

# Spasticity in under 19s: management

Clinical guideline

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## Your responsibility

The recommendations in this guideline represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, professionals and practitioners are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or the people using their service. It is not mandatory to apply the recommendations, and the guideline does not override the responsibility to make decisions appropriate to the circumstances of the individual, in consultation with them and their families and carers or guardian.

Local commissioners and providers of healthcare have a responsibility to enable the guideline to be applied when individual professionals and people using services wish to use it. They should do so in the context of local and national priorities for funding and developing services, and in light of their duties to have due regard to the need to eliminate unlawful discrimination, to advance equality of opportunity and to reduce health inequalities. Nothing in this guideline should be interpreted in a way that would be inconsistent with complying with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

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## Introduction

This guideline covers the management of spasticity and co-existing motor disorders and their early musculoskeletal complications in children and young people (from birth up to their 19th birthday) with non-progressive brain disorders.

Cerebral palsy is the most common condition associated with spasticity in children and young people. The incidence of cerebral palsy is not known, but its prevalence in the UK is 186 per 100,000 population, with a total of 110,000 people affected. The guideline covers the management of spasticity associated with cerebral palsy, but not all aspects of the management of cerebral palsy.

The impact of 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 the person's 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. Management should be tailored to meet the problems faced by the individual child or young person and their individual goals.

There is considerable variation in practice in managing spasticity, including variation in the availability of treatments and the intensity of their use. This guideline will help healthcare professionals to select and use appropriate treatments for individual children and young people.

The guideline will assume that prescribers will use a drug's summary of product characteristics (SPC) to inform decisions made with individual patients.

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.

## Person-centred care

This guideline offers best practice advice on the management of spasticity and co-existing motor disorders and their early musculoskeletal complications in children and young people with non-progressive brain disorders.

Treatment and care should take into account the needs and preferences of children, young people and those who care for them. Children and young people and, as appropriate, their parents or carers, should have the opportunity to make informed decisions about their care and treatment, in partnership with their healthcare professionals. If children and young people do not have the capacity to make decisions, healthcare professionals should follow the [Department of Health's advice on consent](#) and the [code of practice that accompanies the Mental Capacity Act](#). In Wales, healthcare professionals should follow [advice on consent from the Welsh Government](#).

If the child or young person is under 16, healthcare professionals should follow the guidelines in the Department of Health's [Seeking consent: working with children](#).

Good communication between healthcare professionals and children and young people is essential. It should be supported by evidence-based written information tailored to the needs of the child or young person and their parents or carers. Treatment and care, and the information people 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 the Department of Health's [Transition: getting it right for young people](#).

Adult and paediatric healthcare teams should work jointly to provide assessment and services to young people who have spasticity and co-existing motor disorders and their early musculoskeletal complications as a result of a non-progressive brain disorder. 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.

## Terms used in this guideline

### *Botulinum toxin type A*

A neurotoxin produced by the bacterium *Clostridium botulinum* that blocks neurotransmitter release at peripheral cholinergic nerve terminals. Injection into a muscle reduces spasticity.

### *Constraint-induced movement therapy*

An approach to physical therapy in which an unaffected arm is temporarily restrained to encourage use of the other arm.

### *Continuous pump-administered intrathecal baclofen treatment*

Direct administration of baclofen into the fluid-filled space around the spinal cord (the intrathecal space) using a catheter and infusion pump. The pump is implanted in the abdominal cavity and allows a continual controlled delivery of baclofen adjusted according to need.

### *Contracture*

Shortening of muscle tendons, ligaments and soft tissues resulting in a limitation of joint movement. Usually, muscle shortening is the primary abnormality, but prolonged immobility or scarring may also contribute.

### *Dystonia*

Involuntary, sustained, or intermittent muscle contractions that cause twitching and repetitive movements, abnormal postures or both.

### *Equinus deformity*

Abnormal ankle plantarflexion (movement of the foot at the ankle joint in a downward direction). This can, for example, result in the child or young person walking on tiptoe.

### *Fine motor function*

The ability to use small muscle groups, often in coordination with the eyes, to perform precision activities such as writing or fastening buttons.

## *Focal dystonia*

Dystonia involving a specific muscle or group of muscles.

## *Focal spasticity*

Spasticity involving a specific muscle or group of muscles.

## *Function*

The ability to perform normal activities or actions. Such function may be impaired by spasticity and associated motor disorders and by the complications of spasticity.

## *Gait analysis*

A detailed approach to analysing the component phases of walking using instrumentation or video analysis in addition to clinical observation. This is undertaken to evaluate a child or young person's ability and style of walking and to plan or assess treatment.

## *Gross motor function*

The ability to use large muscle groups to perform body movements such as sitting, standing, walking and running.

## *Gross Motor Function Classification System*

A 5-point scale that describes gross motor function: level I, walks without restrictions; level II, walks without assistive devices; level III, walks with assistive devices; level IV, has limited self-mobility; level V, has severely limited self-mobility even with assistive devices.

## *Hip migration*

Movement of the top of the thigh bone that connects with the pelvis (the femoral head) from its normal position in the socket joint of the hip (the acetabulum). This movement is often measured by reporting the degree of displacement seen on X-ray (known as the hip migration percentage).



### *Intrathecal baclofen testing*

Direct injection of baclofen into the fluid-filled space around the spinal cord (the intrathecal space) using a lumbar puncture needle or a temporary spinal catheter in order to assess the likely response to continuous pump-administered baclofen treatment.

### *Kyphosis*

Abnormal curvature of the spine when viewed from the side of the body that results in a hunched or slouching position.

### *Low-load active stretching*

A physical therapy intervention in which the child or young person actively stretches their muscles with the aim of increasing range of movement.

### *Low-load passive stretching*

A physical therapy intervention involving sustained stretching using positioning with equipment, orthoses or serial casting.

### *Muscle tone*

The normal state of continuous passive partial contraction in a resting muscle. Muscle tone is important in maintaining posture. Increased muscle tone (hypertonia) is associated with an abnormal resistance to passive stretch, while reduced muscle tone (hypotonia) is associated with floppiness of the limbs or trunk and poor posture.

### *Network of care*

Linked groups of healthcare professionals and organisations working in an agreed and coordinated manner to deliver a clinical service. A network is not constrained by existing professional, organisational or institutional boundaries.

### *Network team*

A multidisciplinary group of healthcare and other professionals working in a network of care to deliver a clinical service.

## *Orthosis (plural, orthoses)*

An artificial device or appliance used to support, align, prevent, or correct deformities or to improve musculoskeletal function.

## *Passive range of movement*

The degree of motion through which a joint can be moved by an outside force without active participation by the child or young person themselves (for example, movement by another person).

## *Range of movement*

The range of motion, usually measured in degrees, through which a joint can move.

## *Scoliosis*

An abnormal lateral curvature of the spine viewed from in front of or behind the child or young person.

## *Secondary complication of spasticity*

An adverse effect on musculoskeletal structure that occurs as a result of spasticity (for example, a contracture or abnormal torsion).

## *Secondary consequence of spasticity*

Any effect experienced by a child or young person as a result of spasticity. This may be symptomatic (for example, pain or difficulty walking) or a complication affecting the structure of the musculoskeletal system (see secondary complication of spasticity).

## *Selective dorsal rhizotomy*

A neurosurgical procedure in which some of the sensory nerves that contribute to spasticity in the lower limb are cut at the point where they enter the spinal cord.

## *Serial casting*

The successive use of casts with the aim of progressively lengthening muscles and other non-bony tissues such as ligaments and tendons thereby reducing the effect of contractures by passive stretching to gradually improve the range of movement.

## *Spasticity*

A specific form of increased muscle tone (hypertonia) in which one or both of the following are present:

- the resistance to externally imposed movement increases with increasing speed of stretch and varies with the direction of joint movement
- the resistance to externally imposed movement increases rapidly beyond a threshold speed or joint angle.

## *Spinal fusion*

A surgical procedure where two or more vertebrae are joined to prevent movement between them.

## *Task-focused active-use therapy*

A physiotherapy technique where a specific goal is identified and the child or young person carries out exercises or activities using the affected limb or limbs to improve their performance.

## Key priorities for implementation

The following recommendations have been identified as priorities for implementation.

### Principles of 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.
- 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.
- 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.
- 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.
- 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.

### Physical therapy (physiotherapy and/or occupational therapy)

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

### Intrathecal baclofen

- Consider treatment with continuous pump-administered intrathecal baclofen<sup>[1]</sup> 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).

### Orthopaedic surgery

- An assessment should be performed by an orthopaedic surgeon within the network team if:
  - based on clinical findings (see recommendation 1.1.16) or radiological monitoring, there is concern that the hip may be displaced
  - based on clinical or radiological findings there is concern about spinal deformity.

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<sup>[1]</sup> At the time of publication (July 2012), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years, nor did it have UK marketing authorisation for use in the treatment of dystonia associated with spasticity. Where appropriate, informed consent should be obtained and documented.

## 1 Guidance

The following guidance is based on the best available evidence. The [full guideline](#) gives details of the methods and the evidence used to develop the guidance.

### 1.1 *Principles of care*

#### Delivering care

- 1.1.1 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.
- 1.1.2 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/or neurosurgery and paediatric neurology, may be provided locally or regionally.
- 1.1.3 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

- 1.1.4 Following diagnosis, ensure that all children and young people with spasticity are referred without delay to an appropriate member of the network team.
- 1.1.5 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.

- 1.1.6 When formulating a management programme take into account its possible impact on the individual child or young person and their family.
- 1.1.7 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.
- 1.1.8 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 functions
    - body structures
    - activities and participation
    - environmental factors.
- 1.1.9 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.
- 1.1.10 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.

## Supporting the child or young person and their parents or carers

- 1.1.11 Offer contact details of patient organisations that can provide support, befriending, counselling, information and advocacy.
- 1.1.12 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).
- 1.1.13 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.

## Monitoring

- 1.1.14 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.
- 1.1.15 The network of care should have a pathway for monitoring children and young people at increased risk of hip displacement.
- 1.1.16 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.



1.1.17 Offer a hip X-ray to assess for hip displacement:

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

1.1.18 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.

1.1.19 Consider repeating the hip X-ray after 6 months in children and young people where the initial hip migration is greater than 30%, and then consider repeating the hip X-ray every 6 months after this if the hip migration is increasing by more than 10 percentage points per year.

## 1.2 *Physical therapy (physiotherapy and/or occupational therapy)*

### General principles

1.2.1 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.

1.2.2 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.

1.2.3 Give children and young people and their parents or carers verbal and written (or appropriate formats) 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.

1.2.4 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.

1.2.5 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.

1.2.6 Ensure that any equipment or techniques used in the physical therapy programme are safe and appropriate, in particular for 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 unable to walk, malnourished or taking anti-epileptic therapy).

1.2.7 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

1.2.8 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.
- 1.2.9 When using 24-hour postural management strategies consider on an individual basis low-load active stretching or low-load passive stretching.
- 1.2.10 Offer training to parents and carers involved in delivering postural management strategies.
- 1.2.11 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.
- 1.2.12 When undertaking task-focused active-use therapy consider an intensive programme over a short time period (for example, 4–8 weeks).
- 1.2.13 Consider muscle-strengthening therapy where the assessment indicates that muscle weakness is contributing to loss of function or postural difficulties.
- 1.2.14 Direct muscle-strengthening therapy towards specific goals using progressive repetitive exercises performed against resistance.
- 1.2.15 Following treatment with botulinum toxin type A, continuous pump-administered intrathecal baclofen, orthopaedic surgery or selective dorsal rhizotomy, provide an adapted physical therapy programme as an essential component of management.
- 1.2.16 Ensure that children and young people and their parents or carers understand that an adapted physical therapy programme will be an essential component of management following treatment with botulinum toxin type A, continuous pump-administered intrathecal baclofen, orthopaedic surgery or selective dorsal rhizotomy.

## Continuing assessment

- 1.2.17 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.

## 1.3 Orthoses

### General principles

1.3.1 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 efficiency
- 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.

1.3.2 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.

1.3.3 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.

1.3.4 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.

- 1.3.5 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.
- 1.3.6 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.
- 1.3.7 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.
- 1.3.8 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

- 1.3.9 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).
- 1.3.10 Consider ankle-foot orthoses for children and young people with serious functional limitations (GMFCS level IV or V) to improve foot position for sitting, transfers between sitting and standing, and assisted standing.
- 1.3.11 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.
- 1.3.12 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.
- 1.3.13 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.
- 1.3.14 Consider body trunk orthoses for children and young people with co-existing scoliosis or kyphosis if this will help with sitting.
- 1.3.15 Consider the overnight use of orthoses to:
- improve posture
  - prevent or delay hip migration
  - prevent or delay contractures.
- 1.3.16 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.
- 1.3.17 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

1.3.18 The network team should review the use of orthoses at every contact with the child or young person. 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.

## 1.4 Oral drugs

1.4.1 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).

1.4.2 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).

- 1.4.3 If oral diazepam is initially used because of its rapid onset of action, consider changing to oral baclofen if long-term treatment is indicated.
- 1.4.4 Give oral diazepam treatment as a bedtime dose. If the response is unsatisfactory consider:
- increasing the dose or
  - adding a daytime dose.
- 1.4.5 Start oral baclofen treatment with a low dose and increase the dose stepwise over about 4 weeks to achieve the optimum therapeutic effect.
- 1.4.6 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.
- 1.4.7 If adverse effects (such as drowsiness) occur with oral diazepam or oral baclofen, think about reducing the dose or stopping treatment.
- 1.4.8 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.
- 1.4.9 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.
- 1.4.10 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<sup>[2]</sup>, levodopa<sup>[3]</sup> or baclofen<sup>[4]</sup>.



## 1.5 *Botulinum toxin type A*

### General principles

1.5.1 Consider botulinum toxin type A<sup>[5]</sup> 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.

1.5.2 Consider botulinum toxin type A<sup>[5]</sup> 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.

1.5.3 Consider botulinum toxin type A<sup>[5]</sup> treatment after an acquired non-progressive brain injury if rapid-onset spasticity is causing postural or functional difficulties.

1.5.4 Consider a trial of botulinum toxin type A<sup>[5]</sup> treatment in children and young people with spasticity in whom focal dystonia is causing serious problems, such as postural or functional difficulties or pain.

1.5.5 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.

1.5.6 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 (see recommendation 1.2.15).

1.5.7 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.

1.5.8 When considering botulinum toxin type A treatment, give the child or young person and their parents or carers 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

- possible important adverse effects (see also recommendation 1.5.10).

1.5.9 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

1.5.10 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.

1.5.11 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).

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

1.5.13 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.

1.5.14 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.

1.5.15 If 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.

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

## Continuing assessment

1.5.17 Perform an 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.

1.5.18 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.

## 1.6 Intrathecal baclofen

### General principles

1.6.1 Consider treatment with continuous pump-administered intrathecal baclofen<sup>[7]</sup> 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).

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

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

1.6.3 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.

1.6.4 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.

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

1.6.6 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.

1.6.7 When considering continuous pump-administered intrathecal baclofen, inform children and young people and their parents or carers verbally and in writing (or appropriate formats) about:

- 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

1.6.8 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.

1.6.9 Before intrathecal baclofen testing, inform children and young people and their parents or carers verbally and in writing (or appropriate formats) 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.

1.6.10 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.

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

1.6.12 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.

1.6.13 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).

1.6.14 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.

1.6.15 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.

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

## Continuous pump-administered intrathecal baclofen

1.6.17 Before implanting the intrathecal baclofen pump, inform children and young people and their parents or carers, verbally and in writing (or appropriate formats), 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.

1.6.18 Implant 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 1.6.13).

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

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



- 1.6.21 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).
- 1.6.22 Titrate the dose of intrathecal baclofen after pump implantation, if necessary, to optimise effectiveness.
- 1.6.23 If treatment with continuous pump-administered intrathecal baclofen does not result in a satisfactory response (see recommendation 1.6.21), 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.
- 1.6.24 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.
- 1.6.25 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.

## 1.7 *Orthopaedic surgery*

- 1.7.1 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.

- 1.7.2 An assessment should be performed by an orthopaedic surgeon within the network team if:
- based on clinical findings (see recommendation 1.1.16) or radiological monitoring, there is concern that the hip may be displaced
  - based on clinical or radiological findings there is concern about spinal deformity.
- 1.7.3 Consider an assessment by an orthopaedic surgeon in the network team for children and young people with:
- hip migration greater than 30% or
  - hip migration percentage increasing by more than 10 percentage points per year.
- 1.7.4 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.
- 1.7.5 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.
- 1.7.6 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.

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

1.7.8 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.

1.7.9 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.

## 1.8 *Selective dorsal rhizotomy*

1.8.1 Consider selective dorsal rhizotomy to improve walking ability in children and young people with spasticity at GMFCS level II or III:

- Patient selection and treatment should be carried out by a multidisciplinary team with specialist training and expertise in the care of spasticity, and with access to the full range of treatment options.
- Discuss the irreversibility of the treatment, the known complications and the uncertainties over long-term outcomes with children and young people, and their parents and/or carers (see also [Selective dorsal rhizotomy for spasticity in cerebral palsy](#), NICE interventional procedure guidance 373).
- Teams offering selective dorsal rhizotomy should participate in a coordinated national agreed programme to collect information on short- and long-term outcomes on all patients assessed for selective dorsal rhizotomy, whether or not selective dorsal rhizotomy is performed. These recorded outcomes should include measures of muscle tone, gross motor function, neurological impairment, spinal deformity, quality of life and need for additional operations, with nationally agreed consistent definitions.

## More information

You can also see this guideline in the NICE pathway on [spasticity in children and young people](#). To find out what NICE has said on topics related to this guideline, see our web pages on [neurological conditions](#) and [children and young people](#).

<sup>[2]</sup> At the time of publication (July 2012), trihexyphenidyl did not have UK marketing authorisation for use in the treatment of dystonia associated with spasticity, and its use is not recommended in children. However, it is used in the UK for the treatment of dystonia in children and young people with spasticity. Informed consent should be obtained and documented.

<sup>[3]</sup> At the time of publication (July 2012), levodopa (which is always marketed in combination with an extra-cerebral dopa-decarboxylase inhibitor) did not have UK marketing authorisation for use in the treatment of dystonia associated with spasticity, and its use is not recommended in children or young people. However, it is used in the UK for the treatment of dystonia in children and young people with spasticity. Informed consent should be obtained and documented.

<sup>[4]</sup> At the time of publication (July 2012), baclofen did not have UK marketing authorisation for use in the treatment of dystonia associated with spasticity. However, it is used in the UK for the treatment of dystonia in children and young people with spasticity. Informed consent should be obtained and documented.

<sup>[5]</sup> At the time of publication (July 2012), some botulinum toxin type A products had UK marketing authorisation for use in the treatment of focal spasticity in children, young people and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, 2 years of age or older. Other products had UK marketing authorisation only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Botulinum toxin units are not interchangeable from one product to another. Details of licensed indications and doses for individual products are available at the [electronic Medicines Compendium](#). Where appropriate, informed consent should be obtained and documented.

<sup>[6]</sup> At the time of publication (July 2012), botulinum toxin type A did not have UK marketing authorisation for use in the treatment of focal dystonia associated with spasticity. However, it is used in the UK for the treatment of dystonia in children and young people with spasticity. Informed consent should be obtained and documented.

<sup>[7]</sup> At the time of publication (July 2012), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years, nor did it have UK marketing authorisation for use in the treatment of dystonia associated with spasticity. Where appropriate, informed consent should be obtained and documented.

## 2 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](#).

### 2.1 *Inhibitors of functional ability*

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

#### Why this is important

Children and young people with upper motor neurone 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 neurone 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 GMFCS.

### 2.2 *Botulinum toxin type A*

What is the clinical and cost effectiveness of botulinum toxin type A when used routinely or according to clinical need in children and young people who are at GMFCS level I, II or III?

#### Why this is important

The Guideline Development Group's (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 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 achieving clinically important goals (including those related to 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 at GMFCS level I, II or III. Further research is needed to evaluate the effectiveness of botulinum toxin type A in comparison with other treatment options, 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.

### 2.3 *Intrathecal baclofen*

What is the clinical and cost effectiveness of continuous pump-administered intrathecal baclofen compared with usual care in children and young people who are at GMFCS level IV or V?

#### **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 with spasticity (specifically pain or muscle spasms, posture or function, or ease of care). Such children and young people will typically be at GMFCS level IV or V. 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.

### 2.4 *Selective dorsal rhizotomy*

Does selective dorsal rhizotomy followed by intensive rehabilitation performed between the ages of 3 and 9 years in children who are at GMFCS level II or III 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 at GMFCS level II or III (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; and 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.



## Update information

**November 2016:** Recommendation 1.1.8 was amended to update information on the World Health Organization's International Classification of Functioning, Disability and Health (ICF) and its domains. An outdated research recommendation was deleted.

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## Accreditation

