

NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE

SCOPE

1 **Guideline title**

Spasticity in children: the management of spasticity in children and young people with a non-progressive brain injury

Short title

Spasticity in children and young people

2 **The remit**

The Department of Health has asked NICE: 'To prepare a clinical guideline on the management of spasticity in children with a non-progressive brain injury'.

3 **Clinical need for the guideline**

3.1 *Epidemiology*

- a) Spasticity is defined as 'a motor disorder characterised by hyperexcitability of the stretch reflex, resulting in a velocity-dependent increase in tonic stretch reflexes (muscle tone) with exaggerated tendon jerk. It is one component of the upper motor neuron syndrome'.
- b) It is a common and often serious disorder of motor function. Normally when a muscle stretches, a reflex is triggered to pull this muscle back to its 'resting' state. In spasticity, a hyperactive response to the stretch reflex or a or resistance to passive movement results in a rapid and strong contraction of the muscle, causing stiffness. This dysregulation of muscle tone can result in a wide range of clinical manifestations and functional impairments.

- c) Spasticity in children is most often seen in cerebral palsy, although it can also occur with other forms of non-progressive and progressive brain damage; the latter is outside the remit of this guideline.
- d) Cerebral palsy is defined as 'a clinical syndrome characterised by a persistent disorder of posture or movement due to a non-progressive disorder of the immature brain. Examples of non-progressive damage that may affect the fetal or infant brain include brain malformations, prenatal vascular events (stroke) and infections (such as cytomegalovirus), perinatal hypoxic or ischaemic encephalopathy, and postnatal head injury or meningitis.
- e) The prevalence of cerebral palsy in the UK is about 2 per 1000 live births. This figure has not changed significantly in the past 40 years. Around 40% of children with cerebral palsy were born prematurely. In many of these children the precise cause of cerebral palsy is not apparent, but various risk factors can be identified, including maternal illness and postnatal events.
- f) Although in cerebral palsy the causative brain damage is static, the motor manifestations change over time. Typically, abnormalities of movement and posture are first recognised during infancy or early childhood.
- g) Up to 80% of children with cerebral palsy have spasticity. Other types of cerebral palsy include dyskinetic (with athetosis, dystonia and chorea) and ataxic (with abnormalities of coordination and balance). It is quite common for children with spastic cerebral palsy to also have other motor disorders such as dystonia or ataxia.
- h) Non-progressive brain damage may also occur in older children, for example, from head trauma, encephalitis or meningitis. Brain damage of this type is not by definition cerebral palsy, but it can result in motor disorders, including spasticity.

- i) Depending on which parts of the motor cortex are damaged, the imbalance between flexor and extensor muscles may lead to abnormal posture of the joints. It is important to distinguish dynamic postural abnormalities (due to muscle spasticity) from fixed contractures (muscles that have become permanently shortened after long-term spasticity).
- j) Spasticity and cerebral palsy in particular can be categorised according to the parts of the body affected.
- In diplegia the lower limbs are markedly affected with mild involvement of the upper limbs. This pattern is common among low-birth-weight infants who have cerebral palsy, and accounts for about 50% of cases of spastic cerebral palsy. Although therapeutic intervention is required, most children with diplegia and good cognitive function learn to walk independently.
 - In quadriplegia all four limbs are affected, together with the torso and oropharyngeal musculature. There may be extensive cerebral involvement. This pattern of spasticity is sometimes described as 'total body involvement' and accounts for about 30% of spastic cerebral palsy. Although the severity varies considerably, most children do not learn to walk. Many have severe impairment of motor function, together with other manifestations of neurological dysfunction including cognitive impairment, seizures, and lack of bladder and bowel control.
 - In hemiplegia one side of the body is affected, with the upper limb usually more markedly affected than the lower limb. This pattern accounts for about 20% of spastic cerebral palsy.
Children with hemiplegia usually walk independently by 3 years.
- k) The functional abilities of children with spasticity often deteriorate over time. This deterioration is believed to be primarily and specifically caused by the spasticity rather than other commonly associated effects such as muscle weakness (paresis). Effective

treatment of spasticity could therefore be important in preventing functional decline.

- l) The muscular imbalances associated with spasticity often result in abnormal posture, which is initially 'dynamic' with the potential to improve with effective treatment of spasticity. In time the abnormal posturing can become permanent because of contractures, which in turn, may cause fixed joint deformities. Uncorrected deformities in spastic cerebral palsy can cause pain, impair function and reduce mobility, causing difficulties in care for the child. Subluxation or full dislocation of joints can occur. Significant bony deformities can occur, such as kyphosis and scoliosis of the spine.
- m) These changes may substantially worsen the child's functional disability and impair the ability to walk or sit. Children and young people may avoid walking if it becomes unsafe or uncomfortable or if it requires a large effort. Abnormal posturing of the shoulder, elbow, or hand may greatly impair the function of the upper limb with a consequent reduction in the individual's independence, for example in dressing or toileting. A lack of independence leads to an increased need for support by paid carers or family members. It may also reduce employment opportunities.
- n) Successful treatment of spasticity might lead to better motor function, reduction or prevention of contractures and other fixed musculoskeletal deformities, enhanced functional abilities and independence, and ultimately an improvement in the person's quality of life.

3.2 *Current practice*

- a) The aims of managing spasticity are to minimise disability, to promote independence and to achieve as complete an integration into society as possible for the affected child or young person.

- b) Many treatments are used in the management of spasticity, with considerable variation in practice.
- c) Many physiotherapy regimens are commonly used in children with spasticity. These include passive stretching, therapeutic exercises and encouraging the use of compensating movements.
- d) Bracing is common, using a wide variety of orthoses, designed – for example – to hold limbs in an advantageous position to improve functionality and to prevent or treat deformity. Ankle–foot orthoses of various sorts are frequently used. Similar devices are also used to immobilise the knee or to encourage hip abduction. Less frequently, upper limb orthoses may be employed.
- e) Spasticity may be alleviated by a wide range of interventions aimed at modulating the abnormal stretch reflex:
- Oral anti-spastic medications such as baclofen may be used in those with extensive spasticity.
 - Intrathecal baclofen is administered into the cerebrospinal fluid using an implanted pump. It is used for severe spasticity.
 - Local injection with botulinum toxin A may be effective. This works by temporarily blocking the release of the neuromuscular transmitter acetylcholine. There is uncertainty about the optimum regimen for administering this agent, and controversy regarding its benefit in terms of outcomes that are important to patients.
 - Selective dorsal rhizotomy is used to reduce spasticity in the legs by interruption of the spinal reflex, and is covered by ‘Selective dorsal rhizotomy for spasticity in cerebral palsy’, NICE interventional procedure guidance 195 (2006). This procedure has potential adverse effects such as hip instability and spinal deformity.
- f) Orthopaedic surgery has a major role in the management of spasticity. Muscle–tendon lengthening procedures can both release shortened muscles and weaken spastic muscle, thereby improving

the balance of forces influencing joint position. Osteotomy procedures can correct deformities and stabilise hip dislocation. Rotational osteotomy can correct torsional deformities and relieve malaligned muscular forces. Spinal deformities can be treated with fusion and instrumentation techniques. Disorders such as pes equinus and pes varus, scissoring and hip instability can be managed using such techniques. Hip subluxation or dislocation occur in up to 25% of children with cerebral palsy and surgery can be helpful to stabilise joints. Surgical procedures can alleviate many of the consequences of spasticity, resulting in significant functional improvement.

Expertise in and access to various types of treatment varies. Bracing techniques may be employed inappropriately or without evidence of benefit. Conversely, in some areas orthoses are not funded. Placement of intrathecal baclofen pumps is available in certain regional centres only.

4 The guideline

The guideline development process is described in detail on the NICE website (see section 6, 'Further information').

This scope defines what the guideline will (and will not) examine, and what the guideline developers will consider. The scope is based on the referral from the Department of Health.

The areas that will be addressed by the guideline are described in the following sections.

4.1 *Population*

4.1.1 Groups that will be covered

- a) Children and young people from birth up to their 19th birthday who have spasticity as a result of a non-progressive brain injury. It will include those with spasticity resulting from cerebral palsy and those

with spasticity resulting from a non-progressive brain injury acquired later in childhood or adolescence.

- b) Subgroups of this population will be considered in relation to the areas affected by spasticity:
- monoplegia
 - diplegia
 - hemiplegia
 - quadriplegia.

4.1.2 Groups that will not be covered

- a) Adults 19 years and older.
- b) Children and young people with spasticity resulting from a progressive brain disorder. However, many of the recommendations on the management of spasticity might also apply to these children.
- c) Children and young people with comorbidities or other motor disorders such as dystonia. These children will be included in relation to the management of spasticity alone. Awareness of dystonia is important in treating spasticity; if appropriate the guideline may flag changes to the management of spasticity for people who have dystonia.

4.2 *Healthcare setting*

- a) All settings in which NHS care is provided.

4.3 *Clinical management*

4.3.1 Key clinical issues that will be covered

Unless otherwise stated, each issue below will be considered in relation to the subgroups of people with spastic monoplegia, diplegia, hemiplegia and quadriplegia. If clinically appropriate each issue will also be considered in relation to the severity of spasticity: mild, moderate or severe.

- a) Physiotherapy interventions to reduce spasticity and prevent its direct musculoskeletal consequences, for example, muscle shortening.
- b) Orthoses (for example, ankle-foot orthoses, knee splints, and upper limb orthoses) for preventing and treating contractures and improving function (such as mobility).
- c) Oral medications specifically baclofen, benzodiazepines (diazepam, nitrazepam, clonazepam) tizanidine and dantrolene to reduce spastic diplegia, hemiplegia and quadriplegia (not spastic monoplegia).
- d) Long-term use of intramuscular botulinum toxin A to reduce spasticity, maintain motor function and prevent secondary complications.
- e) Whether an effective response to a bolus dose of intrathecal baclofen predicts an effective long-term response in children with moderate to severe spastic diplegia, hemiplegia and quadriplegia (not spastic monoplegia).
- f) The intrathecal baclofen pump to reduce spasticity, maintain motor function and improve quality of life in children with moderate to severe spastic diplegia, hemiplegia and quadriplegia (not spastic monoplegia).
- g) Orthopaedic surgery specifically (tendon lengthening and transfer procedures, and osteotomy) to prevent and correct deformities and prevent joint dislocations.
- h) Multilevel surgery (multiple surgical procedures done at the same time) compared with interval surgery (consecutive operations) to improve quality of life in children with severe spastic diplegia or hemiplegia (not spastic monoplegia and quadriplegia).

4.3.2 Clinical issues that will not be covered

- a) Diagnosis and assessment of spasticity.
- b) Management of spasticity caused by a progressive brain injury or a spinal cord injury.
- c) Holistic management of cerebral palsy or other associated conditions.
- d) Play therapy such as hippotherapy.
- e) Complementary and alternative therapies.
- f) Selective dorsal rhizotomy.
- g) Management of the following complications:
 - wounds
 - scoliosis.
- h) Management of comorbidities, including:
 - other motor disorders
 - cognitive and learning disabilities
 - visual and hearing impairments
 - epilepsy
 - feeding difficulties (including enteral tube feeding)
 - disorders of nutrition and growth
 - impaired bone mineralisation (osteoporosis)
 - urological disorders (voiding difficulties or incontinence)
 - gastrointestinal disorders (including gastro-oesophageal reflux and constipation)
 - respiratory disorders (including apnoea, airway obstruction and chronic aspiration).

4.4 Main outcomes

- a) Reduction of spasticity.

- b) Optimisation of movement and function.
- c) Reduction of pain.
- d) Adverse effects of interventions.
- e) Acceptability and tolerability in children and young people.
- f) Quality of life.

4.5 *Economic aspects*

Developers will take into account both clinical and cost effectiveness when making recommendations involving a choice between alternative interventions. A review of the economic evidence will be conducted and analyses will be carried out as appropriate. The preferred unit of effectiveness is the quality-adjusted life year (QALY), and the costs considered will usually only be from an NHS and personal social services (PSS) perspective. Further detail on the methods can be found in 'The guidelines manual' (see 'Further information').

4.6 *Status*

4.6.1 *Scope*

This is the draft scope for consultation. The consultation dates are 12 March to 9 April 2010.

4.6.2 *Timing*

The development of the guideline recommendations will begin in July 2010.

5 *Related NICE guidance*

5.1 *Published guidance*

- Selective dorsal rhizotomy for spasticity in cerebral palsy. [NICE interventional procedure guidance 195 \(2006\)](#).

6 Further information

Information on the guideline development process is provided in:

- ‘How NICE clinical guidelines are developed: an overview for stakeholders’ the public and the NHS’
- ‘The guidelines manual’.

These are available from the NICE website

(<http://www.nice.org.uk/guidelinesmanual>). Information on the progress of the guideline will also be available from the NICE website (www.nice.org.uk)

Appendix A Glossary

Ataxia: Impaired control of limb movements, often with tremor.

Baclofen: A drug that acts as a muscle relaxant.

Bolus dose: A drug delivered as a single, large dose.

Diplegia: Impaired movements in the legs.

Dyskinetic cerebral palsy (with athetosis, dystonia, chorea): People with dyskinetic cerebral palsy make involuntary movements, because their muscle tone changes rapidly from floppy and loose to tense and still, in a way they cannot control. It affects the movement of the body and presents as slow, rhythmic twisting movements of the trunk, or an arm or leg. It can also include abnormal postures.

Dystonia: Involuntary and uncontrollable muscle spasms that can force affected parts of the body into abnormal, sometimes painful, movements or postures.

Hemiplegia: Impaired movements in the arm and leg down one side of the body, and sometimes one side of the face.

Intrathecal injection: An injection into the fluid surrounding the spinal cord.

Monoplegia: Impaired movements in a single limb.

Multilevel surgery: All necessary surgeries are performed at the same time.

Orthosis: An orthopaedic device used to support, align or improve the function of a moveable part of the body.

Osteotomy: Surgery in which the bone is cut to shorten, lengthen or change its alignment.

Plegia: Complete loss of muscle function.

Quadraplegia: Impaired movements in the arms and legs, and often in the muscles of the face, head and neck.

Torsional deformity: The long bones of the leg are turned inwards or outwards so that the feet do not point straight ahead.