

Cerebral palsy in adults

Ethan is 33 years old, has cerebral palsy and is having increasing problems walking on his right leg. He is usually well and recently moved out of his family home to his own flat. However, he is on sick leave again from work, his muscles feel tight and painful, and he is experiencing a high level of fatigue. This is the second episode in the past year that has led to him needing time off work. He can't work remotely, and is worried as he is finding it difficult to explain the issues to his employers and is concerned about his job security.

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This information comes from the 2019 NICE guideline on cerebral palsy in adults and a BMJ summary of the same NICE guidance (NICE 2019, NG 119, BMJ 2019;364:l806, NICE 2020, QS191).

Headlines

- Cerebral palsy is a non-progressive (but not unchanging) disorder of movement and posture.
- It may affect communication, learning, feeding and vision.
- It is associated with epilepsy, which affects about 30% adults with cerebral palsy, more commonly in those with co-occurring intellectual impairment.
- Cerebral palsy is common, affecting 2–3/1000 live births. This means that 2–3 adults per 1000 have cerebral palsy.
- Impairment of the developing brain can occur in utero, at birth or (less commonly) in the first 2 years of life, and can be due to prematurity, hypoxia or infection.
- About 90% of children with cerebral palsy survive to adulthood; in the UK, about 75% of people with cerebral palsy are adults.

Cerebral palsy can be classified depending on the location of the symptoms, the motor subtype and the severity.- Adults with cerebral palsy with different symptoms will have different needs:

Area affected	Motor subtype	Severity of gross motor impairment (using the Gross Motor Function Classification System)
Unilateral	Spasticity	Level 1: advanced gross motor skills limited, walks without restrictions.
Bilateral	Dyskinesia	Level 2: limitations walking outdoors/in community, walks without assistive devices.
	Dystonia	Level 3: limitations walking outdoors/in community, walks with assistive devices.
		Level 4: requires transport/power mobility outdoors/in community. Self-mobility limited.
		Level 5: even with assistive technology, self-mobility severely limited.

Why does this matter in primary care?

People with cerebral palsy have increased prevalence of long-term conditions

Compared with the adults without cerebral palsy, adults with cerebral palsy have an increased risk of:

Musculoskeletal disorders (Bone 2019;125:30-35)	<ul style="list-style-type: none"> • Osteoporosis (HR 6.19, 95% CI 3.37–11.39). • Osteoarthritis (HR 1.54, 95% CI 1.17–2.02).
Cardiovascular disease (Neurology 2019;93:e1385)	<ul style="list-style-type: none"> • Heart failure (HR 2.62, 95% CI 1.51–4.52). • Hypertension (HR 1.64, 95% CI 1.34–2.01). • IHD (HR 2.32, 95% CI 1.45–3.71). • Cerebrovascular disease (HR 5.53, 95% CI 3.04–10.06).
Respiratory disease (Neurology 2019;93:e1385)	<ul style="list-style-type: none"> • Asthma (HR 2.24, 95% CI 1.82–2.76).
Mental health (JAMA Neurology 2019;76(3):294)	<ul style="list-style-type: none"> • Depression (HR 1.28, 95% CI 1.09–1.51). • Anxiety (HR 1.38, 95% CI 1.15–1.64). <p><i>Note that when studies specifically looked at adults with cerebral palsy with and without intellectual impairment, only adults without intellectual impairment had an increased risk compared with the background population.</i></p>
Dementia	<ul style="list-style-type: none"> • Dementia (HR 2.69, 95% CI 1.44–5.00). <p>This association may be explained by the presence of sensory impairment, intellectual disa-</p>

(BMJ Open 2021;11:e042652)	bility and epilepsy. After adjusting for this, the presence of cerebral palsy alone is not associated with dementia.
Falls (DMCN 2020;62(4):477)	<ul style="list-style-type: none"> • More likely to fall (OR 3.64, 95% CI 2.98–4.45). • Experience more falls (rate ratio 5.83, 95% CI 4.84–7.02).

Morbidity is significant and impacts quality of life

Adults with cerebral palsy commonly experience pain, limited knee and hip mobility, decline in walking function over time and may need to use mobility aids (Arch Phys Med Rehab 2020;101:1041).

The most significant areas of unmet health need in the UK are (Child: care, health and development 2018;44(3):355)

- Pain.
- Bone and joint problems.
- Speech.

Transition from child to adult services can be difficult

Transition from child to adult services can be disjointed and variable, and it is estimated that at least 1/3 people with cerebral palsy have NO ongoing secondary care input. This challenge can be exacerbated because, during childhood, most will have been managed in paediatric services and may not have built a relationship with primary care. Communication and handover between secondary and primary care can also be very variable.

A 5-year UK-based study looked at how effective transition could be facilitated for people with cerebral palsy and type 1 diabetes (Clin Med 2020;20(1):74). Much of what was identified requires organisational and system-level change, but key points were:

- Appropriate parent involvement.
- Building children's confidence in managing their condition and wider health.
- Meeting the 'adult team' before transfer (*this includes primary care as we may be the adult team for some adults with cerebral palsy – can we make this happen?*).

The authors concluded that primary care should be kept better informed by hospital services, especially once the child has reached the age of about 12y following the move to secondary school – this may be a key time to start to build a relationship.

Beware diagnostic overshadowing

This occurs when we assume that a presenting symptom is due to the underlying neurological condition, e.g. that reported hip pain is due to cerebral palsy and spasm, rather than considering the possibility of osteoarthritis. Both clinicians and people living with long-term conditions can do this. We can challenge ourselves to keep an open mind.

NICE guidance on cerebral palsy in adults

The impact of cerebral palsy is completely individual. Some people are fully independent whereas others need 24h care. A person's care and support needs will depend on the severity of their cerebral palsy and associated comorbidities. Their needs may also change with time.

In the UK, 93% of adults have spastic cerebral palsy, and the majority can walk with or without assistive devices. A person's care and support needs will depend on the severity of their cerebral palsy and associated comorbidities. Their needs may also change over time. For example, about half of adults with cerebral palsy report a deterioration in their ability to walk over time.

Follow-up and management of cerebral palsy should be with a specialist, but, in some areas of the UK, services are thin/absent.

Management of comorbidities is likely to be in primary care. We should feel comfortable with what care to provide in primary care and when referral to secondary care is required.

Because of the highly-variable service availability across the UK, we may have to be a little creative and work in partnership with adults with cerebral palsy to get them the support they need.

Until service commissioning catches up, this may include attempting to tap into existing neurology, orthopaedic and rehabilitation pathways with a more specialist multidisciplinary team, e.g. neurophysiotherapy, occupational therapy.

Below I have summarised the parts of the NICE guidance on cerebral palsy in adults that are pertinent to primary care:

Cerebral palsy in adults (NICE 2019, NG 119)

Annual review with a specialist in neurodisabilities (secondary care) is required if **cerebral palsy and complex needs** and any of:

- Communication difficulties or learning disabilities.
- Living in long-term care settings.
- Living in community without sufficient support.
- Multiple comorbidities.

'Complex needs' is defined as: gross motor function classification system level 4 and 5 (see above).

For all other adults with cerebral palsy, consider regular reviews (*NICE does not state where this review should take place*) depending on their needs and preferences.

NICE places emphasis on the changing needs of adults with cerebral palsy as they go through life.

So, what should we cover in primary care? The NICE guidance suggests discussing:

- General function and participation.
- Mental health.
- Musculoskeletal problems, osteoporosis and fracture risk.
- Abnormal muscle tone and respiratory disorders.
- Eating and nutrition.
- Pain.
- Screening services.
- Supporting employment.

Note that [Up: The Adult Cerebral Palsy Movement](#) has produced a fantastic annual self-check form for adults to complete in anticipation of a clinical review. You can also find the link in the useful resources below.

Cerebral palsy review in primary care:

Area to cover	Details
Function and participation:	<ul style="list-style-type: none"> • Ask about any changes in ability to walk. • Ask about any changes in hearing, speech or communication. • Speech, language and communication needs can change over time. Referral to SALT may be required. • Occupational therapy may be required to assess and help with independent living, vocational skills and physical activity. • Electronic assistive devices may be of benefit. • Encourage physical activity to maintain general fitness, physical and mental health. • Ask about current work/if they would like to work and work-related goals. Are there any barriers to this? Refer to occupational therapy/vocational rehabilitation services for support if required.
Mental health problems:	<ul style="list-style-type: none"> • <i>Adults with cerebral palsy have an increased risk of depression and anxiety (BMJ 2019;364:l806).</i> • Ask specifically about mood, irritability, behaviour, social interaction and sleep. • Complex combinations of factors may contribute, e.g. physical problems, pain, communication difficulties, frustration or lack of stimulation. • Ask about mental health and take into account communication difficulties, comorbidities (e.g. epilepsy or pain), learning difficulties and side-effects of drugs when tailoring management. • NICE does not advocate the use/avoidance of any specific treatments in adults with cerebral palsy, and suggests following the current NICE guidance on mental health conditions/learning disabilities as appropriate to that specific individual patient.
Musculoskeletal problems:	<p><i>Adults with cerebral palsy are more likely to develop osteoarthritis and to develop it at a younger age (Bone 2019;125:30-35). Osteoarthritis affects 13% of 31–40-year-olds with cerebral palsy (BMJ 2019;364:l806).</i></p> <p>Musculoskeletal function may deteriorate. Early recognition of bone/joint disorders helps early treatment/improved outcomes.</p> <p>Refer to orthopaedics/musculoskeletal services if a bone/joint disorder is causing pain or affecting posture/function despite conservative measures in primary care. Conditions which may prompt referral:</p> <ul style="list-style-type: none"> • Cervical degenerative changes increase the risk of cervical myelopathy. If cervical myelopathy is suspected, this is a medical emergency: REFER THAT DAY. • Cervical instability or spondylosis. • Spinal deformity, e.g. kyphosis, scoliosis or lordosis. • Osteoarthritis. • Subluxation of hip, wrist or shoulders. • Biomechanical knee problems. • Abnormalities of foot structure. <p>Do not offer -ray for hip subluxation or spinal curvature unless patient in pain, or posture or function affect-</p>

	<p>ed.</p> <p>Orthotics:</p> <ul style="list-style-type: none"> • NICE recognises that devices such as splints are used to help improve position and functioning. • There was no evidence identified on the effectiveness of orthotic devices in adults with cerebral palsy. • NICE highlights this as an area for future research.
<p>Osteoporosis and fracture risk:</p>	<p>Adults with cerebral palsy have a higher risk of osteoporosis (Bone 2019;125:30-35) and falls (DMCN 2020;62(4):477).</p> <p>In this section, NICE refers us to its osteoporosis guideline (which is not the easiest of guidelines to follow). But there are discrepancies between the NICE cerebral palsy guidance and the NICE osteoporosis guidance (see our article on <i>Osteoporosis</i>)! However, there are some important points to remember:</p> <ul style="list-style-type: none"> • Adults with cerebral palsy commonly have low bone mineral density, especially if: <ul style="list-style-type: none"> ◦ Reduced mobility/weight-bearing. ◦ Anticonvulsant or PPI use. ◦ Previous low-impact fracture. • Consider a DEXA if 2 or more of the following features (especially if previous low-impact fracture): <ul style="list-style-type: none"> ◦ Needing help with mobility, e.g. hoisting. ◦ History of falls. ◦ Low BMI. ◦ Previous low-impact fracture. ◦ Other medical factors, e.g. steroids, which can impact bone health. • NICE also says ‘Consider assessing fracture risk if 1 or more of the above risk factors’. We think NICE means doing FRAX/QFracture, although it isn’t totally clear! (And neither risk adjust for cerebral palsy, although QFracture does have a box for ‘Live in a nursing home or care home’ which may be a marker of reduced mobility). • Do remember that in people under the age of 40y, fracture risk assessment tools can underestimate risk, so the NICE guidance on osteoporosis says if <40y, assess bone mineral density with a DEXA scan. • Consider referring to secondary care if a positive DEXA result or a high fracture risk. (<i>NICE does not say why it suggests this; however, we wonder if it is because most trials of osteoporosis drugs involve older people, and also perhaps because some standard osteoporosis treatments such as oral bisphosphonates may be difficult for some with cerebral palsy to take</i>). • Adults with cerebral palsy may have other risk factors for osteoporosis as per the general population – take these into account too.
<p>Eating and nutrition:</p>	<p><i>Adults with cerebral palsy are at risk of obesity or undernutrition (BMJ 2019;364:l806).</i></p> <p>Those with severe spasticity and dyskinesia may have increased metabolic rate and higher risk of malnutrition.</p> <p>Offer regular weight checks and calculation of BMI.</p> <p>At review, ask about:</p> <ul style="list-style-type: none"> • Difficulties with eating, e.g. coughing/choking when eating. • Changes in eating habits, e.g. ability to feed self, changes in appetite, food refusal, increased length of mealtimes. • Bowel function, e.g. constipation (can exacerbate spasticity/dystonia). • Frequency of chest infections: may indicate swallowing difficulties or gastro-oesophageal reflux. <p>The following may affect appetite, eating or weight: changes in carer, depression, pain, reduced physical activity, medication side-effects.</p> <p>Refer to dietician or SALT as required if difficulties with eating or malnutrition.</p>
<p>Abnormal muscle tone:</p>	<p>Main symptoms are of:</p> <ul style="list-style-type: none"> • Spasticity: hypertonia with resistance to externally imposed movement (Paediatrics 2003;111(1):e89). • Dystonia: movement disorder with involuntary/intermittent muscle contractions (Mov Disord 2010;25(11):1538). <p>Discuss the benefits/harms of treatment: some people use their spasticity/dystonia to help posture/ability to stand, walk or transfer. Treatment may affect this.</p> <p>If spasticity/dystonia cause functional impairment, pain or spasms, arrange review:</p> <ul style="list-style-type: none"> • Assess for and address any modifiable factors which may exacerbate spasticity or dystonia: <ul style="list-style-type: none"> ◦ Bladder problems (e.g. UTIs or bladder stones) or constipation. ◦ Emotional distress or pain. ◦ Posture or pressure sores. ◦ Change in home/work environment, e.g. seating. ◦ Medication changes and side-effects.





	<ul style="list-style-type: none"> • Review physical management programme before discussing further options. • If ongoing difficulties with spasticity causing functional impairment, pain or spasms despite the above, consider: <ul style="list-style-type: none"> o Baclofen orally or via a feeding tube (first-line drug treatment). Start low dose and increase gradually over 4w. o If baclofen ineffective/not tolerated, refer to/discuss other drug treatment options with specialist. o Do not offer diazepam for spasticity except if spasticity is causing severe pain/anxiety (acutely: short-term treatment only). o Do not rapidly withdraw muscle relaxant drugs (risk of life-threatening seizures). • If ongoing difficulties with dystonia, such as dystonia affecting function, participation or causing pain despite the above: <ul style="list-style-type: none"> o Refer to a specialist. o Levodopa should not usually be used to manage dystonia in adults with cerebral palsy. o Specialist may suggest specific drugs for dystonia. Do not rapidly withdraw these (risk of life-threatening seizures). • Secondary care may consider Botulinum toxin type A treatment/neurosurgery in certain circumstances for both spasticity and dystonia.
Respiratory disorders:	<p>Adults with cerebral palsy are at increased risk of respiratory failure.</p> <p>Symptoms which may prompt concern:</p> <ul style="list-style-type: none"> • Respiratory: shortness of breath. • Sleep: poor sleep pattern, sleep apnoea, daytime drowsiness. • Changes in behaviour (e.g. poor concentration, irritability), worsening epilepsy, headache on waking, increased frequency of chest infections. <p>If gross motor function classification system level 4 or 5, increased risk of:</p> <ul style="list-style-type: none"> • Aspiration pneumonia. • Chronic cardiorespiratory disorders (e.g. cor pulmonale). • Chronic supportive lung disease. • Kyphoscoliosis. • Poor saliva control. • Recurrent chest infections. <p>Refer to secondary care if concerns about respiratory impairment.</p> <p>If high risk of lower respiratory tract infection, refer to chest physiotherapy.</p> <p>If recurrent chest infections and dysphagia suspected, refer to SALT.</p> <p>Do not offer prophylactic antibiotics for lower respiratory tract infections in primary care. These may be considered by secondary care if high risk of respiratory impairment.</p> <p>Do offer vaccinations to adults with cerebral palsy, and their carers, in line with the national immunisation programme.</p>
Pain and fatigue:	<p>Pain is very common in adults with cerebral palsy, with 65% reporting pain that impacts their quality of life. For some adults with cerebral palsy, there may be difficulties with/barriers to communicating the feeling of pain.</p> <p>Pain assessment tools, e.g. numerical rating/visual analogue/facial pain scales or body maps may help.</p> <p><i>Fatigue is noticeable by its absence in the NICE guideline. Adults with lived experience report that “fatigue is one of the most frequent, and pervasive of symptoms, and often very misunderstood by people outside our community. Understanding and managing fatigue can be one of the most crucial factors in living our best life” (Up: The Adult Cerebral Palsy Movement).</i></p>
Screening services:	<p>Encourage adults with cerebral palsy to attend national screening service appointments for:</p> <ul style="list-style-type: none"> • Females: breast and cervical screening. • Males and females: colorectal screening <p>Screening service providers should ensure services are accessible to people with cerebral palsy.</p> <p><i>NICE does not make specific recommendations about other preventative health strategies, e.g. cardiovascular risk assessment, but we should be aware that adults with cerebral palsy are more likely to experience cardiovascular disease and cardiovascular disease-related death than adults without cerebral palsy. In order to avoid exacerbating health inequalities, we should be proactive in offering appropriate preventative healthcare.</i></p>
When to refer to secondary	<ul style="list-style-type: none"> • Deterioration in ability to carry out usual daily activities (<i>this may present as occupational problems, increased falls, increased pain or fatigue</i>).

care (MDT experienced in neurological impairment):	<ul style="list-style-type: none"> • Considering a neurosurgical/orthopaedic procedure which may affect ability to carry out usual daily activities. • Life circumstances impact on ability to carry out usual daily activities, e.g. pregnancy, parenting, decreased mobility (e.g. due to hip osteoarthritis), loss of care/support from a parent.
General concepts to recognise and address:	<ul style="list-style-type: none"> • Personal barriers to accessing care. Identify/help with these, e.g.: <ul style="list-style-type: none"> ◦ Healthcare professionals with training/expertise in communication skills. ◦ The need for an advocate if required. ◦ Support with social/emotional factors, e.g. fear of stigma. ◦ Support for mental health. • Physical/organisational barriers to accessing care: <ul style="list-style-type: none"> ◦ Access to buildings, appropriate equipment and changing/bathroom facilities as required. ◦ Local community transport services. ◦ Longer appointment times if required. • All with cerebral palsy have the right to a care and support needs assessment in line with the Care Act 2014. • Commissioners/service providers should develop local pathways for provision of the specialist care needed by adults with cerebral palsy.

Experiences of health services

A systematic review explored the experiences of adults with cerebral palsy using the health service. Here, we summarise the key themes for primary care (Dev Med Neurol 2022; 64(4):429). Adults with cerebral palsy:

- Felt frustrated about lack of information and clinician understanding about ageing with cerebral palsy.
- Experienced challenges accessing healthcare, including:
 - Accessibility problems, e.g. lack of disabled parking, reception desk height, examination couches that could not be adjusted, no manual handling equipment to enable transfer.
 - Lack of understanding of how to communicate with people who used communication devices.
- Experienced negative attitudes towards their disability and felt that cerebral palsy overshadowed their reasons for presenting.
- Reported positive experiences when health professionals listened to their needs and spent time working in partnership to problem solve (*we don't have to have perfect solutions and can acknowledge this together*).
- Valued having a consistent GP so they could build relationships and work in collaboration to navigate available services.

	<p>Cerebral palsy in adults:</p> <ul style="list-style-type: none"> • An increasing number of adults are living with cerebral palsy. • In primary care, we should offer review and discuss: <ul style="list-style-type: none"> ◦ General function and participation. ◦ Mental health. ◦ Musculoskeletal problems, osteoporosis and fracture risk. ◦ Abnormal muscle tone and respiratory disorders. ◦ Eating and nutrition. ◦ Pain. ◦ Screening services. • Refer for secondary care annual review if cerebral palsy and complex needs and any of: <ul style="list-style-type: none"> ◦ Communication difficulties or learning disabilities. ◦ Living in long-term care settings. ◦ Living in community without sufficient support. ◦ Multiple comorbidities. • Consider referral to secondary care if: <ul style="list-style-type: none"> ◦ Deterioration in ability to carry out usual daily activities. ◦ Neurosurgical/orthopaedic procedure which may affect ability to carry out usual daily activities. ◦ Concerns about respiratory impairment or considering prophylactic antibiotics for chest. ◦ Input from members of the MDT required, e.g. physio, occupational therapy, SALT, dietician.
	<p>Run a search for adults with cerebral palsy:</p> <ul style="list-style-type: none"> • How can we proactively engage with adults with cerebral palsy at the transition from paediatric care, particularly those who are being discharged without specialist follow-up? • Are you offering annual review to adults with cerebral palsy? • How could the wider primary care team support with this? • Are you aware what local pathways exist for accessing additional multiprofessional support for adults with cerebral palsy?
	<p>Useful resources:</p> <p><i>Websites (all resources are hyperlinked for ease of use in Red Whale Knowledge)</i></p> <ul style="list-style-type: none"> • Up: The Adult Cerebral Palsy Movement - annual self-check resource • Up: The Adult Cerebral Palsy Movement - primary care information leaflet
	

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