Principles of care

Delivering care

Children and young people with spasticity should have access to a network of care that uses agreed care pathways supported by effective communication and integrated team working.

The network of care should provide access to a team of healthcare professionals experienced in the care of children and young people with spasticity. The network team should provide local expertise in paediatrics, nursing, physiotherapy and occupational therapy. Access to other expertise, including orthotics, orthopaedic surgery and paediatric neurology, may be provided locally or regionally.

If a child or young person receives treatment for spasticity from healthcare professionals outside the network team, this should be planned and undertaken in discussion with the network team to ensure integrated care and effective subsequent management.

Management programmes

Following diagnosis, ensure that all children and young people with spasticity are referred without delay to an appropriate member of the network team.

Offer a management programme that is:

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

When formulating a management programme take into account its possible impact on the individual child or young person and their family.

Carefully assess the impact of spasticity in children and young people with cognitive impairments:

- be aware that the possible benefit of treatments may be more difficult to assess in a child or young person with limited communication
- ensure that the child or young person has access to all appropriate services.

Identify and agree with children and young people and their parents or carers assessments and goals that:

- are age and developmentally appropriate
- focus on the following domains of the World Health Organization's International Classification of Functioning, Disability and Health:
  - body function and structure
  - activity and participation.

Record the child or young person's individualised goals and share these goals with healthcare professionals in the network team and, where appropriate, other people involved in their care.

Help children and young people and their parents or carers to be partners in developing and implementing the management programme by offering:

- relevant, and age and developmentally appropriate, information and educational materials
- regular opportunities for discussion **and**
- advice on their developmental potential and how different treatment options may affect this.

Monitoring

Monitor the child or young person's condition for:

- the response to treatments
- worsening of spasticity
- developing secondary consequences of spasticity, for example pain or contractures
- the need to change their individualised goals.

The network of care should have a pathway for monitoring children and young people at increased risk of hip displacement.

Recognise the following clinical findings as possible indicators of hip displacement (hip migration greater than 30%):

- pain arising from the hip
- clinically important leg length difference
- deterioration in hip abduction or range of hip movement
- increasing hip muscle tone
- deterioration in sitting or standing
- increasing difficulty with perineal care or hygiene.

Perform a hip X-ray to assess for hip displacement:

- if there are concerns about possible hip displacement
- at 24 months in children with bilateral cerebral palsy.

Consider repeating the hip X-ray annually in children or young people who are at Gross Motor Function Classification System (GMFCS) level III, IV or V.

Consider repeating the hip X-ray every 6 months in children and young people with a hip migration:

- greater than 15% **or**
- increasing by more than 10 percentage points per year.

Supporting the child or young person and their parents or carers

Offer contact details of patient organisations that can provide support, befriending, counselling, information and advocacy.

Ensure that children and young people have timely access to equipment necessary for their management programme (for example, postural management equipment such as sleeping, sitting or standing systems).

The network team should have a central role in transition to prepare young people and their parents or carers for the young person's transfer to adult services.

Physical therapy (physiotherapy and/or occupational therapy)

General principles

All children and young people with spasticity referred to the network team should be promptly assessed by a physiotherapist and, where necessary, an occupational therapist.

Offer a physical therapy (physiotherapy and/or occupational therapy) programme tailored to the child or young person's individual needs and aimed at specific goals, such as:

- enhancing skill development, function and ability to participate in everyday activities
- preventing consequences such as pain or contractures.

Give children and young people and their parents or carers verbal and written information about the physical therapy interventions needed to achieve the intended goals. This information should emphasise the balance between possible benefits and difficulties (for example, time commitment or discomfort), to enable them to participate in choosing a suitable physical therapy programme.

When formulating a physical therapy programme for children and young people take into account:

- the views of the child or young person and their parents or carers
- the likelihood of achieving the treatment goals
- possible difficulties in implementing the programme
- implications for the individual child or young person and their parents or carers, including the time and effort involved and potential individual barriers.

When deciding 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.

Ensure that any equipment or techniques used in the physical therapy programme are safe and appropriate, in particular in children or young people with any of the following:

- poorly controlled epilepsy
- respiratory compromise
- increased risk of pulmonary aspiration
- increased risk of bone fracture due to osteoporosis (for example, those who are non-ambulatory, malnourished or taking anticonvulsant therapy).

Encourage children and young people and their parents or carers to incorporate physical therapy into daily activities (for example, standing at the sink while brushing teeth in order to stretch leg muscles).

Specific strategies

Consider including in the physical therapy programme 24-hour postural management strategies to:

- prevent or delay the development of contractures or skeletal deformities in children and young people at risk of developing these
- enable the child or young person to take part in activities appropriate to their stage of development.

When using 24-hour postural management strategies consider, on an individual basis, the following techniques:

- low-load active stretching — the child or young person actively stretches their muscles with the aim of increasing range of movement
- low-load passive stretching — sustained stretching using positioning with equipment, orthoses or serial casting.

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

Consider task-focused active-use therapy such as constraint-induced movement therapy (temporary restraint of an unaffected arm to encourage use of the other arm) followed by bimanual therapy (unrestrained use of both arms) to enhance manual skills.

When undertaking task-focused active-use therapy consider an intensive programme over a short time period (for example, 4–8 weeks).

Consider muscle-strengthening therapy where the assessment indicates that muscle weakness is contributing to loss of function or postural difficulties.

Direct muscle-strengthening therapy towards specific goals using progressive repetitive exercises performed against resistance.

Following treatment with botulinum toxin type A, continuous pump-administered intrathecal baclofen or orthopaedic surgery, provide an adapted physical therapy programme as an essential component of management.

Ensure that children and young people and their parents or carers understand that following treatment with botulinum toxin type A treatment, continuous pump-administered intrathecal baclofen treatment or orthopaedic surgery an adapted physical therapy programme will be an essential component of management.

Continuing assessment

Reassess the physical therapy programme at regular intervals to ensure that:

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

Orthoses

General principles

Consider orthoses for children and young people with spasticity based on their individual needs and aimed at specific goals, such as:

- improving posture
- improving upper limb function
- improving walking efficienc
- preventing or slowing development of contractures
- preventing or slowing hip migration
- relieving discomfort or pain
- preventing or treating tissue injury, for example by relieving pressure points.

When considering an orthosis, discuss with the child or young person and their parents or carers the balance of possible benefits against risks. For example, discuss its cosmetic appearance, the possibility of discomfort or pressure sores or of muscle wasting through lack of muscle use.

Assess whether an orthosis 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 its appearance.

Ensure that orthoses are appropriately designed for the individual child or young person and are sized and fitted correctly. If necessary seek expert advice from an orthotist within the network team.

Be aware when considering a rigid orthosis that it may cause discomfort or pressure injuries in a child or young person with marked dyskinesia. They should be monitored closely to ensure that the orthosis is not causing such difficulties.

The network of care should have a pathway that aims to minimise delay in:

- supplying an orthosis once measurements for fit have been performed **and**
- repairing a damaged orthosis.

Inform children and young people who are about to start using an orthosis, and their parents or carers:

- how to apply and wear it
- when to wear it and for how long
    - an orthosis designed to maintain stretch to prevent contractures is more likely to be effective if worn for longer periods of time, for example at least 6 hours a day
    - an orthosis designed to support a specific function should be worn only when needed
- when and where to seek advice.

Advise children and young people and their parents or carers that they may remove an orthosis if it is causing pain that is not relieved despite their repositioning the limb in the orthosis or adjusting the strapping.

Specific uses

Consider the following orthoses 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 non-rigid thumb abduction splint allowing some movement for a child or young person with a ‘thumb in palm’ deformity).

Consider ankle–foot orthoses for children and young people with serious functional limitations (Gross Motor Function Classification System (GMFCS) level IV or V) to improve foot position for sitting, transfers between sitting and standing, and assisted standing.

Be aware that in children and young people with secondary complications of spasticity, for example contractures and abnormal torsion, ankle–foot orthoses may not be beneficial.

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

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

Consider ground reaction force 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.

Consider body trunk orthoses for children and young people with co-existing scoliosis or kyphosis if this will help with sitting.

Consider the overnight use of orthoses to:

- improve posture
- prevent or delay hip migration
- prevent or delay contractures.

Consider the overnight use of orthoses for muscles that control two joints. Immobilising the two adjacent joints provides better stretch and night-time use avoids causing functional difficulties.

If an orthosis is used overnight, check that it:

- is acceptable to the child or young person and does not cause injury
- does not disturb sleep.

Continuing assessment

Review the use of orthoses at every contact with the network team. Ensure that the orthosis:

- is still acceptable to the child or young person and their parents or carers
- remains appropriate to treatment goals
- is being used as advised
- remains well fitting and in good repair
- is not causing adverse effects such as discomfort, pain, sleep disturbance, injury or excessive muscle wasting.

Oral drugs

Consider oral diazepam in children and young people if spasticity is contributing to one or more of the following:

- discomfort or pain
- muscle spasms (for example, night-time muscle spasms)
- functional disability.

Diazepam is particularly useful if a rapid effect is desirable (for example, in a pain crisis).

Consider oral baclofen if spasticity is contributing to one or more of the following:

- discomfort or pain
- muscle spasms (for example, night-time muscle spasms)
- functional disability.

Baclofen is particularly useful if a sustained long-term effect is desired (for example, to relieve continuous discomfort or to improve motor function).

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

Give oral diazepam treatment as a bedtime dose. If the response is unsatisfactory consider:

- increasing the dose **or**
- adding a daytime dose.

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

Continue using oral diazepam or oral baclofen if they have a clinical benefit and are well tolerated, but think about stopping the treatment whenever the child or young person's management programme is reviewed and at least every 6 months.

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

If the response to oral diazepam and oral baclofen used individually for 4–6 weeks is unsatisfactory, consider a trial of combined treatment using both drugs.

If a child or young person has been receiving oral diazepam and/or baclofen for several weeks, ensure that when stopping these drugs the dose is reduced in stages to avoid withdrawal symptoms.

In children and young people with spasticity in whom dystonia is considered to contribute significantly to problems with posture, function and pain, consider a trial of oral drug treatment, for example with trihexyphenidyl,levodopa or baclofen.

Botulinum toxin type A

General principles

Consider botulinum toxin type A treatment in children and young people in whom 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 cosmetic concerns to the child or young person.

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

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

Consider botulinum toxin type A treatment after an acquired non-progressive brain injury if rapid-onset spasticity is causing postural or functional difficulties.

Consider a trial of botulinum toxin type A treatment in children and young people with spasticity in whom focal dystonia is causing serious problems, such as postural or functional difficulties or pain.

Do not offer botulinum toxin type A treatment if the child or young person:

- has severe muscle weakness
- had a previous adverse reaction or allergy to botulinum toxin type A
- is receiving aminoglycoside treatment.

Be cautious when considering botulinum toxin type A treatment if:

- the child or young person has any of the following
    - a bleeding disorder, for example due to anti-coagulant therapy
    - generalised spasticity
    - fixed muscle contractures
    - marked bony deformity **or**
- there are concerns about the child or young person's likelihood of engaging with the post-treatment adapted physical therapy programme.

When considering botulinum toxin type A treatment, perform a careful assessment of muscle tone, range of movement and motor function to:

- inform the decision as to whether the treatment is appropriate
- provide a baseline against which the response to treatment can be measured.

A physiotherapist or an occupational therapist should be involved in the assessment.

When considering botulinum toxin type A treatment, provide the child or young person and their parents or carers with information about:

- the possible benefits and the likelihood of achieving the treatment goals
- what the treatment entails, including:
    - the need for assessments before and after the treatment
    - the need to inject the drug into the affected muscles
    - the possible need for repeat injections
    - the benefits, where necessary, of analgesia, sedation or general anaesthesia
    - the need to use serial casting or an orthosis after the treatment in some cases
- important adverse effects.

Botulinum toxin type A treatment (including assessment and administration) should be provided by healthcare professionals within the network team who have expertise in child neurology and musculoskeletal anatomy.

Delivering treatment

Before starting treatment with botulinum toxin type A, tell children and young people and their parents or carers:

- to be aware of the following rare but serious complications of botulinum toxin type A treatment:
  - swallowing difficulties
  - breathing difficulties
- how to recognise signs suggesting these complications are present
- that these complications may occur at any time during the first week after the treatment and
- that if these complications occur the child or young person should return to hospital immediately.

To avoid distress to the child or young person undergoing treatment with botulinum toxin type A, think about the need for:

- topical or systemic analgesia or anaesthesia
- sedation (see 'Sedation in children and young people', NICE clinical guideline 112).

Consider ultrasound or electrical muscle stimulation to guide the injection of botulinum toxin type A.

Consider injecting botulinum toxin type A into more than one muscle if this is appropriate to the treatment goal, but ensure that maximum dosages are not exceeded.

After treatment with botulinum toxin type A, consider an orthosis to:

- enhance stretching of the temporarily weakened muscle and
- enable the child or young person to practice functional skills.

If the use of an orthosis is indicated after botulinum toxin type A, but limited passive range of movement would make this difficult, consider first using serial casting to stretch the muscle. To improve the child or young person's ability to tolerate the cast, and to improve muscle stretching, delay casting until 2–4 weeks after the botulinum toxin type A treatment.

Ensure that children and young people who receive treatment with botulinum toxin type A are offered timely access to orthotic services.

Continuing assessment

Perform a careful assessment of muscle tone, range of movement and motor function:

- 6–12 weeks after injections to assess the response
- 12–26 weeks after injections to inform decisions about further injections.

These assessments should preferably be performed by the same healthcare professionals who undertook the baseline assessment.

Consider repeat injections of botulinum toxin type A if:

- the response in relation to the child or young person's treatment goal was satisfactory, and the treatment effect has worn off
- new goals amenable to this treatment are identified.

Intrathecal baclofen

General principles

Consider treatment with continuous pump-administered intrathecal baclofen in children and young people with spasticity if, despite the use of non-invasive treatments, spasticity or dystonia are causing difficulties with any of the following:

- pain or muscle spasms
- posture or function
- self-care (or ease of care by parents or carers).

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

- moderate or severe motor function problems (Gross Motor Function Classification System (GMFCS) level III, IV or V)
- bilateral spasticity affecting upper and lower limbs.

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

- the child or young person is too small to accommodate an infusion pump
- local or systemic intercurrent infection.

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

- co-existing medical conditions (for example, uncontrolled epilepsy or coagulation disorders)
- a previous spinal fusion procedure
- malnutrition, which increases the risk of post-surgical complications (for example, infection or delayed healing)
- respiratory disorders with a risk of respiratory failure.

If continuous pump-administered intrathecal baclofen is indicated in a child or young person with scoliosis in whom a spinal fusion procedure is likely to be necessary, implant the infusion pump before performing the spinal fusion.

When considering continuous pump-administered intrathecal baclofen, balance the benefits of reducing spasticity against the risk of doing so because spasticity sometimes supports function (for example, by compensating for muscle weakness). Discuss these possible adverse effects with the child or young person and their parents or carers.

When considering continuous pump-administered intrathecal baclofen, inform children and young people and their parents or carers verbally and in writing about continuous pump-administered intrathecal baclofen. Give information about the following:

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

Intrathecal baclofen testing

Before making the final decision to implant the intrathecal baclofen pump, perform an intrathecal baclofen test to assess the therapeutic effect and to check for adverse effects.

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

- what the test will entail
- adverse effects that might occur with testing
- how the test might help to indicate the response to treatment with continuous pump-administered intrathecal baclofen, including whether:
    - the treatment goals are likely to be achieved
    - adverse effects might occur.

Before performing the intrathecal baclofen test, assess the following where relevant to the treatment goals:

- spasticity
- dystonia
- the presence of pain or muscle spasms
- postural difficulties, including head control
- functional difficulties
- difficulties with self-care (or ease of care by parents or carers).

If necessary, assess passive range of movement under general anaesthesia.

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

Assess the response to intrathecal baclofen testing within 3–5 hours of administration. If the child or young person is still sedated from the general anaesthetic at this point, repeat the assessment later when they have recovered.

When deciding whether the response to intrathecal baclofen is satisfactory, assess the following where relevant to the treatment goals:

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

Discuss with the child or young person and their parents or carers their views on the response to the intrathecal baclofen test. This should include their assessment of the effect on self-care (or ease of care by parents or carers). Consider using a standardised questionnaire to document their feedback.

Intrathecal baclofen testing should be:

- performed in a specialist neurosurgical centre within the network that has the expertise to carry out the necessary assessments
- undertaken in an inpatient setting to support a reliable process for assessing safety and effectiveness.

Initial and post-test assessments should be performed by the same healthcare professionals in the specialist neurosurgical centre.

Continuous pump-administered intrathecal baclofen

Before implanting the intrathecal baclofen pump, 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
- the effects of intrathecal baclofen, possible adverse effects, and symptoms and signs suggesting the dose is too low or too high
- the potential for pump-related complications
- the danger of stopping the continuous pump-administered intrathecal baclofen infusion suddenly
- the need to attend hospital for follow-up appointments, for example to refill and reprogram the infusion pump
- the importance of seeking advice from a healthcare professional with expertise in intrathecal baclofen before stopping the treatment.

Implant the infusion pump and start treatment with continuous pump-administered intrathecal baclofen within 3 months of a satisfactory response to intrathecal baclofen testing.

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 specialist neurosurgical centre.

Monitor the response to continuous pump-administered intrathecal baclofen. This monitoring should preferably be performed by the healthcare professionals in the regional specialist centre who performed the pre-implantation assessments.

When deciding whether the response to continuous pump-administered intrathecal baclofen is satisfactory, assess the following where relevant to the treatment goals:

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

Titrate the dose of intrathecal baclofen after pump implantation, if necessary, to optimise effectiveness.

If treatment with continuous pump-administered intrathecal baclofen does not result in a satisfactory response, check that there are no technical faults in the delivery system and that the catheter is correctly placed to deliver the drug to the intrathecal space. If no such problems are identified, consider reducing the dose gradually to determine whether spasticity and associated symptoms increase.

If continuous pump-administered intrathecal baclofen therapy is unsatisfactory, the specialist neurosurgical centre and other members of the network team should discuss removing the pump and alternative management options with the child or young person and their parents or carers.

As the infusion pump approaches the end of its expected lifespan, consider reducing the dose gradually to enable the child or young person and their parents or carers to decide whether or not to have a new pump implanted.

Orthopaedic surgery

Consider orthopaedic surgery as an important adjunct to other interventions in the management programme for some children and young people with spasticity. Timely surgery can prevent deterioration and improve function.

An assessment should be performed by an orthopaedic surgeon within the network team if:

- based on clinical findings or radiological monitoring, there is concern that the hip may be displaced
- based on clinical or radiological findings there is concern about spinal deformity.

Consider an assessment by an orthopaedic surgeon in the network team for children and young people with:

- hip migration greater than 15% **or**
- hip migration percentage increasing by more than 10 percentage points per year.

Consider an assessment by an orthopaedic surgeon in the network team if any of the following are present:

- limb function is limited (for example, in walking or getting dressed) by unfavourable posture or pain, as a result of muscle shortening, contractures or bony deformities
- contractures of the shoulder, elbow, wrist or hand cause difficulty with skin hygiene
- the cosmetic appearance of the upper limb causes significant concern for the child or young person.

Before undertaking orthopaedic surgery, the network team should discuss and agree with the child or young person and their parents or carers:

- the possible goals of surgery and the likelihood of achieving them
- what the surgery will entail, including any specific risks
- the rehabilitation programme, including:
    - how and where it will be delivered
    - what the components will be, for example a programme of adapted physical therapy, the use of orthoses, oral drugs or botulinum toxin type A.

Orthopaedic surgery should:

- be undertaken by surgeons in the network team who are expert in the concepts and techniques involved in surgery for this group of patients and
- take place in a paediatric setting.

The decision to perform orthopaedic surgery to improve gait should be informed by a thorough pre-operative functional assessment, preferably including gait analysis.

If a child or young person will need several surgical procedures at different anatomical sites to improve their gait, perform them together if possible (single-event multilevel surgery), rather than individually over a period of time,

Assess the outcome of orthopaedic surgery undertaken to improve gait 1–2 years later. By then full recovery may be expected and the outcome of the procedure can be more accurately determined.

Selective dorsal rhizotomy

Offer selective dorsal rhizotomy to improve walking ability in children and young people with spasticity only as part of a national research programme designed to collect data on standardised long-term outcomes.

The guideline will assume that prescribers will use a drug's summary of product characteristics (SPC) to inform decisions made with individual patients. Please refer to footnotes in the recommendations in the full guideline for information about the use of drugs outside their licensed indications.