

NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

SCOPE

1 Guideline title

Cerebral palsy: the diagnosis and management of cerebral palsy in children and young people

1.1 *Short title*

Cerebral palsy

2 The remit

The Department of Health has asked NICE: 'To prepare a clinical guideline on the diagnosis and management of cerebral palsy'. This guideline will take account of the existing NICE guideline on [spasticity in children and young people with non-progressive brain disorders](#).

3 Need for the guideline

3.1 *Epidemiology*

- a) Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation, resulting from non-progressive disturbances (structural abnormalities) that occurred in the developing fetal or infant brain. There is general consensus of an upper age limit of 2 years for onset of the non-progressive brain disturbance and 5 years for clinical or developmental diagnosis. Patterns of motor disorder are generally subdivided into spastic, dyskinetic (including dystonic) and ataxic forms, depending on the area of the brain that is mainly involved.

- b) Although defined primarily as a motor disorder, cerebral palsy is often accompanied by disturbances of sensation, perception, cognition, communication and behaviour, and by epilepsy and musculoskeletal problems. Recognising the interrelationship of these associated disorders and managing them is an essential part of the overall management of cerebral palsy.
- c) Cerebral palsy registers using agreed definitions of the syndrome have shown a prevalence of 2.0–3.5 per 1000 live births in developed countries. Prevalence is inversely associated with gestational age and with birth weight. Prevalence has been reported as 90 cases per 1000 live births in babies with a birth weight of 1000 g, compared with 1.5 cases per 1000 live births for babies weighing 2500 g or more.
- d) Cerebral palsy is attributable mostly to events that occur before birth or in the neonatal period, with about 10–20% of cases resulting from intrapartum asphyxia. Only about 10% of cases arise from later events such as head injury or central nervous system infection (meningitis or encephalitis).
- e) In addition to prematurity and low birth weight, a wide range of risk factors for cerebral palsy exist, including multiple pregnancy and especially stillbirth or infant death of a co-twin, placental abnormalities, birth defects, meconium aspiration, emergency caesarean section, birth asphyxia, neonatal seizures, respiratory distress syndrome, hypoglycaemia and maternal, fetal or neonatal infection.
- f) It is important that disorders resulting from a progressive brain injury are distinguished from cerebral palsy. Although in cerebral palsy the causative brain injury is static, the secondary musculoskeletal problems and motor manifestations change over time. Typically, abnormalities of movement and posture are first recognised during infancy or early childhood, and secondary

disability can then be progressive. Attention should be paid to the evolution of the condition. If this differs from the pattern expected with cerebral palsy then other disorders should be considered, such as genetic and metabolic disorders and disorders resulting from progressive brain injury. In children and young people with dystonia the possibility of a dopamine-responsive disorder should be considered.

- g) Severe cerebral palsy can be associated with a reduced life expectancy. The effect may be minimal, but if gross and fine motor functioning, independent feeding, mental and visual capacities are severely impaired, survival to 40 years of age may be as low as 40%. Causes of early death may include pulmonary aspiration and pneumonia, accidents, associated disorders (for example, congenital heart disease) and delayed recognition of illness. Prognosis is an important issue that should be discussed with people with cerebral palsy and their family members and carers as appropriate. It can also potentially influence the approach to treatment.

3.2 *Current practice*

- a) Management of cerebral palsy depends on a multidisciplinary team of many specialists, across primary, district and regional services. The multidisciplinary team works with the child or young person with cerebral palsy, and their family members and carers as appropriate, to optimise development and minimise the impact of the brain impairment and comorbidities. The focus of social and clinical care during childhood and into young adulthood, which also involves colleagues from social care and education, is on facilitating function and inclusion, minimising 'activity limitation' and enabling individual 'participation'. These concepts are in line with the World Health Organization (WHO) framework, the International Classification of Functioning, Disability and Health, in which participation refers to involvement in life situations across a number

of functional domains, including self-care, relationships, education and, later, employment. This focus on functional ability and quality of life is key to managing cerebral palsy, with the perspective of the child or young person and their family members and carers at the centre of all decisions.

- b) Many specialists and experts may contribute to the recognition, diagnosis and management of cerebral palsy. The movement disorder itself is generally picked up either because of antenatal or neonatal concern about a potential brain impairment (from causes such as infection, epilepsy, prematurity or early hypoxic ischaemic damage) or by concerns raised during routine developmental screening (late sitting, standing and walking or early motor asymmetry).
- c) The primary care service for most families is the local child development team that supports health visitors and GPs. This team includes community paediatricians, physiotherapists, occupational therapists, speech and language therapists, nurses and preschool developmental teams. Other professionals, including specialised therapists, psychologists, orthotists, dietitians, hospital-based paediatricians, a variety of neurology and neurodisability experts, and orthopaedic and general surgeons, are often involved in care.
- d) A variety of care pathways for cerebral palsy exist, depending on the nature and degree of impairment. The spectrum of severity varies with regard to gross and fine motor functioning, bimanual manipulation, feeding, communication and associated disorders. Appropriate assessments and interventions differ depending on the age and level of functional ability of the child or young person.
- e) In addition to difficulties that the child or young person has with movement, posture and mobility, attention may need to be given to aspects such as communication, comfort and overall quality of life. Treatment may be needed for comorbidities such as epilepsy,

gastro-oesophageal reflux, constipation or aspiration pneumonia. In particular, oro-motor problems that affect swallowing and feeding, and hence nutrition, may be of central importance. Difficulties with saliva control that result in drooling can have a serious adverse effect on the wellbeing of the child or young person and their family members and carers. Vision, hearing, cognitive, behavioural and psychological difficulties occur more frequently than in the general population.

- f) Cerebral palsy is a lifelong condition, and this is an important perspective when considering clinical management. Service provision during the transition of healthcare from paediatric services to adult services is of critical importance. Preparing the young person and their family members and carers for this major change is crucial.

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 25th birthday who have cerebral palsy.
- b) Subgroups to be considered:

- recognised subgroups within the cerebral palsy population, depending on level of cognitive disability and functional disability (for example, Gross Motor Function Classification System levels I to V), and age ranges will be considered where appropriate.

4.1.2 Groups that will not be covered

- a) Adults 25 years and older.
- b) Children and young people with a progressive neurological or neuromuscular disorder.

4.2 Setting

- a) All settings in which NHS-commissioned health and social care is provided.

4.3 Management

4.3.1 Key issues that will be covered

Diagnosis and assessment

- a) Determining the key clinical and developmental manifestations of cerebral palsy at first presentation in order to help with early recognition.
- b) Identifying risk factors for cerebral palsy that may:
 - inform the need for enhanced surveillance
 - help in diagnosing the underlying cause of cerebral palsy
 - facilitate early intervention.
- c) Identifying the key information to be obtained from history and examination, including developmental screening to help in determining the underlying cause of cerebral palsy.
- d) Identifying 'red flags' that might suggest a neurodevelopmental disorder other than cerebral palsy, such as progressive neurological or neuromuscular disorders.

- e) Determining the potential value of MRI of the brain in cerebral palsy.
- f) The prognosis for children and young people with cerebral palsy in relation to:
 - ability to walk
 - ability to talk
 - life expectancy.
- g) Identifying common and important comorbidities associated with cerebral palsy and the subgroups most at risk of these comorbidities.
- h) Determining an effective approach to investigating difficulties with eating, drinking and swallowing in children and young people with cerebral palsy, including:
 - clinical observation
 - videofluoroscopic swallow studies and fibroscopic endoscopy.

Interventions

- i) Managing mental health problems in children and young people with cerebral palsy.
- j) Determining the effectiveness of interventions in tackling communication difficulties in children and young people with cerebral palsy.
- k) Determining the effective management of difficulties with eating, drinking and swallowing in children and young people with cerebral palsy.
- l) Determining the effective management of difficulties with saliva control (drooling) in children and young people with cerebral palsy.

- m) Nutritional management in children and young people with cerebral palsy.
- n) Assessing and managing pain, discomfort, distress and sleep disturbance in children and young people with cerebral palsy.
- o) Interventions to reduce the risk of reduced bone mineral density and low-impact fractures in children and young people with cerebral palsy.
- p) Managing difficulties associated with the processing of sensory and perceptual information in children and young people with cerebral palsy.
- q) Identifying social care needs that are specific to children and young people with cerebral palsy and their family members and carers.
- r) Communication, information and support needs that are specific to children and young people with cerebral palsy and their family members and carers.
- s) The role of the multidisciplinary team in the care of children and young people with cerebral palsy.
- t) Aspects of the transition from paediatric to adult health services that are specific to the needs of young people with cerebral palsy and their family members and carers.

Note that guideline recommendations will normally fall within licensed indications; exceptionally, and only if clearly supported by evidence, use outside a licensed indication ('off-label use') may be recommended. The guideline will assume that prescribers will use a drug's summary of product characteristics to inform decisions made with individual patients.

4.3.2 Issues that will not be covered

- a) Management of spasticity and co-existing motor disorders.
- b) Skin care, including management of pressure ulcers.

- c) Laboratory investigations for progressive neurological and neuromuscular disorders.
- d) Management of cognitive impairment and learning difficulties.
- e) Management of bladder dysfunction (urinary retention and incontinence) and bowel dysfunction (constipation and soiling).
- f) Management of gastro-oesophageal reflux disease.
- g) Management of respiratory complications such as pulmonary aspiration.
- h) Management of visual and hearing impairment.
- i) Management of epilepsy.

4.4 *Main outcomes*

- a) Health-related quality of life.
- b) Functional independence, including self-care and independence in activities of daily living.
- c) Ability to communicate.
- d) Participation (including social, education and work).
- e) Psychological wellbeing (for example, depression or anxiety).
- f) Degree of pain.
- g) Nutritional status.
- h) Wellbeing of parents/carers

4.5 *Review questions*

Review questions guide a systematic review of the literature. They address only the key issues covered in the scope, and usually relate to interventions, diagnosis, prognosis, service delivery or patient experience. Please note that

these review questions are draft versions and will be finalised with the Guideline Development Group.

4.5.1 Diagnosis and assessment

- a) What are the key clinical and developmental manifestations of cerebral palsy at first presentation?
- b) What are the risk factors for developing cerebral palsy and what is their prevalence?
- c) What are the causes of cerebral palsy in resource-rich countries?
- d) What clinical manifestations should be recognised as ‘red flags’ that suggest a progressive neurological or neuromuscular disorder rather than cerebral palsy?
- e) In children and young people with cerebral palsy, what is the effectiveness of an MRI scan in determining the cause of cerebral palsy?
- f) In children and young people with cerebral palsy, what is the effectiveness of an MRI scan in determining prognosis?
- g) What comorbidities are associated with cerebral palsy in children and young people and what is their prevalence, including prevalence in relevant subgroups?
- h) In children and young people with cerebral palsy, what are the symptoms and signs of mental health problems?
- i) In children and young people with cerebral palsy, which investigations are useful in evaluating difficulties with eating, drinking and swallowing (including clinical assessment, videofluoroscopic swallow studies and endoscopic examination)?
- j) In children and young people with cerebral palsy who are otherwise unable to communicate, what are the signs that suggest pain, discomfort, distress and sleep disturbance?

- k) In children and young people with cerebral palsy, what are the common causes of pain, discomfort, distress and sleep disturbance?
- l) In children and young people with cerebral palsy, what are the risk factors for reduced bone mineral density and low-impact fractures?
- m) In children and young people with cerebral palsy, what are the clinical and developmental prognostic indicators in relation to:
- the ability to walk
 - the ability to talk
 - life expectancy?

4.5.2 Interventions

- n) In children and young people with cerebral palsy, what interventions are effective in managing of mental health problems?
- o) In children and young people with cerebral palsy, how effective is clinical therapy focusing on oro-motor function in improving speech (for example, speech and language therapy strategies)?
- p) In children and young people with cerebral palsy, what communication systems (alternative or augmentative) are effective in improving communication (for example, eye gaze computerised technologies)?
- q) In children and young people with cerebral palsy, what interventions are effective in managing difficulties with eating, drinking and swallowing?
- r) In children and young people with cerebral palsy, what interventions are effective in managing poor saliva control (drooling)?

- s) In children and young people with cerebral palsy, what interventions are effective in maintaining adequate nutritional status?
- t) In children and young people with cerebral palsy, what interventions are effective for managing problems associated with difficulties in processing of sensory and perceptual information?
- u) In children and young people with cerebral palsy, what interventions are effective in managing pain, discomfort, distress and sleep disturbance with no known cause?
- v) In children and young people with cerebral palsy, what interventions are effective in preventing reduced bone mineral density and low-impact fractures?
- w) What are the specific social care needs of children and young people with cerebral palsy and their family members and carers (for example, use of equipment such as hoists, access to buildings and transport, and respite care)?
- x) What specific information and support is needed by children and young people with cerebral palsy and their family members and carers?
- y) What are the specific elements of the process of transition from paediatric to adult services that are important for young people with cerebral palsy and their family members and carers?

4.6 *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

be only from an NHS and personal social services (PSS) perspective. Further detail on the methods can be found in [The guidelines manual](#).

4.7 Status

4.7.1 Scope

This is the final scope.

4.7.2 Timing

The development of the guideline recommendations will begin in October 2014.

5 Related NICE guidance

5.1 Published guidance

5.1.1 Other related NICE guidance

- [Pressure ulcers](#) (2014) NICE guideline CG179
- [Autism: the management and support of children and young people on the autism spectrum](#) (2013) NICE guideline CG170
- [Urinary incontinence in neurological disease](#) (2012) NICE guideline CG148
- [Spasticity in children and young people with non-progressive brain disorders](#) (2012) NICE guideline CG145
- [The epilepsies](#) (2012) NICE guideline CG137
- [Autism in children and young people: recognition, referral and diagnosis of children and young people on the autism spectrum](#) (2011) NICE guideline CG128
- [Common mental health disorders](#). (2011) NICE guideline CG123
- [Selective dorsal rhizotomy for spasticity in cerebral palsy](#) (2010) NICE interventional procedure guidance 373
- [Constipation in children and young people](#) (2009) NICE guideline CG99
- [Depression in children and young people](#) (2005) NICE guideline CG28

5.2 *Guidance under development*

NICE is currently developing the following related guidance (details available from the [NICE website](#)):

- Gastro-oesophageal reflux disease in children and young people. NICE guideline. Publication expected January 2015.
- Challenging behaviour and learning disabilities. NICE guideline. Publication expected May 2015.
- Transition from children's to adult services. NICE guideline. Publication expected February 2016.

6 Further information

Information on the guideline development process is provided in the following documents, available from the NICE website:

- [How NICE clinical guidelines are developed: an overview for stakeholders the public and the NHS: 5th edition](#)
- [The guidelines manual](#).

Information on the progress of the guideline will also be available from the [NICE website](#).