Spasticity in children and young people with non-progressive brain disorders: management of spasticity and co-existing motor disorders and their early musculoskeletal complications

NICE guideline

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If you wish to comment on this version of the guideline, please be aware that all the supporting information and evidence is contained in the full version.

Contents

Introduction

This guideline covers the management of spasticity and co-existing motor disorders and their early musculoskeletal complications in children (including babies) and young people aged up to 19 years who have non-progressive brain disorders.

Muscle spasticity is defined as an increase in resistance to muscle stretch proportional to the velocity at which the muscle is stretched. Spasticity is a component of the upper motor neurone lesion (UMNL) classically presumed to be caused by a lesion of the pyramidal tract between the motor cortex and the anterior horn cell in the spinal cord. Weakness, poor selective motor control, exaggerated deep tendon reflexes, and difficulties with motor planning are the other components of the UMNL.

Dystonia, chorea and athetosis are motor symptoms caused by lesions to the extra-pyramidal and other motor tracts but can also be symptoms of progressive brain pathologies. In children with cerebral palsy, a broad diagnostic category of dyskinesia is used. Ataxia may be part of cerebral palsy but is more common in children with hydrocephalus and can also be caused by progressive brain disorders.

Cerebral palsy is the most common condition responsible for a UMNL in children and young people. The incidence of cerebral palsy is not known, but its prevalence is 186 per 100,000 population, such that a total of 110,000 people are affected in the UK.

Prematurity is a strong risk factor for the development of a UMNL and cerebral palsy. Forty percent of antenatal or perinatal acquired cerebral palsy occurs in children who are born prematurely and who may have additional non-neurological complications of prematurity (for example, chronic gastrointestinal disorders). Such disorders may worsen spasticity and dystonia.

Difficulties during labour that affect oxygen and blood supply to the fetal brain are a common cause of brain damage leading to cerebral palsy. The strongest

risk factor is the development of severe neonatal encephalopathy in the first few hours after birth.

Acquired brain injury means brain injury that occurs after the neonatal period. The management of spasticity and associated motor disorders acquired after birth or after head injury follows the same principles as in children and young people with antenatal or perinatal causes of their motor disorders.

The impact of the spasticity and co-existing motor disorders and their early musculoskeletal complications on the child or young person varies. Common problems include impaired motor function affecting ability to participate in society, pain from muscle spasms, motor developmental delay and difficulties with daily care due to the onset of secondary complications of spasticity. Therapy should be tailored to meet the problems faced by the individual child or young person.

Motor impairment can also be described in terms of the severity of functional motor impairment. Some of the recommendations refer to the Gross Motor Functional Classification System (GMFCS). This is a five-point scale that describes gross motor function: level 1, walks without restrictions; level 2, walks without assistive devices; level 3, walks with assistive devices; level 4, has limited self-mobility; level 5, has severely limited self-mobility even with assistive devices.

There is considerable variation in practice in managing spasticity, including variation in availability of treatments and the intensity of their use. Planning therapy has become more complex following the increase in the range of treatments available for managing motor disorders during the past two decades. This guideline will help healthcare professionals to select and use appropriate therapies for individual children or young people. Parents and carers also need guidance on choosing the most appropriate therapy, and to ensure that the time, effort and their own resources are used to the best to enhance quality of life for the child or young person and their family.

This guideline recommends some drugs for indications for which they do not have a UK marketing authorisation at the date of publication, if there is good

evidence to support that use. Where recommendations have been made for the use of drugs outside their licensed indications ('off-label use'), these drugs are marked with a footnote in the recommendations.

Patient-centred care

This guideline offers best practice advice on the care of children and young people with spasticity and co-existing motor disorders caused by non-progressive brain disorders.

Treatment and care should take into account patients' needs and preferences. People with spasticity and non-progressive brain disorders should have the opportunity to make informed decisions about their care and treatment, in partnership with their healthcare professionals. If patients do not have the capacity to make decisions, healthcare professionals should follow the Department of Health's advice on consent (available from www.dh.gov.uk/en/DH_103643) and the code of practice that accompanies the Mental Capacity Act (available from www.dh.gov.uk/en/SocialCare/Deliveringsocialcare/MentalCapacity). In Wales, healthcare professionals should follow advice on consent from the Welsh Government (available from www.wales.nhs.uk/consent).

If the patient is under 16, healthcare professionals should follow the guidelines in 'Seeking consent: working with children' (available from www.dh.gov.uk).

Good communication between healthcare professionals and patients is essential. It should be supported by evidence-based written information tailored to the patient's needs. Treatment and care, and the information patients are given about it, should be culturally appropriate. It should also be accessible to people with additional needs such as physical, sensory or learning disabilities, and to people who do not speak or read English.

Families and carers should be given the information and support they need.

Care of young people in transition between paediatric and adult services should be planned and managed according to the best practice guidance described in 'Transition: getting it right for young people' (available from www.dh.gov.uk).

Adult and paediatric healthcare teams should work jointly to provide assessment and services to young people with spasticity and co-existing motor disorders caused by non-progressive brain disorders. Diagnosis and management should be reviewed throughout the transition process, and there should be clarity about who is the lead clinician to ensure continuity of care.

Key priorities for implementation

The following recommendations have been identified as priorities for implementation.

Principles of care

- Offer immediate referral to a local multidisciplinary child development team that can be accessed when needed and is linked to regional specialist centres. **[1.1.1]**

- Offer a management programme that is:
  - individualised
  - goal focused
  - developed and implemented in partnership with the child or young person and their family or carers. **[1.1.4]**

- Local multidisciplinary child development teams and regional specialist centres should enable children and young people (and when appropriate their parents or carers) to be partners in the development and implementation of management programmes by offering:
  - relevant information and educational materials
  - regular opportunities for discussion and
  - advice on the child or young person's developmental potential and how different treatment options may affect this potential. **[1.1.5]**

- Monitor the child or young person for:
  - progression of spasticity
  - development of secondary consequences of spasticity
  - response to treatments
  - the need for changes to individualised goals and
  - the need for timely referral to regional specialist centres. **[1.1.9]**

- Offer adjunctive physical therapy following treatments involving botulinum toxin type A, continuous pump-administered intrathecal baclofen,

orthopaedic surgery or selective dorsal rhizotomy to ensure effectiveness of these treatments. **[1.1.11]**

- Before starting treatment, regional specialist centres should ensure that local multidisciplinary child development teams have allocated resources for locally provided post-treatment services. **[1.1.13]**

Physical therapy (physiotherapy and occupational therapy)

- Offer to refer children and young people to a physiotherapist who is a member of the local multidisciplinary child development team. **[1.2.1]**

Intrathecal baclofen

- Consider treatment with continuous pump-administered intrathecal baclofen if, despite the use of non-invasive treatments, spasticity, with or without dystonia, is causing difficulties with any of the following:
  - pain or muscle spasms
  - posture or function
  - self-care (or ease of care in the case of parents or carers)[1]. **[1.6.1]**

Orthopaedic surgery

- Offer children and young people referral to an orthopaedic surgeon if there is clinical or radiological evidence of hip displacement or spinal deformity. **[1.7.1]**

- Monitor children and young people to identify displacement of the hip and spinal deformity. **[1.7.4]**

[1] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

1 Guidance

The following guidance is based on the best available evidence. The full guideline ([hyperlink to be added for final publication]) gives details of the methods and the evidence used to develop the guidance.

1.1 Principles of care

1.1.1 Offer immediate referral to a local multidisciplinary child development team that can be accessed when needed and is linked to regional specialist centres.

1.1.2 The local multidisciplinary child development team should be experienced in the management of spasticity in children and young people and include a paediatrician, a paediatric physiotherapist and have access to a paediatric occupational therapist.

1.1.3 Access to a paediatric occupational therapist is needed for children and young people with spasticity that affects the upper limb.

1.1.4 Offer a management programme that is:

- individualised
- goal focused
- developed and implemented in partnership with the child or young person and their family or carers.

1.1.5 Local multidisciplinary child development teams and regional specialist centres should enable children and young people (and when appropriate their parents or carers) to be partners in the development and implementation of management programmes by offering:

- relevant information and educational materials
- regular opportunities for discussion and
- advice on the child or young person's developmental potential and how different treatment options may affect this potential.

1.1.6 When formulating a management programme take into account the impact of treatment schedules on family circumstances.

1.1.7 Identify and agree with children and young people (and where appropriate their parents or carers) goals and assessments that:

- are appropriate for their age and development
- will aim to improve their body function and structure and activity and participation in line with the domains of the World Health Organization's International Classification of Functioning, Disability and Health[2].

1.1.8 Record and communicate the child or young person's individualised goals within the local multidisciplinary child development team and with all healthcare professionals who care for them in different settings.

1.1.9 Monitor the child or young person for:

- progression of spasticity
- development of secondary consequences of spasticity
- response to treatments
- the need for changes to individualised goals and
- the need for timely referral to regional specialist centres.

1.1.10 Do not offer botulinum toxin type A, continuous pump-administered intrathecal baclofen, orthopaedic surgery or selective dorsal rhizotomy to children and young people unless they are participating actively in a programme of care and physical therapy.

1.1.11 Offer adjunctive physical therapy following treatments involving botulinum toxin type A, continuous pump-administered intrathecal baclofen, orthopaedic surgery or selective dorsal rhizotomy to ensure effectiveness of these treatments.

[2] World Health Organization International Classification of Functioning, Disability and Health (ICF), available from www.who.int/classifications/icf/en/

1.1.12 Healthcare professionals in regional specialist centres who assess children and young people's suitability for oral drugs, botulinum toxin type A, continuous pump-administered intrathecal baclofen, orthopaedic surgery or selective dorsal rhizotomy should communicate with the child or young person's local multidisciplinary child development team to ensure compatibility and continuity of local and specialist services.

1.1.13 Before starting treatment, regional specialist centres should ensure that local multidisciplinary child development teams have allocated resources for locally provided post-treatment services.

1.2 Physical therapy (physiotherapy and occupational therapy)

1.2.1 Offer to refer children and young people to a physiotherapist who is a member of the local multidisciplinary child development team.

1.2.2 Offer children and young people a physical therapy programme tailored to their individual needs and aimed at specific goals, such as:

- enhancing skill development and improving function
- enhancing the ability to participate in everyday activities
- preventing or delaying the onset of complications such as contractures.

1.2.3 When formulating physical therapy programmes for children and young people take account of:

- the views of the child or young person and their parents or carers
- the likelihood of achieving the intended goals of treatment
- the implications for the child or young person and their family in implementing the plan, including the time and effort involved and

potential barriers (for example, barriers associated with particular cultural practices).

1.2.4 Consider task-focused active-use therapies such as constraint-induced movement therapy followed by bimanual therapy to enhance manual skills.

1.2.5 Consider structuring task-focused active-use therapy as an intensive programme over a short time period (for example, 4–8 weeks).

1.2.6 Consider muscle-strengthening therapy where assessment suggests that muscle weakness is contributing to loss of function or joint deformity.

1.2.7 Direct muscle-strengthening therapies towards specific goals and incorporate progressive repetitive exercises performed against resistance.

1.2.8 Consider postural management strategies to:

- prevent or slow the development of contractures in children and young people at risk of developing these
- enable the child or young person to take part in activities appropriate to the child or young person’s stage of development.

1.2.9 As part of postural management consider an individualised physical therapy programme that includes:

- resting positions and
- low-load active or passive stretching over 24 hours.

1.2.10 Offer training to parents and carers involved in delivering postural management programmes.

1.2.11 Assess whether any equipment or techniques used in the physical therapy plan is safe and appropriate, for example in children or young people with any of the following:

- poorly controlled co-existing epilepsy
- respiratory compromise
- risk of aspiration
- risk of bone fracture due to osteoporosis (for example, children and young people who are non-ambulatory, malnourished or taking anticonvulsant therapy).

1.2.12 For children and young people who are at risk of bone fractures due to osteoporosis (for example, children and young people who are non-ambulatory, malnourished or taking anticonvulsant therapy), consider sustained low-load stretching to prevent or limit contractures and joint deformity. Depending on the individual child or young person's circumstances (for example recent history of fractures, bone pain, broken skin), consider low-load stretching and weight bearing including use of orthoses or serial casting.

1.2.13 Monitor children and young people at risk of developing functional difficulties related to their condition. Consider a programme of daily maintenance activities for children and young people with or at risk of developing functional difficulties.

1.2.14 Consider the use of serial casting after botulinum toxin type A treatment to improve passive range of movement if muscle tightness is identified alongside dynamic spasticity. To improve the cast's tolerability and allow better stretch of muscle, do not apply serial casts for 2-4 weeks after botulinum toxin type A treatment.

1.2.15 Offer children and young people and their parents or carers verbal and written information about physical therapy interventions needed to achieve intended goals. This information should emphasise possible advantages as well as difficulties and possible

adverse effects (for example time commitment and discomfort) to enable them to participate in choosing a suitable physical therapy programme.

1.2.16 Reassess at regular intervals all children and young people receiving a programme of physical therapy to ensure that:

- the intended goals are being achieved
- the therapy programme remains appropriate to the child or young person's individual needs.

1.2.17 When considering who should deliver physical therapy, take into account:

- whether the child or young person and their parents or carers are able to deliver the specific therapy
- what training the child or young person or their parents or carers might need
- the wishes of the child or young person and their parents or carers.

1.2.18 Physical therapists should have a central role in preparing young people (and their parents or carers) for transition and transfer to adult physical therapy services (for example, helping them to take responsibility for their own physical therapy).

1.3 Orthoses

General principles

1.3.1 Consider orthoses for children and young people with spasticity to:

- improve posture
- facilitate upper limb function
- improve walking efficiency
- prevent or slow development of contractures
- prevent or slow hip migration.

1.3.2 Determine realistic goals for treatment with orthoses based on a careful individual assessment, and discuss the options, risks and benefits of wearing them with children and young people and their parents or carers.

1.3.3 Ensure that orthoses have been designed, sized and fitted correctly.

1.3.4 Inform children and young people with orthoses and their parents and carers:

- how to apply them
- when to wear them and for how long
- when and where to seek further advice.

1.3.5 Ensure that an orthotist is involved when a custom-made orthosis is being used.

1.3.6 Minimise delays in the supply of orthoses after measurement and in the repair of orthoses.

1.3.7 Review orthotic use at every contact with the local multidisciplinary child development team to ensure that orthoses:

- are still acceptable to the child or young person and their parents or carers
- remain in good repair
- remain appropriate to intended treatment goals
- remain well fitting
- are being used as advised
- are not causing discomfort or pain
- are not causing injury
- are not causing sleep disturbance.

Cautions in the use of orthoses

1.3.8 Assess whether orthoses might:

- cause difficulties with self-care or care by others
- cause difficulties in relation to hygiene
- be unacceptable to the child or young person because of their appearance.

1.3.9 Advise about the risk of pressure sores with orthoses.

1.3.10 Inform children and young people and their parents or carers to remove orthoses that are causing pain that cannot be relieved immediately through repositioning of the limb in the orthosis or adjustment of the strapping.

1.3.11 When deciding whether to offer an orthosis, balance the benefits against the risks and potential consequences of muscle wasting through lack of muscle use. Discuss these with the child or young person and their parents or carers.

1.3.12 Be cautious in offering rigid orthoses to children and young people with severe spasticity or dyskinesis because rigid orthoses are often poorly tolerated in this group.

Botulinum toxin type A injection and orthoses

1.3.13 Consider an orthosis after treatment with botulinum toxin type A.

1.3.14 Consider treatment with botulinum toxin type A if this is likely to improve the tolerability of an orthosis.[3]

[3] At the time of publication (October 2011), some botulinum toxin type A products were licensed for use in focal spasticity in children and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, two years of age or older. Other products were licensed only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Prescribers should refer to the summaries of product characteristics when considering or offering botulinum toxin type A and, where appropriate, informed consent should be obtained and documented.

Overnight use of orthoses

1.3.15 Consider overnight use of orthoses. If an orthosis is used overnight:

- check that overnight use does not disturb sleep
- use night resting splints for muscles that control two joints (for example, the ankle and knee, in the case of the gastrocnemius muscle).

Lower limb orthoses

1.3.16 When deciding whether to offer an ankle–foot orthosis, balance the benefits against the risk of worsened gait in children and young people with:

- hip or knee contractures
- femoral or tibial anteversion.

Discuss these with the child or young person and their parents and carers.

1.3.17 Consider ground reaction ankle–foot orthoses to assist with walking if the child or young person has a crouch gait and good passive range of movement at the hip and knee.

1.3.18 For children and young people with equinus deformities that impair their gait consider:

- a solid ankle–foot orthosis if they have good control of knee or hip extension
- a hinged ankle–foot orthosis if they have poor control of knee or hip extension.

1.3.19 In children whose motor development is between 8 months and 2 years consider offering supramalleolar orthoses or supportive orthotic footwear instead of ankle–foot orthoses.

1.3.20 Consider ankle–foot orthoses for children and young people with serious functional limitations (GMFCS levels 4 and 5) to improve

foot position for sitting, transfers between sitting and standing, and assisted standing.

1.3.21 Inform children and young people and their parents and carers that ankle–foot orthoses intended to stretch muscles (for example, rigid, hinged or ground-reaction force ankle–foot orthoses) should usually be worn for at least 6 hours each day.

1.3.22 Consider knee gaiters for children and young people with knee flexion deformities.

1.3.23 Consider hip orthoses:

- to improve function if scissoring is causing difficulties with sitting, standing or walking
- to limit hip adduction and reduce the risk of hip migration.

Upper limb and trunk orthoses

1.3.24 Consider the following for children and young people with upper limb spasticity:

- elbow gaiters to maintain extension and improve function
- rigid wrist orthoses to prevent contractures and limit wrist and hand flexion deformity
- dynamic orthoses to improve hand function (for example, a thumb abduction splint if the child or young person has a ‘thumb in palm’ deformity).

1.3.25 Consider offering body trunk orthoses to children and young people for the management of spasticity with co-existing scoliosis or kyphosis if this will help with sitting.

1.4 Oral drugs

1.4.1 Consider oral diazepam if spasticity is contributing to:

- discomfort or pain
- muscle spasms (for example night-time muscle spasms)

- functional disability and
- a rapid effect is desirable (for example, in pain crisis).

1.4.2 Consider oral baclofen if spasticity is contributing to:

- discomfort or pain
- muscle spasms
- functional disability and
- a sustained long-term effect is desired (for example, to relieve continuous discomfort or to improve motor function).

1.4.3 Start oral diazepam treatment with a single dose at bedtime. If the clinical response is unsatisfactory consider:

- increasing the dose or
- adding a daytime dose.

1.4.4 Start oral baclofen treatment with a low dose and increase the dose stepwise over about 4 weeks to achieve the optimum therapeutic effect.

1.4.5 If oral diazepam is used because of its rapid onset of action, consider changing to oral baclofen if long-term treatment is indicated.

1.4.6 Continue using oral diazepam or oral baclofen if they have a clinical benefit and are well tolerated, but consider whether to stop treatment every time the child or young person's management programme is reviewed and at least every 6 months.

1.4.7 If adverse effects (such as drowsiness) occur with oral diazepam or oral baclofen consider reducing the dose or stopping treatment.

1.4.8 If the clinical response to oral diazepam or oral baclofen used alone is unsatisfactory within 4–6 weeks, stop using the drug or consider a trial of combination treatment with both oral diazepam and oral baclofen.

1.5 Botulinum toxin type A

When to use botulinum toxin type A

1.5.1 Consider botulinum toxin type A where focal spasticity of the upper limb is:

- impeding fine motor function
- compromising care and hygiene
- causing pain
- impeding tolerance of other treatments, such as orthoses
- causing concerns about appearance to the child or young person.[4]

1.5.2 Consider botulinum toxin type A where focal spasticity of the lower limb is:

- impeding gross motor function
- compromising care and hygiene
- causing pain
- disturbing sleeping patterns
- impeding tolerance of other treatments, such as orthoses and use of equipment to support posture
- causing cosmetic concerns to the child or young person.[5]

1.5.3 Do not offer botulinum toxin type A in children and young people:

- with severe muscle weakness

[4] At the time of publication (October 2011), some botulinum toxin type A products were licensed for use in focal spasticity in children and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, two years of age or older. Other products were licensed only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Prescribers should refer to the summaries of product characteristics when considering or offering botulinum toxin type A and, where appropriate, informed consent should be obtained and documented.

[5] At the time of publication (October 2011), some botulinum toxin type A products were licensed for use in focal spasticity in children and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, two years of age or older. Other products were licensed only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Prescribers should refer to the summaries of product characteristics when considering or offering botulinum toxin type A and, where appropriate, informed consent should be obtained and documented.

- with a previous adverse reaction or allergy
- who are currently taking aminoglycosides.

1.5.4 Consider botulinum toxin type A with caution if:

- the child or young person has any of the following
  - a bleeding disorder or is receiving anti-coagulation therapy
  - generalised spasticity
  - fixed muscle contractures
  - marked bony deformity **or**
- where there are concerns about the child or young person engaging with post-treatment adjunctive therapy.[6]

1.5.5 Consider using botulinum toxin type A to treat rapid-onset spasticity causing abnormal postures and soft-tissue shortening after acquired brain injury.[7]

Assessment

1.5.6 Local multidisciplinary child development teams and regional specialist centres involved in the assessment and administration of botulinum toxin type A should have expertise in child neurology, child development and musculoskeletal assessment in order to decide on:

- the need for botulinum toxin type A
- administration of botulinum toxin type A
- offering repeat injections.

[6] At the time of publication (October 2011), some botulinum toxin type A products were licensed for use in focal spasticity in children and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, two years of age or older. Other products were licensed only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Prescribers should refer to the summaries of product characteristics when considering or offering botulinum toxin type A and, where appropriate, informed consent should be obtained and documented.

[7] At the time of publication (October 2011), some botulinum toxin type A products were licensed for use in focal spasticity in children and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, two years of age or older. Other products were licensed only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Prescribers should refer to the summaries of product characteristics when considering or offering botulinum toxin type A and, where appropriate, informed consent should be obtained and documented.

1.5.7 Include movement and motor function in assessments for treatment with botulinum toxin type A and involve a paediatric physiotherapist or paediatric occupational therapist.

1.5.8 Ensure that children and young people who receive treatment with botulinum toxin type A are offered timely access to orthotic services (see recommendation 1.3.13).

Treatment

1.5.9 Consider using ultrasound-guided injection or electrical muscular stimulation when injecting botulinum toxin type A into muscles.[8]

1.5.10 Minimise distress to the child or young person undergoing treatment with botulinum toxin type A by considering the need for the:

- topical or systemic analgesia or anaesthesia
- sedation.

1.5.11 Local multidisciplinary child development teams and regional specialist centres involved in the assessment and administration of botulinum toxin type A should:

- monitor effectiveness of the first botulinum toxin type A injection by repeating pre-injection assessment 6-12 weeks after the injection (both assessments should preferably be performed by the same healthcare professionals)
- monitor effectiveness of subsequent botulinum toxin type A injections and the need for further injections at 3–6 months.

[8] At the time of publication (October 2011), some botulinum toxin type A products were licensed for use in focal spasticity in children and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, two years of age or older. Other products were licensed only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Prescribers should refer to the summaries of product characteristics when considering or offering botulinum toxin type A and, where appropriate, informed consent should be obtained and documented.

1.5.12 If the clinical response to treatment is satisfactory review the child or young person's goals and consider repeat injections if:

- the problem that prompted initial treatment returns after treatment wears off
- new goals are identified.[9]

1.5.13 Inform children and young people and their parents and carers:

- how to recognise serious but rare complications associated with botulinum toxin type A (swallowing difficulties and breathing difficulties)
- that these complications may arise during the first week after botulinum toxin type A treatment, and
- that the child or young person should return to hospital immediately if they occur.

1.5.14 Consider injecting botulinum toxin type A into more than one muscle, but ensure that:

- maximum doses are not exceeded
- a clear functional goal is identified
- the child or young person and their parents or carers understand the possible side effects.[10]

[9] At the time of publication (October 2011), some botulinum toxin type A products were licensed for use in focal spasticity in children and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, two years of age or older. Other products were licensed only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Prescribers should refer to the summaries of product characteristics when considering or offering botulinum toxin type A and, where appropriate, informed consent should be obtained and documented.

[10] At the time of publication (October 2011), some botulinum toxin type A products were licensed for use in focal spasticity in children and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, two years of age or older. Other products were licensed only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Prescribers should refer to the summaries of product characteristics when considering or offering botulinum toxin type A and, where appropriate, informed consent should be obtained and documented.

1.6 Intrathecal baclofen

When to consider intrathecal baclofen

1.6.1 Consider treatment with continuous pump-administered intrathecal baclofen if, despite the use of non-invasive treatments, spasticity, with or without dystonia, is causing difficulties with any of the following:

- pain or muscle spasms
- posture or function
- self-care (or ease of care in the case of parents or carers). [11]

1.6.2 Be aware that children and young people who benefit from continuous pump-administered intrathecal baclofen typically have:

- moderate to severe motor function problems (GMFCS level 3-5)
- bilateral spasticity affecting upper and lower limbs. [12]

1.6.3 When considering continuous pump-administered intrathecal baclofen, balance the benefits against the risk of reducing spasticity if that spasticity supports function (for example, by compensating for muscle weakness) which may have adverse consequences. Discuss this with the child or young person and their parents and carers. [13]

Intrathecal baclofen testing

1.6.4 In children and young people being considered for treatment with continuous pump-administered intrathecal baclofen perform

[11] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

[12] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

[13] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

intrathecal baclofen testing to assess therapeutic effect and to check for adverse effects. [14]

1.6.5 Before starting intrathecal baclofen testing inform children and young people and their parents or carers verbally and in writing about:

- what the test will entail
- how the test might predict successful treatment with continuous pump-administered intrathecal baclofen and achievement of individualised goals
- adverse effects of continuous pump-administered intrathecal baclofen that might be predicted by testing
- adverse effects that might be associated with intrathecal baclofen testing. [15]

1.6.6 Inform children and young people and their parents or carers verbally and in writing about continuous pump-administered intrathecal baclofen. The information should include all of the following:

- the surgical procedure used for implantation of the infusion pump
- the need for regular hospital follow-up visits
- requirements for pump maintenance
- risks associated with implantation of the pump, pump-related complications, and adverse effects that might be associated with continuous pump-administered intrathecal baclofen infusion.

[14] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

[15] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

1.6.7 Intrathecal baclofen testing should be:

- performed by a regional specialist centre that is able to carry out the necessary assessments
- undertaken in an inpatient setting to ensure safety and to allow a thorough assessment of outcomes. [16]

1.6.8 Before intrathecal baclofen testing, a pre-test assessment should be performed, including where necessary, an assessment of joint range of movement while the child or young person is under general anaesthesia.

1.6.9 The test dose or doses of intrathecal baclofen should be administered using a catheter inserted under general anaesthesia.[17]

1.6.10 Assess the response to intrathecal baclofen testing using standardised outcome measures within 3-5 hours of administration or later if the effects of the general anaesthetic have not worn off. Take account of individualised goals and the following criteria for a satisfactory response to intrathecal baclofen:

- reduction in spasticity or dystonia
- reduction in pain or muscle spasms
- improved posture and function, including head control
- improved self-care (or ease of care in the case of parents or carers).

1.6.11 Discuss with the child or young person and their parents or carers their subjective assessments of the response to intrathecal baclofen testing. Subjective assessments should include reports on self-care (or ease of care in the case of parents or carers).

[16] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

[17] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

Consider using a standardised questionnaire to document their assessments.

1.6.12 Pre- and post-test assessments should be performed by the same healthcare professionals in the regional specialist centre.

Continuous pump-administered intrathecal baclofen

1.6.13 Perform implantation of the infusion pump and start treatment with continuous pump-administered intrathecal baclofen within 3 months of a satisfactory response to intrathecal baclofen testing (see recommendation 1.6.10). [18]

1.6.14 Be aware of the following potential contraindications to treatment with continuous pump-administered intrathecal baclofen:

- the child or young person is too small to accommodate an infusion pump
- co-existing medical conditions (for example, uncontrolled epilepsy and coagulation disorders)
- intercurrent infections (systemic or around operative sites) which can increase the risks associated with continuous pump-administered intrathecal baclofen temporarily
- spinal fusion
- malnutrition which increases the risk of post-surgical complications (including infection and delayed healing)
- some respiratory conditions

1.6.15 Support children and young people receiving treatment with continuous pump-administered intrathecal baclofen and their parents or carers by offering regular follow-up and a consistent point of contact with the regional specialist centre.

[18] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

1.6.16 Monitor the response to continuous pump-administered intrathecal baclofen. Take account of individualised goals and the criteria for a satisfactory response to intrathecal baclofen (see recommendation 1.6.10).

1.6.17 Inform children and young people and their parents or carers verbally and in writing:

- about safe and effective management of continuous pump-administered intrathecal baclofen
- about the effects of intrathecal baclofen, possible adverse effects, and symptoms and signs suggesting the dose is too low or too high
- about safe and effective management of the infusion pump, including correct pump settings and the potential for pump-related complications
- that it is dangerous to stop the continuous pump-administered intrathecal baclofen infusion suddenly
- that the child or young person will need to attend hospital for follow-up appointments, for example to refill and reprogram the infusion pump
- that continuous pump-administered intrathecal baclofen should not be stopped before seeking advice from a healthcare professional.

1.6.18 If the response to continuous pump-administered intrathecal baclofen is unsatisfactory (see recommendation 1.6.10) offer continued support from the local multidisciplinary care team and consider referral for specialist support.

1.6.19 In children and young people with spasticity and co-existing scoliosis exercise caution and if the child or young person:

- has not yet undergone spinal fusion, implant the infusion pump before performing spinal fusion

- has undergone spinal fusion be aware that the operative procedure for implanting the pump will be more difficult technically and may not be possible.[19]

1.6.20 Titrate the dose of intrathecal baclofen after continuous pump-administered intrathecal baclofen pump implantation to optimise effectiveness and reassess the child or young person's achievement of their individualised goals.[20]

1.6.21 Repeat assessments after titration to determine the response to the new dose. The post-titration assessment should be performed by the same healthcare professionals in the regional specialist centre that performed the pre- and post-implantation assessments.

1.6.22 If treatment with continuous pump-administered intrathecal baclofen is judged to be unsatisfactory (see recommendation 1.6.10) and the infusion pump system has been confirmed to be working, consider reducing the dose gradually to determine whether spasticity and associated symptoms increase.

1.6.23 When the infusion pump is coming to the end of its lifespan, consider reducing the dose gradually to enable the child or young person to decide whether or not to have a new pump.[21]

[19] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.
[20] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.
[21] At the time of publication (October 2011), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years. Informed consent should be obtained and documented.

1.7 Orthopaedic surgery

Referral

1.7.1 Offer children and young people referral to an orthopaedic surgeon if there is clinical or radiological evidence of hip displacement or spinal deformity.

1.7.2 Consider referring a child or young person for an orthopaedic opinion if any of the following indications is present:

- the posture of an upper limb is causing difficulties with putting on or taking off clothing
- hand or upper limb function is limited by functionally short muscles (where spasticity prevents muscles stretching to their full length during functional tasks), pain or an unfavourable limb posture
- a contracture of the shoulder, elbow, wrist or hand causes difficulty with skin crease hygiene
- lower limb function is limited by functionally short muscles or an unfavourable limb posture
- walking is limited by functionally short lower limb muscles, joint contracture, abnormal torsion of the femur or tibia, foot deformity, or lower limb pain
- the cosmetic appearance of the upper limb causes significant concern for the child or young person.

1.7.3 Consider orthopaedic surgery as an adjunct to other interventions because timely surgery can prevent deterioration and ameliorate function.

Monitoring

1.7.4 Monitor children and young people to identify displacement of the hip and spinal deformity.

1.7.5 Clinically monitor all children and young people for signs of hip migration and recognise the following as evidence of hip displacement:

- abnormal hip migration percentage (more than 30%)
- increasing hip migration percentage
- deterioration in hip abduction
- pain arising from the hip
- reduced range of hip movement
- increased hip muscle tone
- decreased ability or tolerance for sitting or standing because of worsening hip joint contracture or bony deformity
- clinically important leg length difference
- increasing difficulty of perineal care or hygiene.

1.7.6 Perform a hip X-ray to monitor hip migration:

- by the age of 18 months in children with bilateral cerebral palsy
- in children with poor prognosis for walking (total body involved), delayed walking or who are using an external support for spastic diplegia
- in children or young people with signs of hip displacement (see recommendation 1.7.5).

1.7.7 Repeat the hip X-ray every 6 months in children and young people with hip migration percentage greater than 15% or in whom hip migration percentage is increasing by more than 10% per year.

Before undertaking orthopaedic surgery

1.7.8 Before undertaking orthopaedic surgery discuss and agree with the child or young person and their parents or carers a rehabilitation

programme and how and where it will be delivered. The programme may include:

- inpatient care and subsequent follow-up
- physical therapy
- orthoses
- other adjunctive treatments, such as oral drugs and botulinum toxin type A.

Performing orthopaedic surgery

1.7.9 Orthopaedic surgery should:

- be undertaken by surgeons experienced in the concepts and techniques of performing such surgery in this group of patients and
- take place in a paediatric setting.

1.7.10 Aim to perform single-event multilevel orthopaedic surgery to improve gait (rather than as staged surgical episodes) informed by a thorough preoperative functional assessment, preferably including a pre-operative gait analysis and interpretation of the results by a surgical team with experience in such analyses.

Assessment

1.7.11 Assess outcomes of gait-improvement orthopaedic surgery 1–2 years after performing the surgery. Use the same criteria for pre- and post-operative assessments.

1.8 Selective dorsal rhizotomy

1.8.1 Offer selective dorsal rhizotomy to improve walking ability only in the context of clinical research.

2 Notes on the scope of the guidance

NICE guidelines are developed in accordance with a scope that defines what the guideline will and will not cover. The scope of this guideline is available from www.nice.org.uk/[NICE to add details]. For the final guideline this should read, "The scope of this guideline is available from www.nice.org.uk/guidance/CG[XX] – click on 'How this guidance was produced'."

How this guideline was developed

NICE commissioned the National Collaborating Centre for Women's and Children's Health to develop this guideline. The Centre established a Guideline Development Group (see appendix A), which reviewed the evidence and developed the recommendations. An independent Guideline Review Panel oversaw the development of the guideline (see appendix B).

There is more information about how NICE clinical guidelines are developed on the NICE website (www.nice.org.uk/HowWeWork). A booklet, 'How NICE clinical guidelines are developed: an overview for stakeholders, the public and the NHS' (fourth edition, published 2009), is available from NICE publications (phone 0845 003 7783 or email publications@nice.org.uk and quote reference N1739).

3 Implementation

NICE has developed tools to help organisations implement this guidance (see www.nice.org.uk/guidance/CG[XX])'.

4 Research recommendations

The Guideline Development Group has made the following recommendations for research, based on its review of evidence, to improve NICE guidance and patient care in the future. The Guideline Development Group's full set of research recommendations is detailed in the full guideline (see section 5).

4.1 Selective dorsal rhizotomy

Does selective dorsal rhizotomy followed by intensive rehabilitation performed between the ages of 3 and 9 years in children who are in GMFCS level 2 or 3 result in good community mobility as a young adult?

Why this is important

The available evidence relating to selective dorsal rhizotomy suggests that the procedure results in some short- and medium-term improvements in motor function. The effects reported were not consistent across all studies nor sustained across all durations of follow-up investigated (6–24 months). The GDG considered that if the observed improvements could be maintained through to adult life then the outcomes of selective dorsal rhizotomy would be clinically important. Further research is urgently needed to evaluate long-term outcomes (including adverse effects) of selective dorsal rhizotomy followed by an intensive rehabilitation programme involving physical therapy (and prioritising targeted strength training) compared with physical therapy alone. The research could be conducted using a range of designs, including randomised controlled trials and audits of outcomes from procedures already performed. The research should focus on selective dorsal rhizotomy performed: between the ages of 3 and 9 years in children who are in GMFCS level 2 or 3 (because these children are likely to benefit most from selective dorsal rhizotomy); and before the development of significant contractures at the ankles, knees and hips. The research should: be coordinated through a multicentre research programme; use nationally agreed outcome measures (such as incidence of neurological impairment and spinal deformity, the need for additional operations, and assessment of disability, social inclusion and quality of life) and follow-up periods to facilitate national audit; include assessment of the child's clinical condition before and after selective dorsal rhizotomy using the same formally validated assessment techniques. The full guideline includes further considerations relating to criteria for identifying children who could be included in the research, the timing of selective dorsal rhizotomy in relation to other treatments such as orthopaedic surgery, and information that should be given to children and their parents or carers to facilitate informed decision making about participation in research.

4.2 Inhibitors of functional ability

What are the greatest inhibitors of functional ability in children and young people with upper motor neuron lesions?

Why this is important

Children and young people with upper motor neuron lesions may experience:

- reduced muscle strength
- selective muscle control
- spasticity.

The relationships between these factors, and the extent to which the child or young person can develop or maintain functional ability, remain unclear. Prospective cohort studies, or large cross-sectional studies, are needed to explore the relationships between positive and negative effects of upper motor neuron lesions and to determine which factor is the greatest inhibitor of functional ability. The studies should incorporate classification of functional ability based on validated scales, such as the gross motor functional classification system (GMFCS).

4.3 Postural management

What is the optimal postural management programme using a standing frame in children aged 1–3 years?

Why this is important

Children who are in GMFCS level 4 or 5 may benefit from using a standing frame as part of a postural management programme. Clinical benefits might include improved weight bearing and walking and, as a result, reduced hip migration. Postural management programmes involving the use of standing frames are part of established clinical practice. However, the individual elements that optimise the effectiveness of such programmes merit further research. The research should compare the effectiveness of postural management programmes that incorporate different durations and timings of standing frame use. For example, what is the effectiveness of 1 hour per day in a single session compared with two sessions of 30 minutes per day? The

research should be conducted in children and aged 1–3 years. These children are likely to benefit most from using standing frames (in terms of developing well-formed femoral heads and acetabulums) and they should find the use of standing frames acceptable (because they are lighter than older children, they do not have severe contractures and they are usually easily occupied).

4.4 Botulinum toxin type A

What is the effectiveness of botulinum toxin type A when used routinely or according to clinical need in children and young people who are in GMFCS levels 1 to 3?

Why this is important

The GDG's recommendation to consider offering botulinum toxin type A to children and young people with focal spasticity of an upper or lower limb reflected available evidence relating to the safety and effectiveness of botulinum toxin type A. In making their recommendations, the GDG emphasised the importance of establishing individualised functional goals that justify the use of this potentially harmful toxin to treat spasticity. The cost of the procedure combined with the risk of side effects means that clear treatment goals that will positively influence the child or young person's life should be identified before offering this treatment. The evidence reviewed for the guideline provided limited support for botulinum toxin type A in terms of improving function, and this discouraged the GDG from making a strong recommendation to offer treatment with botulinum toxin type A to all children and young people who are in GMFCS levels 1 to 3. Further research is needed to evaluate the effectiveness of botulinum toxin type A, particularly when used over long time periods (for example, 10 years) and involving repeat injections, in this population of children and young people. Outcomes relating to improvements in gross motor function and participation in activities, and the psychological impacts of these factors, should be evaluated as part of the research.

4.5 Intrathecal baclofen

What is the effectiveness of continuous pump-administered intrathecal baclofen compared with usual care in children and young people who are in GMFCS level 4 or 5?

Why this is important

The GDG's recommendation to consider offering continuous pump-administered intrathecal baclofen focused on children and young people in whom the use of appropriate non-invasive treatments did not relieve difficulties associated spasticity (specifically pain or muscle spasms, posture or function, or ease of care). Such children and young people will typically be in GMFCS level 4 or 5. Further research is needed to evaluate the clinical and cost effectiveness of continuous pump-administered intrathecal baclofen compared with usual care in these children and young people. Relevant research designs include randomised controlled trials, prospective cohort studies and qualitative studies. The outcomes to be investigated as part of the research include: quality of life; reduction of pain; reduction of tone; acceptability and tolerability; participation or inclusion; and adverse effects and their association with any potential predisposing factors.

5 Other versions of this guideline

5.1 Full guideline

The full guideline, 'Spasticity in children and young people with non-progressive brain disorders: management of spasticity and co-existing motor disorders and their early musculoskeletal complications' contains details of the methods and evidence used to develop the guideline. It is published by the National Collaborating Centre for Women's and Children's Health, and is available from our website (www.nice.org.uk/guidance/CG[XX]/Guidance).

Note: these details will apply to the published full guideline.

5.2 NICE pathway

The recommendations from this guideline have been incorporated into a NICE pathway, which is available from http://pathways.nice.org.uk/pathways/[xxx]

Note: these details will apply when the guideline is published.

5.3 'Understanding NICE guidance'

A summary for patients and carers ('Understanding NICE guidance') is available from www.nice.org.uk/guidance/CG[XX]/PublicInfo

For printed copies, phone NICE publications on 0845 003 7783 or email publications@nice.org.uk (quote reference number N[XXXX]). **Note: these details will apply when the guideline is published.**

We encourage NHS and voluntary sector organisations to use text from this booklet in their own information about spasticity and co-existing disorders caused by non-progressive brain disorders in children and young people.

6 Related NICE guidance

Published

- Selective dorsal rhizotomy for spasticity in cerebral palsy. NICE interventional procedure guidance 373 (2010). Available from www.nice.org.uk/guidance/IPG373

7 Updating the guideline

NICE clinical guidelines are updated so that recommendations take into account important new information. New evidence is checked 3 years after publication, and healthcare professionals and patients are asked for their views; we use this information to decide whether all or part of a guideline needs updating. If important new evidence is published at other times, we may decide to do a more rapid update of some recommendations. Please see our website for information about updating the guideline.

Appendix A: The Guideline Development Group, National Collaborating Centre and NICE project team

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Appendix B: The Guideline Review Panel

The Guideline Review Panel is an independent panel that oversees the development of the guideline and takes responsibility for monitoring adherence to NICE guideline development processes. In particular, the panel ensures that stakeholder comments have been adequately considered and responded to. The panel includes members from the following perspectives: primary care, secondary care, lay, public health and industry.

NICE to add

[Name; style = Unnumbered bold heading]
[job title and location; style = NICE normal]