

NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

SCOPE

1 Guideline title

Cerebral palsy: the diagnosis and management of cerebral palsy

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 guidance (which may be updated) on [spasticity in children and young people with non-progressive brain disorders](#) (NICE clinical guideline 145).

3 Need for the guideline

3.1 *Epidemiology*

- a) Cerebral palsy is a syndrome of motor impairment that results from a lesion in the developing brain. It has been defined as 'a group of permanent disorders of the development of movement and posture, causing activity limitation, and attributable to non-progressive disturbances that occurred in the developing fetal or infant brain'. There is a lack of consensus about when the brain stops developing rapidly, but there is general agreement on an arbitrary upper limit of between 3 and 5 years of age. Patterns of movement disorder are generally subdivided into spastic, 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 and managing these associated disorders is central to managing 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 and 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 neonatal infections.
- 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 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) Cerebral palsy can be associated with a reduced life expectancy. The effect may be minimal, but if gross and fine motor, independent feeding, mental and visual capacities are severely impaired then 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) Clinical 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 core service for most families is the local child development centre that supports health visitors and GPs. This team includes community paediatricians, physiotherapists, occupational therapists, speech and language therapists, nurses and portage workers (who provide a home-visiting educational service for preschool children with additional support needs). Hospital services, including specialised therapists, psychologists, 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 motor functioning, bimanual manipulation and communication. 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 child and young person 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).

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

- 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) Identifying common and important comorbidities associated with cerebral palsy.
- g) Determining the effectiveness of interventions in improving communication difficulties in children and young people with cerebral palsy.
- 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.
- i) Determining the effective management of difficulties with eating, drinking and swallowing in children and young people with cerebral palsy.
- j) Determining the effective management of difficulties with saliva control (drooling) in children and young people with cerebral palsy. 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.
- k) Nutritional management in children and young people with cerebral palsy.
- l) Assessing signs of pain, discomfort or distress in children and young people with cerebral palsy.
- m) Managing difficulties with sensory planning and perception in children and young people with cerebral palsy.

- n) The prognosis for children and young people with cerebral palsy in relation to:
- the ability to walk
 - the ability to talk
 - life expectancy.
- o) Social care needs that are specific to children and young people with cerebral palsy and their family members and carers.
- p) Communication, information and support needs that are specific to children and young people with cerebral palsy and their family members and carers.
- q) The role of the multidisciplinary team in the care of children and young people with cerebral palsy.
- r) Aspects of the transition from paediatric to adult services that are specific to the needs of young people with cerebral palsy and their family members and carers.

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 sores.
- 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) Social participation.
- e) Psychological wellbeing (for example, depression or anxiety).
- f) Degree of pain.
- g) Nutritional status.

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.

- a) What are the 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) What is the value of an MRI scan in determining the cause of cerebral palsy?
- f) What are the comorbidities associated with cerebral palsy and what is their prevalence?
- g) How effective is clinical therapy (for example, speech and language therapy strategies) focusing on oro-motor function in improving speech in children and young people with cerebral palsy?
- h) What communication systems (alternative or augmentative) are effective in improving communication (for example, eye gaze computerised technologies) in children and young people with cerebral palsy?
- i) What investigations are useful in evaluating difficulties with eating, drinking and swallowing in children and young people with cerebral palsy, including clinical assessment, videofluoroscopic swallow studies and endoscopic examination?
- j) What interventions are effective in managing difficulties with eating, drinking and swallowing in children and young people with cerebral palsy?
- k) What interventions are effective in managing poor saliva control (drooling) in children and young people with cerebral palsy?
- l) What interventions are effective in maintaining adequate nutritional status in children and young people with cerebral palsy?
- m) What clinical manifestations suggest the presence of pain, discomfort or distress in children and young people with cerebral palsy who are otherwise unable to communicate?
- n) What interventions are effective in managing problems associated with difficulties in sensory planning and perception in children and young people with cerebral palsy?

- o) What is the prognosis for children and young people with cerebral palsy in relation to:
- the ability to walk
 - the ability to talk
 - life expectancy?
- p) 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)?
- q) What specific information and support is needed by children and young people with cerebral palsy and their family members and carers?
- r) 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 consultation draft of the scope. The consultation dates are 7 July to 4 August 2014.

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](#). NICE clinical guideline 179 (2014).
- [Autism: the management and support of children and young people on the autism spectrum](#). NICE clinical guideline 170 (2013).
- [Urinary incontinence in neurological disease](#). NICE clinical guideline 148 (2012).
- [Spasticity in children and young people with non-progressive brain disorders](#). NICE clinical guideline 145 (2012).
- [The epilepsies](#). NICE clinical guideline 137 (2012).
- [Autism in children and young people: recognition, referral and diagnosis of children and young people on the autism spectrum](#). NICE clinical guideline 128 (2011).
- [Selective dorsal rhizotomy for spasticity in cerebral palsy](#). NICE interventional procedure guidance 373 (2010).
- [Nocturnal enuresis](#). NICE clinical guideline 111 (2010).
- [Constipation in children and young people](#). NICE clinical guideline 99 (2009).
- [Depression in children and young people](#). NICE clinical guideline 28 (2005).

5.2 *Guidance under development*

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

- Gastro-oesophageal reflux in children and young people. NICE clinical guideline. Publication expected January 2015.

- Transition from children's to adult services. NICE social care 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](#).