Specialist Pharmacy Service

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Cerebral palsy in under 25s

NICE NG62: 2017

This guideline covers the assessment and management of cerebral palsy in under 25s.

Definition of terms

MDT	multidisciplinary team
GMFCS	gross motor function classification system
GORD	gastro-oesophageal reflux disease
DEXA	dual energy X-ray absorptiometry
U	unlicensed indication

Causes of cerebral palsy – see NICE pathway

Risk factors

- Recognise independent risk factors for cerebral palsy – see **Box 1**
- Provide an enhanced clinical and developmental follow-up programme for children who have any of the risk factors.

Box 1: Risk factors

Risk factors for cerebral palsy

Antenatal	Perinatal	Postnatal
♦ preterm birth (increased risk with decreasing gestational age) ♦ chorioamnionitis ♦ maternal respiratory tract or genito-urinary infection treated in hospital	♦ low birth weight ♦ chorioamnionitis ♦ neonatal encephalopathy ♦ neonatal sepsis (particularly with a birth weight <1.5 kg) ♦ maternal respiratory tract or genito-urinary infection treated in hospital	♦ meningitis

Assessment and management

Looking for signs of cerebral palsy

- Recognise the following as possible early motor features in the presentation of cerebral palsy:
  - unusual fidgety movements or other abnormalities of movement, including asymmetry or paucity of movement,
  - abnormalities of tone, including hypotonia (floppiness), spasticity (stiffness) or dystonia (fluctuating tone),
  - abnormal motor development, including late head control, rolling, and crawling,
  - feeding difficulties.
- Recognise that the most common delayed motor milestones in children with cerebral palsy are:
  - not sitting by 8 months*,
  - not walking by 18 months*,
  - early asymmetry of hand function (hand preference) before 1 year*.
- If there are concerns that a child may have cerebral palsy but a definitive diagnosis cannot be made, discuss this with their parents/carers and explain that an enhanced clinical and developmental follow-up programme will be necessary to try to reach a definite conclusion.
- Refer all children with the following to a child development service for further assessment:
  - delayed motor milestones,
  - persistent toe walking.

* corrected for gestational age

Red flags

- Review a diagnosis of cerebral palsy if clinical signs or the child's development do not follow the pattern expected, taking into account that functional and neurological manifestations of cerebral palsy change over time.
- Recognise the following as red flags for neurological disorders other than cerebral palsy and refer the child/young person to a specialist in paediatric neurology if any of these are observed:
  - absence of known risk factors,
  - family history of a progressive neurological disorder,
  - loss of already attained cognitive or developmental abilities,
  - development of unexpected focal neurological signs,
  - MRI findings suggestive of a progressive neurological disorder or not in keeping with clinical signs of cerebral palsy.

Information on prognosis – see NICE pathway

Multidisciplinary care

- Refer all children with suspected cerebral palsy to a child development service for urgent multidisciplinary assessment, to facilitate early diagnosis and intervention.
- Recognise that children/young people with cerebral palsy and their parents/carers have a central role in decision-making and care planning.
- Ensure that the child/young person has access to a local integrated core MDT able to meet their individual needs within agreed care pathways and who can provide expertise and access to other services, as appropriate, through a local network of care – see NICE pathway

Managing comorbidities

General principles

- Assess children/young people regularly for developmental and clinical comorbidities, and recognise that these can have an important impact on wellbeing, function and participation.
- Manage comorbidities, and refer the child/young person for further specialist care if necessary e.g. if a management programme is unsuccessful.
- Also see NICE pathway: Multimorbidity.
- For guidance on the safe and effective use of medicines, see NICE pathway: Medicines optimisation.

Epilepsy

- Advise children and their parents/carers that epilepsy may be associated with cerebral palsy. Useful information to discuss includes the following:
  - epilepsy occurs in around 1 in 3 children with cerebral palsy,
  - it may occur in children/young people with any functional level or motor subtype, but prevalence increases with increasing severity of motor impairment,
  - it is reported in around 1 in 2 children with dyskinetic cerebral palsy.
- Ensure that dyskinetic movements are not misinterpreted as epilepsy in children with cerebral palsy.
- For guidance on identifying and managing epilepsy - see NICE pathway: Epilepsy.

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Low bone mineral density

In children/young people with cerebral palsy:

- Recognise that the following are independent risk factors for low bone mineral density:
  - non-ambulant (GMFCS level IV or V),
  - vitamin D deficiency,
  - presence of eating, drinking and swallowing difficulties or concerns about nutritional status,
  - low weight for age (below the 2nd centile),
  - history of low-impact fracture,
  - use of anticonvulsant medication.
- Recognise that there is an increased risk of low-impact fractures in children/young people who are non-ambulant or have low bone mineral density.
- Inform children/young people and their parents/carers if they are at an increased risk of low-impact fractures.

Management

- If a child/young person with cerebral palsy has ≥1 risk factors for low bone mineral density assess dietary intake of calcium and vitamin D and consider the following investigations:
  - serum calcium, phosphate and alkaline phosphatase,
  - serum vitamin D,
  - urinary calcium/creatinine ratio.
- Create an individualised care plan for children/young people with cerebral palsy who have ≥1 risk factors for low bone mineral density.
- Consider the following as possible interventions to reduce the risk of reduced bone mineral density and low-impact fractures:
  - an active movement programme,
  - active weight bearing,
  - dietetic interventions as appropriate, including nutritional support and calcium and vitamin D supplementation,
  - minimising risks associated with movement and handling.
- Consider a DEXA scan under specialist guidance for children/young people with cerebral palsy who have had a low-impact fracture.
- Refer children/young people with cerebral palsy with reduced bone density and a history of low-impact fracture to a specialist centre for consideration of bisphosphonate therapy.
- **Do NOT** offer vibration therapy solely to prevent low bone mineral density.
- **Do NOT** offer standing frames solely to prevent low bone mineral density.

Nutritional status

- Regularly review the nutritional status of children/young people with cerebral palsy, including measuring height and weight (or consider alternative anthropometric measurements, if height and weight cannot be measured).
- Provide timely access to assessment and nutritional interventional support from a dietitian if there are concerns about oral intake, growth or nutritional status.
- If oral intake is still insufficient to provide adequate nutrition, refer the child/young person to be assessed for enteral tube feeding by a MDT with relevant expertise.
- For guidance on nutritional interventions and enteral tube feeding in over 18s – see NICE pathway: Nutrition support in adults.

Managing functional issues

Vomiting, regurgitation and reflux

- Advise parents/carers that vomiting, regurgitation and gastro-oesophageal reflux are common in children/young people with cerebral palsy. If there is a marked change in the pattern of vomiting, assess for a clinical cause.

For guidance on identifying and managing GORD - see NICE pathway: Dyspepsia and gastro-oesophageal reflux disease.

Saliva control

- Assess factors that may affect drooling such as positioning, medication history, reflux and dental issues before starting drug therapy
- Consider the use of anticholinergic drug treatment to reduce the severity and frequency of drooling:
  - glycopyrronium bromide oral solution**U** or by enteral tube (off-label), **OR**
  - transdermal hyoscine hydrobromide**U**, **OR**
  - trihexyphenidyl hydrochloride**U** for children with dyskinetic cerebral palsy, but only with input from specialist services.
- NICE has published an evidence summary on severe sialorrhoea (drooling) in children/young people with chronic neurological disorders.
- When choosing a medicine take into account preferences of the child/young person and their parents /carers, and the age range and indication covered by the marketing authorisations.
- Regularly review effectiveness, tolerability and side effects of all drug treatments used for saliva control.
- If anticholinergic drug treatments are contraindicated, not tolerated or not effective, refer the child/young person to a specialist service to consider other treatments.
- Consider use of botulinum toxin A**U** injections to salivary glands with ultrasound guidance to reduce severity and frequency of drooling if anticholinergic drugs provide insufficient benefit or are not tolerated.
- Advise children/young people and their parents/carers that high-dose botulinum toxin A**U** injection to saliva glands can rarely cause swallowing difficulties, and to return to hospital immediately if breathing or swallowing difficulties occur.
- Consider referring young people for a surgical opinion, after an assessment confirming clinically safe swallow, if there is:
  - potential need for lifelong drug treatment, **OR**
  - insufficient benefit or non-tolerance of anticholinergic drugs and botulinum toxin A injections.

Pain, discomfort and distress

- Explain to children/young people and their parents/carers that pain is common in people with cerebral palsy, especially those with more severe motor impairment, and this should be recognised and addressed.
- Recognise that the most common condition-specific causes of pain, discomfort and distress in children/young people with cerebral palsy include:
  - musculoskeletal problems e.g. scoliosis, hip subluxation and dislocation,
  - increased muscle tone (including dystonia and spasticity),
  - muscle fatigue and immobility,
  - constipation,
  - vomiting,
  - GORD.
- Recognise that usual causes of pain, discomfort and distress that affect children/young people also occur in those with cerebral palsy, and that difficulties with communication and perception may make identifying the cause more challenging.
- Common types of pain in children/young people include:
  - non-specific back pain or abdominal pain,
  - headache,
  - dental pain,
  - dysmenorrhea.

Recommendations – wording used such as 'offer' and 'consider' denote the strength of the recommendation.

Drug recommendations – the guideline assumes that prescribers will use a drug's Summary of Product Characteristics (SPC) to inform treatment decisions.

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Sleep disturbances

- Explain to parents/carers that, in children/young people with cerebral palsy, sleep disturbances are common e.g. difficulties with falling asleep and staying asleep or daytime sleepiness and may be caused by factors such as environment, hunger and thirst.
- Recognise that the most common condition-specific causes of sleep disturbances include:
  - sleep-induced breathing disorders, such as obstructive sleep apnoea,
  - seizures,
  - pain and discomfort ,
  - need for repositioning because of immobility,
  - poor sleep hygiene (poor night-time routine and environment),
  - night-time interventions, including overnight tube feeding or the use of orthoses,
  - comorbidities, including adverse effects of medication.
- When identifying and assessing sleep disturbances in children/young people with cerebral palsy:
  - recognise that parents and familiar carers have the primary role in this,
  - consider using sleep questionnaires or diaries.
- Always ask about pain, sleep and distress as part of any clinical consultation.

Management

- Optimise sleep hygiene.
- Manage treatable causes of sleep disturbances that are identified.
- If no treatable cause is found, consider a trial of melatoninU to manage sleep disturbances particularly for problems with falling asleep.
- **Do NOT** offer regular sedative medication to manage primary sleep disorders without seeking specialist advice.
- **Do NOT** offer sleep positioning systems solely to manage primary sleep disorders.
- Refer the child/young person to specialist sleep services for MDT assessment and management if there are ongoing sleep disturbances.

Constipation

- Be aware that around 3 in 5 children/young people with cerebral palsy have chronic constipation and:
  - discuss this with children/young people and their parents/carers,
  - carry out regular clinical assessments for constipation.
- For guidance on identifying and managing constipation in under 18s - see NICE pathway: Constipation.

Eating, drinking and swallowing difficulties

- If eating, drinking and swallowing difficulties are suspected in a child/young person with cerebral palsy, carry out a clinical assessment as first-line investigation to determine the safety, efficiency and enjoyment of eating and drinking. This should include:
  - taking a relevant clinical history, including asking about any previous chest infections,
  - observation of eating and drinking in a normal mealtime environment by a speech and language therapist with training in assessing and treating dysphagia.
- Refer the child/young person to a local specialist MDT with training in assessing and treating dysphagia if there are clinical concerns such as:
  - coughing, choking, gagging altered breathing pattern or change in colour while eating or drinking,
  - recurrent chest infection,
  - mealtimes regularly being stressful or distressing for the child/young person or their parents/carers,
  - prolonged meal duration.
- **Do NOT** use videofluoroscopy or fibroscopic endoscopy for the initial assessment of eating, drinking and swallowing difficulties in children/young people with cerebral palsy.
- For recommendations on when to consider videofluoroscopy – see NICE pathway.

Management

- Develop strategies and goals in partnership with the child/young person and their parents/carers/other family members for interventions to improve eating, drinking and swallowing.
- Create an individualised plan for managing eating, drinking and swallowing difficulties, taking into account understanding, knowledge and skills of parents/carers and any other people involved in feeding the child/young person. Assess the role of the following:
  - postural management and positioning when eating,
  - modifying fluid and food textures and flavours,
  - feeding techniques, such as pacing and spoon placement,
  - equipment, such as specialised feeding utensils,
  - optimising the mealtime environment,
  - strategies for managing behavioural difficulties,
  - strategies for developing oral motor skills,
  - communication strategies,
  - modifications to accommodate visual or other sensory impairment that affect eating, drinking and swallowing,
  - the training needs of people who care for the child/young person particularly outside the home.
- Advise parents/carers that intra-oral devices have not been shown to improve eating, drinking and swallowing in children/young people with cerebral palsy.
- Use outcome measures important to the child/young person and their parents/carers to review:
  - whether individualised goals have been achieved,
  - the clinical and functional impact of interventions to improve eating, drinking and swallowing.

Mental health

- Follow relevant NICE recommendations when identifying and managing mental health problems and psychological and neurodevelopmental disorders in children/young people with cerebral palsy.

Assessment

- Take into account that parents and familiar carers have a central role in recognising and assessing emotional difficulties and mental health problems in children/young people with cerebral palsy.
- Recognise that children/young people with cerebral palsy have an increased prevalence of:
  - mental health and psychological problems, including depression, anxiety and conduct disorders,
  - behaviours that challenge, which may be triggered by pain, discomfort or sleep disturbances,
  - neurodevelopmental disorders, including autism spectrum disorder and attention deficit hyperactivity disorder.
- Recognise that emotional and behavioural difficulties e.g. low self-esteem, are reported in up to 1 in 4 children/young people with cerebral palsy.
- Any MDT should:
  - recognise that mental health problems and emotional difficulties can be as important as physical health problems for children/young people with cerebral palsy,
  - explore such difficulties during consultations,
  - recognise that assessing psychological problems can be challenging in children/young people with communication difficulties or learning disability (intellectual disability).

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- Think about and address the following contributory factors if a change in emotional state occurs in a child/young person with cerebral palsy:
  - pain or discomfort,
  - frustration associated with communication difficulties,
  - social factors, such as a change in home circumstances or care provision.
- Use validated tools, such as the Child Health Questionnaire and the Strengths and Difficulties Questionnaire, to assess mental health problems in children/young people with cerebral palsy.

Management

- Refer the child/young person with cerebral palsy for specialist psychological assessment and ongoing management if emotional and behavioural difficulties persist or there are concerns about their mental health.
- Work in partnership with the child/young person and their parents and primary carers, when assessing and managing mental health problems and setting goals.
- When making an individual management plan to address the mental health needs of a child/young person with cerebral palsy, take into account ways of providing support to parents/carers.
- Recognise that there are specific challenges in managing and minimising the impact of mental health problems in children/young people with cerebral palsy. These include:
  - communication difficulties,
  - comorbidities, particularly epilepsy and pain,
  - side effects and drug interactions of multiple medications (polypharmacy),
  - adverse effects of medicines used for managing mental health problems on motor function,
  - adverse effects of medicines used for managing motor function on mental health,
  - specific social care needs.
- For information on reviewing medicines and shared-decision making about medicines – see NICE pathway: Medicines optimisation.

Behavioural difficulties

- Talk to children/young people and their parents/carers about behavioural difficulties that can be associated with cerebral palsy.
- Useful information to discuss includes that around 2 to 3 in 10 children/young people with cerebral palsy have ≥1of the following:
  - emotional and behavioural difficulties that have an effect on the child/young person's function and participation,
  - problems with peer relationships,
  - difficulties with attention, concentration and hyperactivity,
  - conduct behavioural difficulties.
- Recognise that difficulties with registering or processing sensory information may present as behavioural difficulties.
- Support children/young people with cerebral palsy and their families/carers to recognise behavioural difficulties.
- Manage routine behavioural difficulties within the MDT, and refer the child /young person to specialist services if difficulties persist.
- See also:

NICE pathway: Attention deficit hyperactivity disorder
NICE pathway: Challenging behaviour and learning disabilities
NICE pathway: Mental health problems in people with learning disabilities

Visual and hearing impairment – see NICE pathway

Learning disability (intellectual disability) – see NICE pathway

Information and support

- Ensure that information and support focuses as much on the functional abilities of the child/young person with cerebral palsy as on any functional impairment.
- Provide clear, timely and up-to-date information to parents or carers on the following topics:
  - diagnosis,
  - aetiology,
  - prognosis,
  - expected developmental progress,
  - comorbidities,
  - availability of specialist equipment,
  - resources available and access to financial, respite, social care and other support for children/young people and their parents, carers and siblings,
  - educational placement (including specialist preschool and early years settings),
  - transition from children to adult services.
- Ensure that clear information about the 'patient pathway' is shared with the child/young person and their parent/carers e.g.by providing them with copies of correspondence. See NICE guidance: Patient experience in adult NHS services.
- Provide information to the child/young person, and their parents/carers, on an ongoing basis. Adapt communication methods and information resources to take account of the needs and understanding of the child/young person and their parents/carers. Think about using 1 or more of the following:
  - oral explanations,
  - written information and leaflets,
  - mobile technology, including apps,
  - augmentative and alternative communication systems.
- Work with the child/young person and their parents/carers to develop and maintain a personal 'folder' in their preferred format (electronic or otherwise) containing relevant information that can be shared with their extended family and friends and used in health, social care, educational and transition settings.
- Ensure that the child/young person and their parents/carers are provided with information, by a professional with appropriate expertise, about the following topics relevant to them tailored to their individual needs:
  - menstruation,
  - fertility,
  - contraception,
  - sex and sexuality,
  - parenting.
- Provide information to the child/young person and their parents/carers, and to all relevant teams around them, about:
  - local and regional services available e.g. sporting clubs, respite care and specialist schools for children/young people with cerebral palsy, and how to access them,
  - local support and advocacy groups to the child/young person and their parents/gcarers.

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Resources:
NICE Evidence summary (ES5): Severe sialorrhoea (drooling) in children and young people with chronic neurological disorders; oral glycopyrronium bromide.