

NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE

SCOPE

1 Guideline title

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

1.1 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 a sign found in some motor disorders which is 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 components of the upper motor neuron syndrome.
- b) Spasticity is a common and often serious abnormality affecting motor function. Spasticity results in an increased resistance to passive movement of a muscle through hyperactive stretch reflexes causing rapid and strong contraction of the muscle. This dysregulation of tone with movement can result in a wide range of clinical manifestations and functional impairments.

- c) Spasticity in children and young people is most often seen in cerebral palsy, although it can also occur with other forms of non-progressive and progressive brain disorders, the latter is outside the remit of this guideline.
- d) In children and young people with cerebral palsy, the motor disorder can be characterised using the following approaches:
- Anatomic distribution of motor disorder
 - Unilateral involvement or bilateral involvement
 - Description of involvement of each limb, trunk and oropharynx
 - Nature of motor disorder
 - Spastic, dyskinetic or ataxic as predominant abnormality
 - Dyskinetic further divided into dystonic or choreoathetosis
 - Additional tone or movement problems listed as secondary types
 - Functional motor ability
 - Gross Motor Function Classification System (GMFCS) used to assess ambulation
 - Manual Ability Classification System (MACS) used to assess hand and arm function
 - Accompanying Impairments

This system of classification was developed by the Surveillance of Cerebral Palsy in Europe (SCPE) project and replaces the previous classification where the following terms were used to describe anatomic distribution:

- Hemiplegia – one side of body affected, arm usually more severely than leg
- Diplegia – legs predominantly affected, mild to moderate upper limb impairment
- Quadriplegia – severe impairment of arms and legs, often with trunk weakness and oropharyngeal involvement

As the guideline will be referring to literature over the last few decades, these terms will still be used in the assessment of the evidence for management of spasticity.

- e) Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation that are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems.
- f) 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.
- g) 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 and progressive disability can occur.
- h) Up to 80% of children with cerebral palsy have a spastic motor impairment. Other types of motor impairment in 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.
- i) Examples of non-progressive disorders that may affect the brain of a fetus or infant include brain malformations, prenatal vascular events (stroke) and infections (such as cytomegalovirus), perinatal hypoxic or ischaemic encephalopathy, and postnatal head injury or meningitis. When this damage occurs in the developing brains of

children under three years of age, it is referred to in this guideline as cerebral palsy.

- j) Non-progressive disturbances may also occur in older children and young people, for example, from head trauma, encephalitis or meningitis. Non-progressive disturbances affecting movement and posture occurring after this age are defined in this guideline as being “acquired”.
- k) 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).
- l) The functional abilities of children with spasticity often deteriorate over time. The cause of the progression is not often identified. It may include weakness, posturing, contracture, dystonia, ataxia or other motor disorders. Incorrect diagnosis and high expectations can all lead to functional deterioration. Effective management of spasticity and other motor problems could be important in preventing functional decline.
- m) 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, reduce mobility and cause difficulties in caring for the child.
- n) Subluxation or full dislocation of joints arise most commonly in the hips, but shoulder, elbow and ankle dislocations also occur though infrequently. Significant bony deformities can form such as kyphosis and scoliosis of the spine.

- o) These changes may substantially worsen the child's functional disability and impair the ability to walk or sit. Postural management or other specialist equipment may be necessary. 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. These functional deteriorations can cause a consequent reduction in the individual's independence, for example in dressing or toileting or in access to education or play. A lack of independence leads to an increased need for support by paid carers or family members. It may also reduce employment opportunities.
- p) Progressive disability requires acknowledgment, surveillance, prevention and management, especially during the transition to young adulthood when the demands of normal teenage life become more dominant in determining the health of the individual.
- q) 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 the effect that it has on the child – to treat pain, improve motor function, improve ease of care, and prevent the consequences of spasticity. In combination with other interventions dealing with the child's associated motor disorders and co-morbidities, the aim is 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 and young people with spasticity. These include passive stretching, muscle strengthening therapeutic exercises, serial casting, using splints and discouraging and preventing postures and movement that lead to disability and deformity, and encouraging postures and movement that improve function.
- d) Orthoses, aids and appliances are used to manage seating and posture or– 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. 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.
 - 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 373 (2010). This procedure has potential adverse effects such as hip instability and spinal deformity.
- f) Orthopaedic surgery has a major role in the management of early and late consequences 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 occurs 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.

- g) 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 disorder. 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 anatomic distribution of the motor disorder and the nature of the motor disorder.

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 and young people.
- c) Children with a pure dystonia or other motor disorders which do not co-exist with spasticity.

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 will be considered in relation to the subgroups of people with unilateral spasticity and bilateral spasticity. If clinically appropriate, each issue will also be considered in relation to the severity of the functional impairment using GMFCS and MACS. However, as this classification system has only recently come into general use, we will also use the older classification system (of spastic monoplegia, diplegia, hemiplegia and quadriplegia with severity graded as mild, moderate, or severe) as necessary to describe the reported evidence.

- a) Physiotherapy and occupational therapy interventions that have a direct effect to reduce spasticity, its musculoskeletal

consequences, or accompanying motor disorders for example, muscle shortening.

- b) Orthoses (for example, ankle-foot orthoses, knee splints, serial casting and upper limb orthoses) for preventing and treating contractures and improving function (such as mobility).
- c) Oral medications specifically baclofen, benzodiazepines (diazepam, nitrazepam, clonazepam), levodopa, tizanidine and dantrolene
- d) Long-term use of intramuscular botulinum toxin A and B 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 and young people with moderate to severe spasticity.
- f) The intrathecal baclofen pump to reduce spasticity, maintain motor function, to improve posture and improve health related quality of life in children and young people with moderate to severe spasticity.
- 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 health related quality of life in children and young people.
- i) Selective dorsal rhizotomy.

4.3.2 Clinical issues that will not be covered

- a) Diagnosis and assessment of spasticity and co-existing motor disorders.

- b) Management of spasticity and co-existing motor disorders caused by a progressive brain disorder or a spinal cord injury.
- c) Management of motor disorders which do not co-exist with spasticity.
- d) Holistic management of cerebral palsy or other non-progressive brain disorders.
- e) Play therapy.
- f) Complementary and alternative therapies.
- g) Management of the following complications:
 - kyphosis
 - scoliosis.
- h) Management of comorbidities, including:
 - cognitive and learning disabilities
 - visual, hearing and speech impairments
 - epilepsy
 - feeding difficulties (including enteral tube feeding)
 - disorders of nutrition and growth
 - impaired bone mineralisation (osteoporosis)
 - pressure sores
 - 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) Health related 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 final scope.

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 373 (2010). Available from

www.nice.org.uk/guidance/IPG373.

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

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