Assessment and management of cerebral palsy in the under-25s

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In January, NICE published its first guideline on cerebral palsy (NG62), with the aim of improving the diagnosis and treatment of children and young people across England. This article provides a summary of the guidance.

Cerebral Palsy in Under 25s: Assessment and Management (NG62) is the first NICE guideline to tackle this condition.[1] Work on this 47-page guideline began in 2014 and a glance at its scope shows the next three years was time well spent. The guidance covers recognition and assessment, the management of physical and mental health co-morbidities, dealing with personal and family needs, and the transition to adult services. Its intention is to ensure individuals "get the care and treatment they need… so that they can be as active and independent as possible."

The guideline, which contains 19 sections and a wealth of detail, is intended for health professionals, social care workers, families and carers, and the children and young people affected. There is a version written for the public on the NICE website[2] – a better place to start for the nonspecialist (service user or provider) – but even this is a lengthy document. Those looking for a shorter introduction should first read the summary on page 43 of the main guidance document and the introduction to the draft scope guideline (in the history section on the NICE website).[3]

NICE covers the management of problems with movement and posture in children and young people with cerebral palsy in *Spasticity in Under 19s: Management* (CG145, 2012).[4]

- Early history
- Motor subtype and limb involvement
- Functional abilities
- Interventions
- Medication
- Co-morbidities
- Preferred methods of communication
- Any specialist equipment that is used or needed
- Care plans
- Emergency contact details

Table 1. Recommended contents of a personal folder

Diagnosis and assessment

Signs of cerebral palsy may not be evident at birth so a clinical and developmental programme up to the age of two years should be established for children with risk factors. These may be antenatal, *eg* preterm birth, perinatal (low birth weight) or postnatal (meningitis). Early signs of cerebral palsy to look out for include unusual fidgeting, abnormalities of tone, abnormal motor development and feeding difficulties.

A diagnosis of cerebral palsy is not always clear. If the child's development does not follow the pattern expected for cerebral palsy, red flags indicating other neurological disorders should be considered. These include the absence of known risk factors, a family history or MRI findings suggestive of progressive neurological disorder, the loss of cognitive or developmental abilities, the development of unexpected focal neurological signs, and MRI findings not consistent with the clinical signs of cerebral palsy.

Providing care and support

All children in whom cerebral palsy is suspected should be referred to a multidisciplinary child development service for an urgent assessment. When this happens,

there is a risk that control is wrested from the family but NICE recommends that services should "recognise that children and young people with cerebral palsy and their parents or carers have a central role in decision-making and care planning." The multidisciplinary team should include a range of professionals with expertise that can meet individual needs. As well as general and specialist services in a local care network, this means teaching support for preschool and school-age children, including home teaching for preschool children.

All this adds up to a complex care pathway, so effective communication and clear routes of access should be established. If people with cerebral palsy and their families are to have any influence on this process, they need to be well informed about cerebral palsy and its management – in other words, they need to be familiar with what is in this guideline, with the services they will need and how care will change over time. Individuals and families should maintain a personal "folder" – electronic or otherwise – that they can share with their family and friends and use when dealing with care services (see Table 1). Parents and carers need to know about a child's prognosis, particularly as it affects walking, speech and life expectancy. In addition, appropriately trained health professionals should provide information about issues that become important with maturity such as menstruation, sex and sexuality, fertility and contraception, and parenting. All should also be told about local and regional support, including advocacy groups, leisure and sports facilities.

Management of physical and mental health problems

Clinical concerns about eating, drinking and swallowing should prompt a referral to the multidisciplinary team. There are several indications for videofluoroscopy (*eg* safety concerns) but this should be neither the first nor the routine strategy. All this should result in a management plan providing access to tertiary services for ongoing problems, developed jointly with the individual and family, and with agreed outcomes to measure performance.

About half of children and young people with cerebral palsy have communication difficulties, 10% need to use signs, symbols and speech-generating devices and a further 10% are unable to do so. Regular assessment of speech, language and communication, referral and specialist management are recommended. Interventions should be introduced early and include physical training (*eg* posture, breath control), use of support devices and communication training for the family.

Concerns about oral intake, growth or nutritional status should prompt assessment by a dietitian. If nutritional interventional support is unsuccessful, enteral feeding may be necessary. For over-18s, this is covered by the NICE guidance *Nutrition Support for Adult*s (CG32, 2006).[5]

Before considering drugs to reduce drooling, possible contributory factors such as positioning, medication, dental issues and reflux should be excluded. Anticholinergic drugs can reduce saliva flow; options include glycopyrronium bromide oral solution, transdermal hyoscine hydrobromide and (for children with dyskinetic cerebral palsy but only with input from specialist services) oral trihexyphenidyl hydrochloride. Sialanar (glycopyrronium bromide oral solution) received a licence for the symptomatic treatment of severe sialorrhoea in children aged three years and older with chronic neurological disorders in September 2016 and NICE recently published an evidence summary for it, concluding that it is better than placebo, adverse effects are common, almost a third of children discontinue use, and it costs over £400 per month (ES5, 2017).[6] Treatment with anticholinergic drugs (which are not licensed for all age groups) should be monitored and reviewed; if they are contraindicated, ineffective or not tolerated, injection of botulinum toxin type A into the salivary glands is an option. Surgery should be considered when it seems that lifelong drug treatment might be needed, or if drugs are not working well enough or are poorly tolerated.

- Nonambulant
- 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

Table 2. Risk factors for low bone mineral density in children and young people with cerebral palsy

- Musculoskeletal problems (*eg* scoliosis, hip subluxation and dislocation)
- Increased muscle tone (including dystonia and spasticity)
- Muscle fatigue and immobility
- Constipation
- Vomiting
- Gastro-oesophageal reflux disease

Table 3. Aspects of cerebral palsy associated with pain

Children and young people with cerebral palsy are at risk of low bone mineral density. Anyone with one or more risk factors (see Table 2) should have a review of their vitamin D and calcium status to inform a care plan that includes a programme of active movement, weight-bearing activity, nutritional support and supplementation, while minimising risks from movement and handling. A fracture indicates the need to consider a DEXA scan and bisphosphonate therapy.

Pain is common in people with cerebral palsy – due either to the problems associated with the condition (see Table 3) or the everyday niggles that affect everyone (nonspecific back pain, headache, nonspecific abdominal pain, dental pain and dysmenorrhoea). Problems with communication make it harder to identify and assess pain. Pain management is much the same as for any person: after addressing any underlying cause, it should accommodate the impact of anxiety, depression or other possible mental health problems and use the traditional stepped approach, beginning with simple analgesics like paracetamol and ibuprofen. If this does not work, a referral to a specialist pain team is indicated.

Sleep disturbance is also common. It may be due to nonspecific or specific causes (see Table 4). If addressing these and sleep hygiene measures are insufficient, a trial of melatonin should be considered (especially if the problem is falling asleep). Sedating drugs should not be prescribed unless recommended

by a specialist and sleep positioning should not be the sole intervention. Refractory problems warrant specialist referral.

NICE also makes recommendations for managing other co-morbidities (see Table 5). These problems tend to be more frequent among individuals with greater motor impairment. Children and young people and their families should be informed about these risks. NICE recommends its guideline *Medicines Optimisation* (NG5, 2015)[7] for advice on ensuring the safe and effective use of medicines.

A quarter of children and young people with cerebral palsy have emotional and behavioural difficulties such as low self-esteem. The role of family and carers in identifying and assessing these issues should be recognised. NICE refers to seven guidelines covering anxiety, depression, learning difficulty, autism, ADHD and challenging behaviour and recommends referral to a specialist psychologist if difficulties persist or if there are mental health concerns. Management may be complicated by communication difficulties, adverse effects and medication interactions, social factors and co-morbidities. Support for families and carers should not be forgotten.

Utilising other services

Individuals and their families should be assessed at diagnosis and regularly thereafter for their needs for social care (particularly important to facilitate participation and independence), financial support, welfare rights information, support groups, and respite and hospice services. Mobility equipment, transport, and toileting and changing facilities should be assessed to ensure they help individuals join in activities in the home, school, workplace and community. Tailored care such as pain management, rehabilitation and equipment should be in place after any major surgical intervention.

The guideline *Transition from Children's to Adults' Services for Young People Using Health or Social Care Services* (NG43, 2016)[8] sets out how this process can work well during what may well be more than a single step as the individual moves between resident educational, college and adult home settings. The problems of cerebral palsy continue into adulthood and need to be planned for in the transition process, which should include provision of named key workers and staff with appropriate expertise.

References

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2. National Institute for Health and Care

Nonspecific
• Environment • Hunger • Thirst
Specific
• Sleep-induced breathing disorders (*eg* 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 (*eg* overnight tube feeding or the use of orthoses) • Co-morbidities (including adverse effects of medication)

Table 4. Causes of sleep disturbance in cerebral palsy

Visual impairment	• Half of children and young people are affected and one-fifth have cerebral visual impairment • Refer for ophthalmological and optometric assessment • Regularly assess
Hearing impairment	• Affects 10% of children and young people • Regular ongoing assessment needed
Learning disability	• Affects half of children and young people, and severe (IQ<50) in a quarter • Problems with knowledge acquisition, memory, and understanding and use of language • Discuss with individual and family
Behavioural difficulties	• 20–30% of children and young people affected • Problems processing information may manifest this way • Provide support and manage within multidisciplinary team; specialist referral an option
Vomiting, regurgitation and reflux	• Common • Covered by NICE guidelines NG1 (2015) and CG184 (2014)
Constipation	• Affects 60% of children and young people • Assess regularly and discuss • Covered by NICE guideline CG99 (2010)
Epilepsy	• Affects a third of children and young people, but a half of those with dyskinetic cerebral palsy • Do not confuse with dyskinesias • Covered by NICE guideline CG137 (2012)

Table 5. Recommendations for managing other co-morbidities

Excellence. *Cerebral palsy in under 25s: assessment and management. Information for the public*. January 2017. Available from: https://www.nice.org.uk/guidance/ng62/resources/cerebral-palsy-in-under-25s-assessment-and-management-5646702664645
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Declaration of interests

None to declare.

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