximise the level of benefits – health effects – relative to the resources available. It should be used to inform and support<br>The decision-making process; it is not supposed to replace the judgement of healthcare professionals.<br>There are several types of economic evaluation: cost-benefit analysis, cost consequence analysis, cost-effectiveness analysis, cost-minimisation<br>Analysis and cost–utility analysis. They use similar methods to define and evaluate costs, but differ in the way they estimate the benefits of a particular drug, programme or intervention. |
| Effect (as in effect measure, treatment effect, estimate of effect, effect size) | A measure that shows the magnitude of the outcome in 1 group compared with that in a control group. For example, if the absolute risk reduction is shown to be 5% and it is the outcome of interest, the effect size is 5%. The effect size is usually tested, using statistics, to find out how likely it is that the effect is a result of the treatment and has not just happened by chance. |
| Effectiveness | How beneficial a test or treatment is under usual or everyday conditions. |
| Effectiveness reviews | Evaluation of how beneficial a test or treatment is |

| Term | Definition |
|---|---|
| | under everyday conditions. |
| Efficacy | How beneficial a test, treatment or public health intervention is under ideal conditions (for example in a laboratory). |
| Encephalopathy | A disorder of the brain |
| End of life care | Care that helps all those with advanced, progressive, incurable illnesses to live as well as possible until they die. It includes management of pain and other symptoms and provision of psychological, social, spiritual and practical support. (definition from National Council for Palliative Care) |
| Enteral tube | A method of feeding via a tube inserted into the stomach |
| Epidemiological study | The study of a disease within a population, defining its incidence and prevalence and examining the roles of external influences (for example infection, diet) and interventions. |
| EQ-5D (EuroQol 5 dimensions) | A standardised instrument used to measure health-related quality-of-life. It can be used across a range of health conditions and treatments and provides a simple descriptive profile and a single index value for health status. A standardised instrument used to measure health-related quality-of-life. It<br>Provides a single index value for health status. |
| Equivalence study | A trial designed to determine whether the response to 2 or more treatments differs by an amount that is clinically unimportant. This is usually demonstrated by showing that the true treatment difference is likely to lie between a lower and an upper equivalence level of clinically acceptable differences. |
| Evidence | Information on which a decision or guidance is based. Evidence is obtained from a range of sources including rcts, observational studies, expert opinion (of clinical professionals or patients). |
| Exclusion criteria (clinical study) | Criteria that define who is not eligible to participate in a clinical study. |
| Exclusion criteria (literature review) | Explicit standards used to decide which studies should be excluded from consideration as potential sources of evidence. |
| Extended dominance | If Option A is both more clinically effective and costly than Option B but has a lower cost per unit of effect, when both are compared with a do-nothing<br>Alternative then Option A is said to have extended dominance over Option B. Option A is therefore more cost effective and should be preferred, other things remaining equal. If Option A is both more clinically effective than Option B and has a lower cost per unit of effect, when both are compared with a do-nothing alternative then Option A is said to have extended dominance over Option B. Option A is therefore more cost effective and should be preferred, other things remaining equal. |
| Extended family | Includes all those important to the child or young person, for example grandparents, other relatives, foster carers |

| Term | Definition |
|---|---|
| Extrapolation | An assumption that the results of studies of a specific population will also hold true for another population with similar characteristics. |
| Extubation | Removal of a tube from the airway providing ventilation |
| False negative | A diagnostic test result that incorrectly indicates that an individual does not have the disease of interest, when they do actually have it. |
| False positive | A diagnostic test result that incorrectly indicates that an individual has the disease of interest, when they actually do not have it. |
| Fixed-effect model | In meta-analysis, a model that calculates a pooled effect estimate using the assumption that all observed variation between studies is caused by random sample variability. Studies are assumed to estimating the same overall effect. |
| focus group | A group of people assembled discuss about an area of interest |
| Follow-up | Observation over a period of time of an individual, group or initially defined population whose appropriate characteristics have been assessed in order to observe changes in health status or health-related variables. |
| Forest plot | A graphical representation of the individual results of each study included in a meta-analysis together with the combined meta-analysis result. The plot also allows readers to see the heterogeneity among the results of the studies. The results of individual studies are shown as squares centred on each study's point estimate. A horizontal line runs through each square to show each study's confidence interval. The overall estimate from the meta-analysis and its confidence interval are shown at the bottom, represented as a diamond. The centre of the diamond represents the pooled point estimate, and its horizontal tips represent the confidence interval. |
| Generalisability | The extent to which the results of a study hold true for groups that did not participate in the research. |
| Glasgow Coma Scale | A neurological scale which aims to give a reliable and objective way of recording the conscious state of a person for initial as well as subsequent assessment. |
| Gold standard | A method, procedure or measurement that is widely accepted as being the best available to test for or treat a disease. |
| GRADE, GRADE profile | A system developed by the GRADE Working Group to address the short-comings of present grading systems in healthcare. The GRADE system uses a common, sensible and transparent approach to grading the quality of evidence. The results of applying the GRADE system to clinical trial data are displayed in a table known as a GRADE profile. |
| Haemoptysis | Coughing up blood |
| Harms | Adverse effects of an intervention. |
| Hazard ratio | A hazard is the rate at which events happen, so that the probability of an event happening in a short time interval is the length of time multiplied by the hazard. |

| Term | Definition |
|---|---|
| | Although the hazard may vary with time, the assumption in proportional hazard models for survival analysis is that the hazard in 1 group is a constant proportion of the hazard in the other group. This proportion is the hazard ratio. |
| Health economics | A branch of economics that studies decisions about the use and distribution of healthcare resources. Study or analysis of the cost of using and distributing healthcare resources. |
| Health-related quality of life (hrqol) | This is a concept with domains that relate to A measure of the effects of an illness to see how it affects someone's day to-day life, physical, mental, emotional, and social functioning. Its particular focus is the impact health status has on quality of life. |
| Hematologic | Relating to the blood |
| Heterogeneity | The term is used in meta-analyses and systematic reviews to describe when the results of a test or treatment (or estimates of its effect) differ |
| Homogenous group | A group of similar people |
| Hydromorphone | A medication to help with pain |
| Hypothesis | The proposed explanation at the start of an investigation made on the basis of prior evidence. It is something that can be tested by the investigation. |
| Hypoxia | Low levels of oxygen |
| Imprecision | Results are imprecise when studies include relatively few patients and few events and thus have wide confidence intervals around the estimate of effect. |
| Incidence | The incidence of a disease is the rate at which new cases occur in a population during a specified period. |
| Inclusion criteria | Characteristics that people must have to be included in the study |
| Inclusion criteria (clinical study) | Specific criteria that define who is eligible to participate in a clinical study. |
| Inclusion criteria (literature review) | Explicit criteria used to decide which studies should be considered as potential sources of evidence. |
| Incremental cost | The extra cost linked to using 1 test or treatment rather than another, or the additional cost of performing a test or providing a treatment more frequently. |
| Incremental cost effectiveness ratio (ICER) | This measure is used to summarise the cost effectiveness of a healthcare intervention. It is defined by the difference in cost between 2 possible interventions, divided by the difference in their effect. |
| Incremental net benefit (INB) | The value (usually in monetary terms) of an intervention net of its cost<br>Compared with a comparator intervention. The INB can be calculated for a<br>Given cost-effectiveness (willingness to pay) threshold. If the threshold is<br>£20,000 per QALY gained then the INB is calculated as: (£20,000 x Incremental QALYs<br>Gained) – Incremental cost. |
| Indirectness | The available evidence is different to the review question being addressed, in terms of population, intervention, comparison and outcome (PICO). |

| Term | Definition |
|---|---|
| Infants | A child aged under the age of 2 |
| Intensive care | A unit to provide a high level of care, often including artificial ventilation |
| Intention-to-treat analysis (ITT) | An assessment of the people taking part in a clinical trial, based on the group they were initially (and randomly) allocated to. This is regardless of whether or not they dropped out, fully complied with the treatment or switched to an alternative treatment. Intention-to-treat analyses are often used to assess clinical effectiveness because they mirror actual practice: that is, not everyone complies with treatment and the treatment people receive may be changed according to how they respond to it. |
| Internal validity | How well an experiment is done and if it is clear that the variable being tested is what is causing the measured effect. |
| Intervention | In medical terms this could be a drug treatment, surgical procedure, diagnostic or psychological therapy. Examples of public health interventions could include action to help someone to be physically active or to eat a more healthy diet. |
| Intervention GRADE approach | GRADE is a systematic and explicit approach to making judgements about quality of evidence and strength of recommendations. |
| Intractable seizures | Seizures that do not respond to all normal management |
| Intravenous | Via a cannula inserted into a vein |
| Invasive techniques | A technique which is some way 'invades' the body, this can be anything from an injection to surgery |
| Kappa statistic | A statistical measure of inter-rater agreement that takes into account the agreement occurring by chance |
| Length of stay | The total number of days a patient stays in hospital. |
| Licence | See Product licence. |
| Life years gained | Mean average years of life gained per person as a result of the intervention<br>Compared with an alternative intervention |
| Life-limiting condition | Any condition which either generally or in a particular individual is thought likely to result in early death |
| Life-threatening condition | Any condition for which curative treatment is not possible or might fail |
| Likelihood ratio | The likelihood ratio combines information about the sensitivity and specificity. It tells you how much a positive or negative result changes the likelihood that a patient would have the disease. The likelihood ratio of a positive test result (LR+) is sensitivity divided by (1 minus specificity). |
| Limitations of a study | Influences on a study that the researcher cannot control which limit the conclusions which can be made |
| Loss to follow-up | Patients who have withdrawn from the clinical trial at the point of follow-up. |
| Malignancy | An illness that is a cancer |
| Markov model | A method for estimating long-term costs and effects for recurrent or chronic |

| Term | Definition |
|---|---|
| | Conditions, based on health states and the probability of transition between<br>Them within a given time period (cycle). |
| Maximal therapy | The most treatment that can be offered |
| Mean | An average value, calculated by adding all the observations and dividing by the number of observations. |
| Mean difference | In meta-analysis, a method used to combine measures on continuous scales (such as weight), where the mean, standard deviation and sample size in each group are known. The weight given to the difference in means from each study (for example how much influence each study has on the overall results of the meta-analysis) is determined by the precision of its estimate of effect. |
| Median | The value of the observation that comes half-way when the observations are ranked in order. |
| Medical sedation | Reducing a patients conscious state for medical reasons such as procedures |
| Medically assisted nutrition | Nutrition provided other than by eating, e.g. via feeding tube |
| Memory-making activities | Activities to help families make memories of their child, e.g. photos, hand or foot prints, locks of hair, recording of voice etc. |
| Meta-analysis | A method often used in systematic reviews. Results from several studies of the same test or treatment are combined to estimate the overall effect of the treatment. |
| Methodology | Systematic, theoretical analysis of the methods applied to a field of study |
| Minimal important difference (MID) | Threshold for clinical importance which represents the minimal important difference for benefit or for harm; for example the threshold at which drug A is less effective than drug B by an amount that is clinically important to patients. |
| Monte carlo | A technique used to approximate the probability of certain outcomes by running multiple simulations using random variables. |
| Multivariate model | A statistical model for analysis of the relationship between 2 or more predictors, (independent) variables and the outcome (dependent) variable. |
| Nasogastric tube | A tube passed through the nose into the stomach to allow provision of fluid, medication and nutrition |
| Neonatal period | The first 28 days of life |
| Neonates | As above |
| Net monetary benefit (NMB) | The value (usually in monetary terms) of an intervention net of its cost. The<br>NMB can be calculated for a given cost-effectiveness (willingness to pay)<br>Threshold. If the threshold is £20,000 per QALY gained then the NMB is<br>Calculated as: (£20,000 x qalys gained) – cost. |
| Network meta-analysis | Meta-analysis in which multiple treatments (that is, 3 or more) are being compared using both direct |

| Term | Definition |
|---|---|
| | comparisons of interventions within rcts and indirect comparisons across trials based on a common comparator. |
| Neuro-disability | Conditions associated with impairment involving the nervous system and includes conditions such as cerebral palsy, autism and epilepsy; |
| Neuroleptics | A group of drugs normally used to manage psychosis |
| Neuropathic | Pain resulting from damage or dysfunction of the nerves |
| Nociceptive | Sensory nervous system responses to stimuli |
| Non-inferiority trial | A trial designed to determine whether the effect of a new treatment is not worse than a standard treatment by more than a pre-specified amount. A one-sided version of an equivalence trial. |
| Non-randomised | When subjects of a study are not allocated to a specific treatment/group at random |
| Number needed to treat (NNT) | The average number of patients who need to be treated to get a positive outcome. For example, if the NNT is 4, then 4 patients would have to be treated to ensure 1 of them gets better. The closer the NNT is to 1, the better the treatment. For example, if you give a stroke prevention drug to 20 people before 1 stroke is prevented, the number needed to treat is 20. |
| Observational before-and-after study | A study where the dependent variables are measured before and after an intervention. |
| Observational retrospective study | Investigators observe and measure variables of interest from past records so the treatment that each person received is beyond the control of the investigator. |
| Observational study | Individuals or groups are observed or certain factors are measured. No attempt is made to affect the outcome. For example, an observational study of a disease or treatment would allow 'nature' or usual medical care to take its course. Changes or differences in 1 characteristic (for example whether or not people received a specific treatment or intervention) are studied without intervening. There is a greater risk of selection bias than in experimental studies. |
| Odds ratio (OR) | Odds are a way to represent how likely it is that something will happen (the probability). An odds ratio compares the probability of something in 1 group with the probability of the same thing in another.<br>An odds ratio of 1 between 2 groups would show that the probability of the event (for example a person developing a disease, or a treatment working) is the same for both. An odds ratio greater than 1 means the event is more likely in the first group. An odds ratio less than 1 means that the event is less likely in the first group.<br>Sometimes probability can be compared across more than 2 groups – in this case, one of the groups is chosen as the 'reference category' and the odds ratio is calculated for each group compared with the reference category. For example, to compare the risk of dying from lung cancer for non-smokers, occasional smokers and regular smokers, non-smokers could be |

| Term | Definition |
|---|---|
| | used as the reference category. Odds ratios would be worked out for occasional smokers compared with non-smokers and for regular smokers compared with non-smokers.<br>See also Confidence interval, Relative risk. |
| Oedema | Swelling of tissues die to collection of fluid |
| Oedematous skin | As above |
| Oncology | Related to cancer |
| Open-ended questions | Questions which require thought and more than a simple 1-word answer. |
| Opioids | A class of pain medication related to morphine |
| Opportunity cost | The loss of other healthcare programmes displaced by investment in or<br>Introduction of another intervention. This may be best measured by the<br>Health benefits that could have been achieved had the money been spent<br>On the next best alternative healthcare intervention. |
| Organ and tissue donation | The donation of body parts after death to assist others |
| Organ failure | Organs stopping working resulting in illness |
| Outcome | The impact that a test, treatment, policy, programme or other intervention has on a person, group or population. Outcomes from interventions to improve the public's health could include changes in knowledge and behaviour related to health, societal changes (for example a reduction in crime rates) and a change in people's health and wellbeing or health status. In clinical terms, outcomes could include the number of patients who fully recover from an illness or the number of hospital admissions, and an improvement or deterioration in someone's health, functional ability, symptoms or situation. Researchers should decide what outcomes to measure before a study begins. |
| P value | The p value is a statistical measure that indicates whether or not an effect is statistically significant. For example, if a study comparing 2 treatments found that 1 seems more effective than the other, the p value is the probability of obtaining these results by chance. By convention, if the p value is below 0.05 (that is, there is less than a 5% probability that the results occurred by chance) it is considered that there probably is a real difference between treatments. If the p value is 0.001 or less (less than a 1% probability that the results occurred by chance), the result is seen as highly significant. If the p value shows that there is likely to be a difference between treatments, the confidence interval describes how big the difference in effect might be. |
| Paediatric | Relating to children |
| Palliative care | Care provided to the advanced progressive illness. Includes care for the families or carers of the person, management of pain, provision of psychological and social support. |
| Paracetamol | Pain relief medication |
| Parental responsibility | Refers to all the rights, duties, powers, responsibilities |

| Term | Definition |
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| | and authority which by law a parent of a child has in relation to the child and his property. |
| Patient controlled analgesia | A method of pain relief which allows the person to administer their own mediation. |
| Performance bias | Systematic differences between intervention groups in care provided apart from the intervention being evaluated. Blinding of study participants (both the recipients and providers of care) is used to protect against performance bias. |
| Pharmacological treatment | Medication |
| Placebo | A fake (or dummy) treatment given to participants in the control group of a clinical trial. It is indistinguishable from the actual treatment (which is given to participants in the experimental group). The aim is to determine what effect the experimental treatment has had over and above any placebo effect caused because someone has received (or thinks they have received) care or attention. |
| Placebo effect | A beneficial (or adverse) effect produced by a placebo and not due to any property of the placebo itself. |
| A posteriori | Reasoning from observed facts |
| Post-hoc analysis | Statistical analyses that are not specified in the trial protocol and are generally suggested by the data. |
| Postnatal | The time period after birth |
| Power (statistical) | The ability to demonstrate an association when one exists. Power is related to sample size; the larger the sample size, the greater the power and the lower the risk that a possible association could be missed. |
| Pre-condition | A medical condition which existed before the condition being examined |
| Prevalence | The prevalence of a disease is the proportion of a population that are cases at a point in time. |
| Primary care | Healthcare delivered outside hospitals. It includes a range of services provided by GPs, nurses, health visitors, midwives, and other healthcare professionals and allied health professionals such as dentists, pharmacists and opticians. |
| Primary caregiver | The person most involved in looking after the child or young person |
| Primary outcome | The outcome of greatest importance; usually the one in a study that the power calculation is based on. |
| Product licence | An authorisation from the Medicines and Healthcare Products Regulatory Agency (MHRA) to market a medicinal product. |
| Prognosis | A probable course or outcome of a disease. Prognostic factors are patient or disease characteristics which influence the course. Good prognosis is associated with a low rate of undesirable outcomes, whereas poor prognosis is associated with a high rate of undesirable outcomes. |
| Prospective study | A research study in which the health or other characteristic of participants is monitored (or 'followed up') for a period of time, with events recorded as they happen. This contrasts with retrospective studies. |

| Term | Definition |
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| Protocol (review) | A document written prior to commencing a review that details exactly how evidence to answer a review question will be obtained and synthesised. It defines in detail the population of interest, the interventions, the comparators/controls and the outcomes of interest (PICO). |
| Proxy outcomes | A way of gauging the gauges progress of research, predicting probable results. |
| Psychological intervention | See the chapter |
| Psychological support groups | A series of regular meetings for children and young people with a life-limiting condition and/or their family members, convened and facilitated by a practitioner with relevant knowledge, skills and expertise in facilitating group activities and conversations using recognised psychological theories and approaches to address the psychological support needs of the group members. |
| Publication bias | Publication bias occurs when researchers publish the results of studies showing that a treatment works well and don't publish those showing it did not have any effect. If this happens, analysis of the published results will not give an accurate idea of how well the treatment works. This type of bias can be assessed by a funnel plot. |
| Qualitative | A type of data that records qualities that are descriptive, subjective or difficult to measure in some way. |
| Quality adjusted life year (QALY) | A measure of health outcome that looks at both length of life and quality of life. Qalys are calculated by estimating the years of life remaining for a patient following a particular care pathway and weighting each year with a quality of life score (on a 0 to 1 scale). One QALY is equal to 1 year of life in perfect health, 2 years at 50% health, and so on. |
| Quality of life | See ‘Health-related quality of life’. |
| Quantitative | Data based on quantities obtained using a measurable process. |
| Random effect model | In meta-analysis, a model that calculates a pooled effect estimate using the assumption that each study is estimating a different true treatment effect due to real differences between studies. Observed variation in effects are therefore caused by a combination of random sample variability (within-study variation) and heterogeneity between studies (between-study variation). The overall effects is an average of the estimated true study effects. |
| Randomisation | Assigning participants in a research study to different groups without taking any similarities or differences between them into account. For example, it could involve using a random numbers table or a computer-generated random sequence. It means that each individual (or each group in the case of cluster randomisation) has the same chance of receiving each intervention. |
| Randomised controlled trial (RCT) | A study in which a number of similar people are randomly assigned to 2 (or more) groups to test a |

| Term | Definition |
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| | specific drug or treatment. One group (the experimental group) receives the treatment being tested, the other (the comparison or control group) receives an alternative treatment, a dummy treatment (placebo) or no treatment at all. The groups are followed up to see how effective the experimental treatment was. Outcomes are measured at specific times and any difference in response between the groups is assessed statistically. This method is also used to reduce bias. |
| Recollection bias | A systematic error caused by differences in the accuracy or completeness of recollections by people regarding events or experiences from the past. |
| Recruitment bias | When proper randomisation is not achieved when recruiting individuals, meaning that the sample obtained may not be representative of the population intended to be analysed. |
| Reference standard | The test that is considered to be the best available method to establish the presence or absence of the outcome – this may not be the one that is routinely used in practice. |
| Relative risk (RR) | The ratio of the risk of disease or death among those exposed to certain conditions compared with the risk for those who are not exposed to the same conditions (for example the risk of people who smoke getting lung cancer compared with the risk for people who do not smoke). If both groups face the same level of risk, the relative risk is 1. If the first group had a relative risk of 2, subjects in that group would be twice as likely to have the event happen. A relative risk of less than 1 means the outcome is less likely in the first group. Relative risk is sometimes referred to as risk ratio. |
| Reporting bias | See Publication bias. |
| Resilience | Resilience is the process of adapting well in the face of adversity, trauma, tragedy, threats or significant sources of stress (such as family and relationship problems, serious health problems or workplace and financial stressors). It means withstanding and "bouncing back" from difficult experiences, becoming more resourceful and better able to deal with other difficulties in the future. Building resilience involves fostering the ability to struggle well, surmount obstacles and go on to live well and sustain good relationships. The skills and resources of resilience enable individuals and families to respond successfully to crises and persistent challenges, to adapt and to “grow” from these experiences. |
| Resource implication | The likely impact in terms of finance, workforce or other NHS resources. |
| Respiratory distress | Feeling breathless |
| Respiratory rate | How fast you are breathing |
| Retrospective cohort study | A study of a group of individuals that share a common exposure factor to determine its effect on the development of a disease. |
| Retrospective study | A research study that focuses on the past and present. The study examines past exposure to suspected risk factors for the disease or condition. Unlike prospective |

| Term | Definition |
|---|---|
| | studies, it does not cover events that occur after the study group is selected. |
| Review protocol | A document that sets out the reviewers' intentions with regard to the topic and the methods to be used for inclusion in the review |
| Review question | The plan or set of steps to be followed in a study. A protocol for a systematic review describes the rationale for the review, the objectives and the methods that will be used to locate, select and critically appraise studies, and to collect and analyse data from the included studies. |
| Risk ratio | The ratio of the probability of an event occurring in an exposed group to the probability of the even occurring in a non-exposed group. |
| Sample | A set of data collected from a defined population |
| Secondary care | Care provided in hospitals. |
| Secondary outcome | An outcome used to evaluate additional effects of the intervention deemed a priori as being less important than the primary outcomes. |
| Seizure | Often termed fit or convulsion. A seizure is the physical effect or change in behaviour which happens after abnormal electrical activity in the brain. Specific symptoms depend on which parts of the brain are involved. |
| Selection bias | Selection bias occurs if:<br>• The characteristics of the people selected for a study differ from the wider population from which they have been drawn; or<br>• There are differences between groups of participants in a study in terms of how likely they are to get better. |
| Self-selection bias | When individuals have selected themselves into a group, causing a biased sample. See ‘bias’ |
| Sensitivity | How well a test detects the thing it is testing for. If a diagnostic test for a disease has high sensitivity, it is likely to pick up all cases of the disease in people who have it (that is, give a 'true positive' result). But if a test is too sensitive it will sometimes also give a positive result in people who don't have the disease (that is, give a 'false positive'). For example, if a test were developed to detect if a woman is 6 months pregnant, a very sensitive test would detect everyone who was 6 months pregnant but would probably also include those who are 5 and 7 months pregnant. If the same test were more specific (sometimes referred to as having higher specificity), it would detect only those who are 6 months pregnant and someone who was 5 months pregnant would get a negative result (a 'true negative'). But it would probably also miss some people who were 6 months pregnant (that is, give a 'false negative'). |
| Sensitivity analysis | A means of representing uncertainty in the results of an analysis. Uncertainty may arise from missing data, imprecise estimates or methodological controversy. Sensitivity analysis also allows for exploring the generalisability of results to other settings. The |

| Term | Definition |
|---|---|
| | analysis is repeated using different assumptions to examine the effect on the results. |
| Serum electrolyte concentrations | Blood test |
| Significance (statistical) | A result is deemed statistically significant if the probability of the result occurring by chance is less than 1 in 20 ($p<0.05$). |
| Specificity | The proportion of true negatives that are correctly identified as such. For example, in diagnostic testing the specificity is the proportion of non-cases correctly diagnosed as non-cases. In terms of literature searching a highly specific search is generally narrow and aimed at picking up the key papers in a field and avoiding a wide range of papers. |
| Stakeholder | An organisation with an interest in a topic on which NICE is developing a clinical guideline or piece of public health guidance. Organisations that register as stakeholders can comment on the draft scope and the draft guidance. Stakeholders may be: |
| Standard deviation (SD) | A measure of the spread or dispersion of a set of observations, calculated as the average difference from the mean value in the sample. |
| Structured interview | When each interviewee is presented with exactly the same questions in the same order. |
| Subcutaneous infusion | Administering drugs into tissues via a needle |
| Subgroup analysis | An analysis in which the intervention effect is evaluated in a defined subset of the participants in a trial, or in complementary subsets. |
| Systematic review | A review in which evidence from scientific studies has been identified, appraised and synthesised in a methodical way according to predetermined criteria. It may include a meta-analysis. |
| Team around the child | Professionals involved in co-ordinating and delivering integrated services for children and young people (defined by Children's Workforce Development Council) |
| Thematic analysis | A type of analysis which records patterns within data. Themes are patterns across sets of data that are important to the description of a phenomenon and are associated with a specific research question. |
| Time horizon | The time span over which costs and health outcomes are considered in a<br>Decision analysis or economic evaluation. |
| Tissue donation | See organ donation |
| Transdermal | Administering medication via application to the skin |
| Treatment allocation | Assigning a participant to a particular arm of a trial. |
| True negative | A diagnostic test result that correctly indicates that an individual does not have the disease of interest when they actually do not have it. |
| True positive | A diagnostic test result that correctly indicates that an individual has the disease of interest when they do actually have it. |
| Univariate | Analysis which separately explores each variable in a data set. |
| Utility | In health economics, a 'utility' is the measure of the |

| Term | Definition |
|---|---|
| | preference or value that an individual or society places upon a particular health state. It is generally a number between 0 (representing death) and 1 (perfect health). The most widely-used measure of benefit in cost–utility analysis is the quality-adjusted life year (QALY), but other measures include disability-adjusted life years (DALYs) and healthy-year equivalents (HYEs). |
| Young people | A person aged 16-18 years |

## 13.2 Abbreviations

**Table 106: Abbreviations**

| Abbreviation | Description |
|---|---|
| ACP | Advance Care Plan |
| AFS | Affective Facial Score |
| CI | Confidence interval |
| DNR | Do not resuscitate |
| HCP | Healthcare professional |
| HADS | Hospital Anxiety and Depression Scale |
| HTA | health technology assessment |
| ICER | incremental cost-effectiveness ratio |
| ICYP | Infant, child or young person |
| CYP | Child or young person |
| ITU | Intensive care unit |
| LLC | Life-limiting condition |
| MD | Mean difference |
| MDT | Multi-disciplinary team |
| MID | Minimal important difference |
| NC | Not calculable |
| NHS | National Health Service |
| NHS EED | NHS Economic Evaluation Database |
| NICE | The National Institute for Health and Care Excellence |
| NICU | Neonatal intensive care unit |
| PCA | Patient controlled analgesia |
| PICU | Paediatric intensive care unit |
| PPC | Paediatric palliative care |
| PPHC | Palliative home care |
| QALY | quality-adjusted life year |
| QoL | Quality of life |
| QOLLTI-F | Quality of life in life-threatening illness-Family carer version |
| RCT | Randomized control trial |
| RR | Risk Ratio |
| TENS | Transcutaneous electrical nerve stimulation |
| TFSL | Together for Short Lives |
| VAS | Visual analogue scale |
| WHO | World Health Organisation |
| WTE | Whole time equivalent |

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# 14 Appendices

The appendices are presented in 4 separate documents; Appendices G, L and K are in individual documents and the fourth contains all the remaining appendices.