Cerebral palsy

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Accepted 4 January 2016
Published Online First 2 February 2016

ABSTRACT

Cerebral palsy has always been known as a disorder of movement and posture resulting from a non-progressive injury to the developing brain; however, more recent definitions allow clinicians to appreciate more than just the movement disorder. Accurate classification of cerebral palsy into distribution, motor type and functional level has advanced research. It also facilitates appropriate targeting of interventions to functional level and more accurate prognosis prediction. The prevalence of cerebral palsy remains fairly static at 2–3 per 1000 live births but there have been some changes in trends for specific causal groups. Interventions for cerebral palsy have historically been medical and physically focused, often with limited evidence to support their efficacy. The use of more appropriate outcome measures encompassing quality of life and participation is helping to deliver treatments which are more meaningful for people with cerebral palsy and their carers.

INTRODUCTION

A working party led by Rosenbaum and Bax in 2005 proposed a definition for cerebral palsy:

Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances occurring in the developing fetal or infant brain. Its motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication and behaviour, by epilepsy and by secondary musculoskeletal problems.[1]

This holistic approach is reflected in the way that cerebral palsy is classified, by motor type and distribution and also by functional level. Functional descriptors have enabled more meaningful research into its natural history and benefit of interventions.

Cerebral palsy has no cure and few disease-modifying interventions; symptom management is the mainstay of treatment. To ensure that interventions have meaningful outcomes for people with cerebral palsy, these must relate to the International Classification of Functioning, Disability and Health framework for disability: body structure, activity and participation.[2]

Diagnostics, assessment and interventions have come a long way since 1843, when William Little described stiffness associated with contractures in children with injuries to their developing brains.[3] Yet many interventions still have only a limited evidence base, focusing on body structure; many measures are taken by proxy due to communication and learning difficulties faced by people with cerebral palsy.[4] Life expectancy relates to the number of comorbidities; a significant proportion of those with severe comorbidities live to adult life, and thus cerebral palsy is an important entity for the adult physician.[5]

EPIDEMIOLOGY

Cerebral palsy registries from developed countries suggest its prevalence is 2–3 per 1000 live births.[6] The prevalence is significantly higher in children born prematurely: 40–100 per 1000 live births for those born below 28 weeks gestation.[6] In many surveillance studies, low birthweight is often quoted rather than gestational age as it is a more accurate measure; however, low birthweight often implies preterm birth. The prevalence of cerebral palsy in children born at term is fairly static but its prevalence in those with low birthweight has fallen and more recently plateaued.[7] Due to the increased survival of more preterm and severely affected children, the overall prevalence of cerebral palsy has not changed significantly with time.[8]

Multiple pregnancies are a recognised risk factor but the data are often confounded by gestational age and birthweight.[9]

McIntyre *et al*[10] in 2013 reported 10 risk factors significantly associated with

To cite: Wimalasundera N, Stevenson VL. *Pract Neurol* 2016;**16**:184–194.

cerebral palsy for children born at term: placental abnormalities, major and minor birth defects, low birthweight, meconium aspiration, emergency caesarean section, birth asphyxia, neonatal seizures, respiratory distress syndrome, hypoglycaemia and neonatal infections.

PATHOPHYSIOLOGY

The disturbance (injury) to the developing brain may occur in utero, around the time of delivery, in the post-neonatal period, or later in early childhood. The injury often results from hypoxia, infection, stroke or hypotension, with the subsequent inflammatory cascade following the original insult.

There is no international consensus on the upper age limit of brain injury for the definition of postnatal cerebral palsy, but in practice, injuries up to the age of 2 years are commonly accepted as being cerebral palsy. It is important to note, however, that the pattern of injury and resulting disability can be very different in children with later-acquired brain injuries. Older children with acquired brain injury may already have developed skills such as walking and talking and therefore can build on these established skills during rehabilitation.

Several causes of cerebral palsy are rare in most developed countries, such as iodine deficiency, and rhesus disease leading to kernicterus and maternal infection.

Contrary to popular belief, hypoxia around the time of birth in term infants contributes to only around 10% of cases. To confirm this as a cause, there also needs to be evidence of encephalopathy (hypoxic–ischaemic encephalopathy).[11]

Around 10% of cerebral palsy is attributable to post-neonatal causes, including infection, hypoglycaemia, stroke and trauma, both accidental and non-accidental.[8]

Around 80% of cerebral palsy is caused by an in utero event with brain injury. These can broadly be divided into three distinct patterns, depending on when the brain injury occurred (box 1).

Early gestational disturbances, often before 20 weeks, can cause brain maldevelopment as they interfere with the migration of cells to their final destinations. This may result from infection, hypoxia or stroke but there are now genetic factors identified that interact with environmental influences.[12] Brain maldevelopments often result in more severe cerebral palsy phenotypes involving the whole body with a combination of spasticity and dystonia, and significant comorbidities such as epilepsy, dysphagia, cognitive and communication impairment. (figure 1A–E)

Early mid-trimester injuries (24–32 weeks gestation) usually result in injury to the periventricular white matter. At this gestation, the periventricular area has the most vulnerable blood supply and is therefore compromised following hypoxia, infection or hypotension. In preterm infants, this area is also susceptible to injury following intraventricular haemorrhage and haemorrhagic parenchymal infarction of the surrounding cortical tissue. Due to the periventricular anatomy in relation to the homunculus—with leg fibres passing closest to the ventricle edge—injury to this area usually results in a leg-dominant spastic motor pattern (spastic diplegia). The larger the injury to the white matter the more extensive is the limb involvement; very extensive lesions involve the optic radiations affecting oromotor function (figure 2).

Injuries to the brain occurring around the time of birth (peripartum) can give phenotypes of varying severity, depending on the duration of the insult. Near to term, the brain is structurally and vascularly more robust and therefore injury occurs first in the areas of highest metabolic activity: this is typically the basal ganglia. The resulting pattern of cerebral palsy is characteristic: bilateral and dyskinetic (dystonia or choreoathetoid). In some situations other cortical structures and cognition are preserved. However, following prolonged hypoxia, there may be a mixed motor pattern with spasticity and dystonia, combined with significant comorbidities (figure 3).

Strokes, primarily infarcts, may also occur around the time of birth. Thrombi may form in the venous circulation and enter the arterial vasculature through arteriovenous connexions in the fetal circulation. This often results in middle cerebral artery territory infarction.[13] These children present with a unilateral spastic dystonic pattern, with the arm often more affected

Box 1 Patterns and causes of brain injury at different stages

Early brain injuries resulting in brain maldevelopment (often before 20 weeks gestation)
- Maternal infection—cytomegalovirus—polymicrogyria
- *COL4A1* genetic mutations—porencephalic cysts, schizencephaly
- *LIS1* genetic mutations—lissencephaly
- *GPR56* gene—polymicrogyria

Injuries in early/mid-pregnancy (24–32 weeks gestation)
- White matter disease of prematurity (periventricular leukomalacia)
- Hypoxia, hypotension, sepsis
- Pressure from intraventricular haemorrhage (haemorrhagic parenchymal infarction)

Late injuries to the developed brain
- Peripartum asphyxia
- Maternal infection
- Stroke

Post-neonatal brain injuries
- Meningitis/encephalitis
- Stroke
- Trauma—accidental/non-accidental
- Hypoxia—near drowning

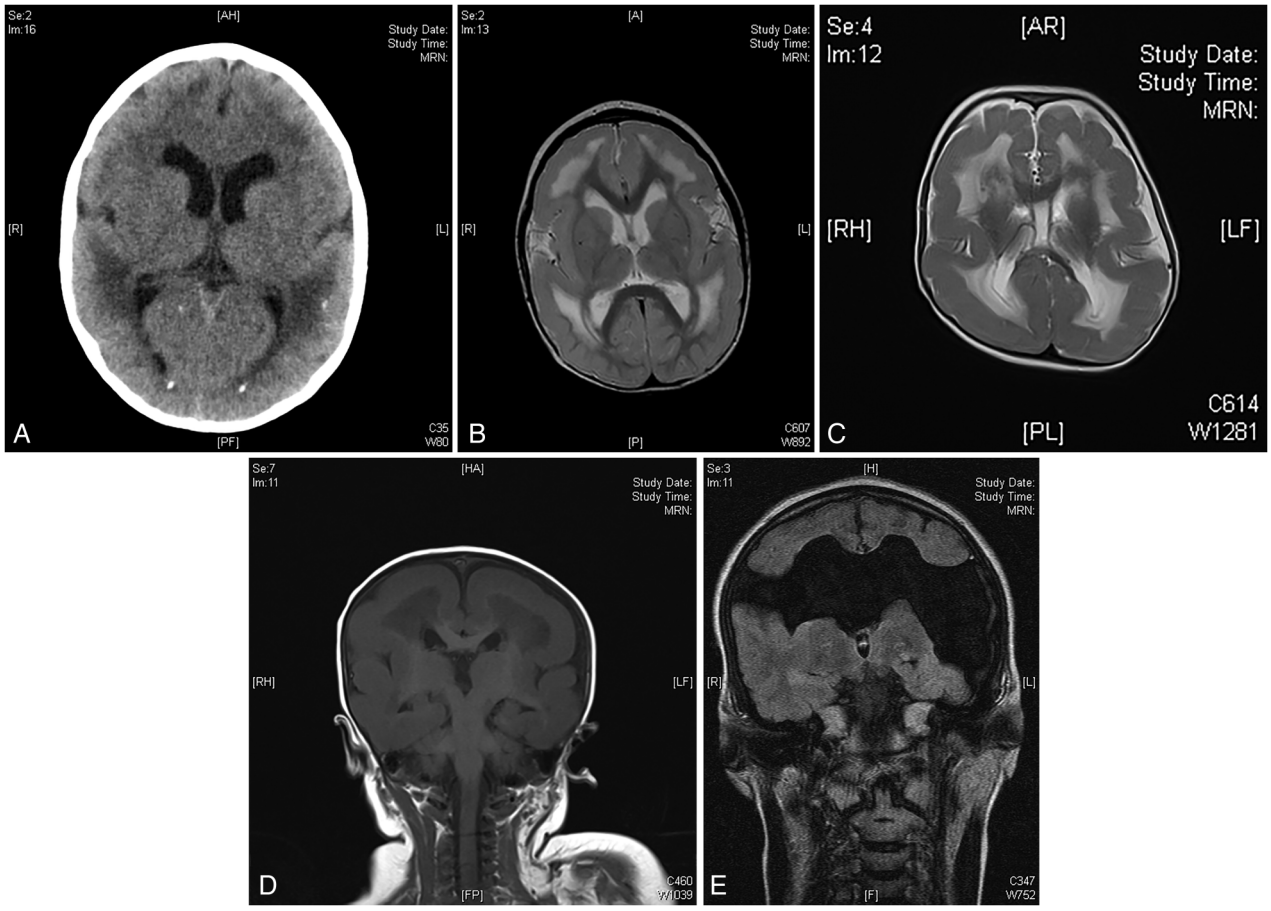

Figure 1 (A) CT scan of head showing intracranial calcification associated with early cytomegalovirus infection. (B) MR scan of brain (T2-weighted) showing extensive polymicrogyria typical of a congenital cytomegalovirus infection. (C) MR scan of brain (T2-weighted) showing lissencephaly, a cerebral maldevelopment often associated with the *LIS1* gene mutation. (D) MR scan of brain (T1-weighted) showing lissencephaly. (E) MR scan of brain (T1-weighted) showing bilateral open-lipped schizencephaly, which can be associated with *COL4a1* genetic mutation predisposing to early infarcts.

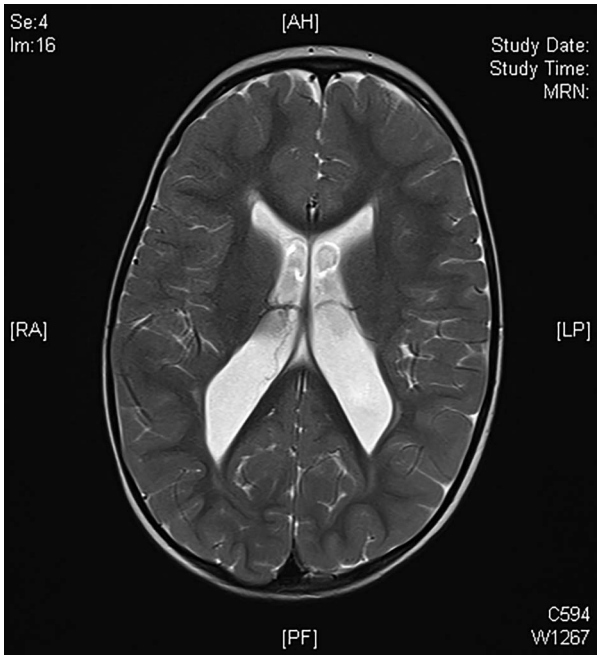

Figure 2 MR scan of brain (T2-weighted) showing periventricular leukomalacia associated with preterm (24–32 weeks gestation) injury to the brain.

than the leg. Facial weakness and dysphagia are uncommon due to bilateral innervation in these congenital injuries (figure 4).

DIAGNOSING CEREBRAL PALSY

When a clinician meets a person with cerebral palsy for the first time, the initial thoughts should always be, 'How robust is this diagnosis?' and 'What is the cause?' (box 2). These are particularly important questions for adult physicians as the person or their family (or even the medical records) might not remember appropriate details. Also, many adults with a diagnosis of cerebral palsy have not had neuroimaging.

Establishing the diagnosis and cause is essential to enable meaningful decisions regarding management and prognosis. Clinicians cannot assume that an expert in cerebral palsy made the original diagnosis, or that there were appropriate neurometabolic tests and neuroimaging.

Cerebral palsy is a clinical diagnosis but international guidelines suggest that the cause should be investigated with neuroimaging.[14] The extent of further neurometabolic tests to exclude other

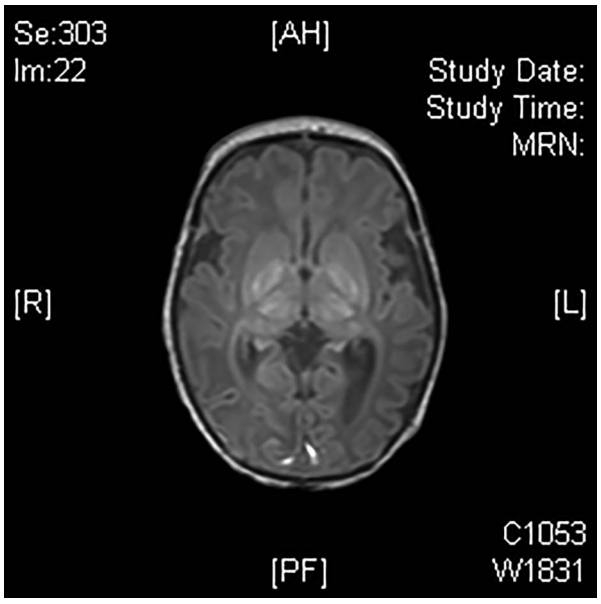

Figure 3 MR scan of brain (T1-weighted) showing bilateral basal ganglia enhancement associated with hypoxic–ischaemic injury to the term brain.

potential diagnoses depends on individual presentations. Around 10% of patients with cerebral palsy have normal neuroimaging.[15] Depending on the clinical context, this may suggest alternative diagnoses with, for example, hereditary spastic paraparesis. In reality, however, if the clinical diagnosis of cerebral palsy is robust from history and examination and neuroimaging is negative, then metabolic testing has a very low diagnostic yield.[16]

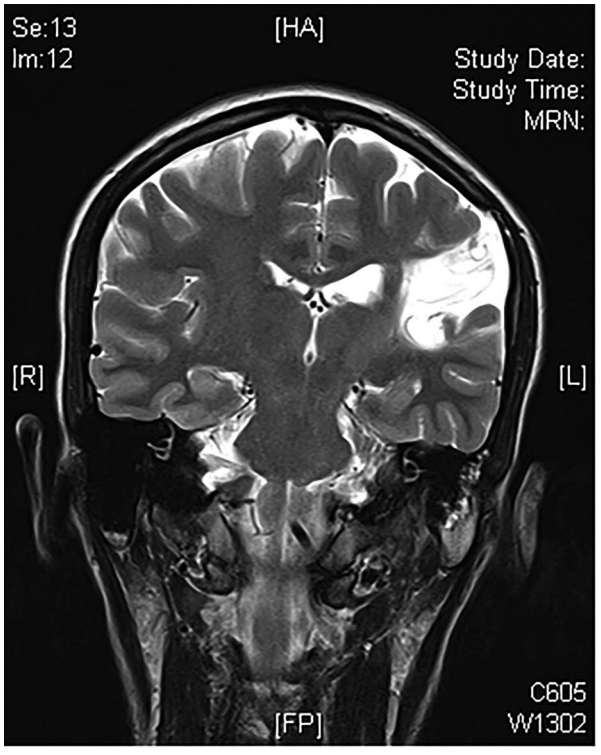

Figure 4 MR scan of brain (T2-weighted) showing typical mature changes following a perinatal middle cerebral artery infarction.

Box 2 Questions always to ask when establishing if cerebral palsy is the correct diagnosis

1. Do the clinical and antenatal histories suggest cerebral palsy?
2. Do the examination features match the clinical history?
3. Do the neuroimaging features fit with the clinical history and physical signs?
4. What is the functional level and does it fit the clinical pattern?
5. Are there features to suggest an alternative diagnosis?

The diagnosis of cerebral palsy rests on identifying and classifying the movement disorder. In some cases this can be recognised from as early as 5 months by a standardised assessment of general movements.[17] However, the diagnosis is usually made a little later, often in the second year of life when there are consistent signs and once neuroimaging is available. In the early years, the main clinical features may relate more to feeding than to overt motor difficulty.[18]

Ataxic cerebral palsy is an infrequent variant, often with normal neuroimaging. Many paediatric neurologists regard it as a diagnosis of exclusion, initially searching for alternative diagnoses.

CLASSIFICATION OF CEREBRAL PALSY

Despite the most recent definition highlighting that it is more than a movement disorder, cerebral palsy is classified by its motor type and distribution. Without this subclassification, the term cerebral palsy is too heterogeneous to be useful. Historically, it was grouped simply into mild, moderate and severe, or ambulant and non-ambulant, each of which had different meanings to different clinicians around the world. A significant advance was Palisano *et al*'s[19] functional motor classification in the mid-1990s. Classifying by motor type, distribution and functional level allows comparisons of like groups and more meaningful outcome studies.

The European surveillance of cerebral palsy group (SCPE) reviewed data from 16 countries in their initial epidemiology studies and described the distribution of limb involvement as unilateral or bilateral.[6] This terminology is now superseding terms such as hemiplegia, diplegia and quadriplegia as a more robust description of distribution.

The motor type is then described as spastic, dyskinetic (dystonia, chorea and athetosis), ataxic and mixed pattern[6 8] (box 3).

Box 3 Physical classification of tone and movement disorders in cerebral palsy, as recommended by surveillance of cerebral palsy in Europe collaboration[6 18]

Topographical (distributional) description for spastic cerebral palsy

- Unilateral
- Bilateral

Classifications of tone and movement abnormality

Spastic (85%–90% of cerebral palsy is spastic; one-third is unilateral, two-thirds are bilateral)

Velocity-dependent increased tone with hyper-reflexia and upper motor neuron signs

Tone increased but not necessarily constantly

Dyskinetic (7% of cerebral palsy)

Recurring, uncontrolled and involuntary movements that may be stereotyped

Tone abnormality varies

Dyskinetic cerebral palsy may be:

Dystonic—characterised by hypokinesia (reduced activity) and hypertonia (increased tone) resulting in stiff movements

Choreoathetotic—characterised by hyperkinesia (increased activity) and hypotonia (reduced tone) resulting in uncoordinated writhing and jerky movements

Ataxic (4% of cerebral palsy)

Generalised hypotonia with loss of muscle coordination —characterised by abnormal force, rhythm and control or accuracy of movement

Mixed forms

No single tone abnormality and movement disorder predominates. The most common mixed type has a combination of spasticity with dyskinesia.

Topographical and motor classifications do not give a sense of activity or participation; this is a clear advantage of functional classifications. The gross motor function classification system (GMFCS) measure is age-dependent and describes five groups according to the patient's level of mobility.[19] Level 1 indicates the person has minimal disability and level 5 indicates total dependence on equipment or carers to maintain posture (figure 5). The levels are independent of the cerebral palsy motor type and distribution. They do not indicate the quality of movement but simply the main way that the person mobilises. There are similar classifications for upper limb function, including the manual ability classification system; again level 1 indicates independence and level 5 indicates no hand function.[20]

The gross motor function measure scores physical activity, looking at activities such as rolling, walking, running and jumping. Gross motor curves have been developed from these scores for children at different GMFCS levels.[21] These graphs identify at what age there is peak motor function. For GMFCS level 1, 90% of potential motor development occurs at around 5 years; 4.5 years for level 2; 3.75 years for level 3; 3.5 years for level 4 and 2.75 years for level 5. Motor ability plateaus in the years after this, sooner in the more severe GMFCS levels, so this information can help to give a prognosis for walking. For example, a child with cerebral palsy aged 4 and GMFCS level 4 is unlikely to achieve independent walking in later life.

TREATMENT OF CEREBRAL PALSY

There are no 'cures', and symptom management is the mainstay of treatment, maximising developmental potential and minimising musculoskeletal deformity. The goal of any intervention is to modify the course of the disease but there are very few interventions in cerebral palsy with good evidence to support them.[4]

Improved antenatal and neonatal care have contributed to reduced severity of cerebral palsy in developed countries. There is good evidence that postnatal head cooling following hypoxic–ischaemic injury in term infants can improve survival and disability outcomes.[22] This is now common practice in the UK.

There are current stem cell trials in cerebral palsy management, targeting both the acute injury and also later in life. The evidence is only anecdotal at this stage, despite many services around world offering this treatment.[23]

All interventions offered to people with cerebral palsy should be done within the framework of the international classification of functioning, disability and health[2] (figure 6). Most interventions focus on body structure (eg, spasticity, contracture) but the most meaningful treatments then affect activity (eg, walking or running) and, most importantly, participation (facilitating social and employment engagement). The outcomes that clinicians regard as important are often not those that are important to people with cerebral palsy and their families.[24]

Treatments are generally matched to the GMFCS levels: mobility is a priority for the more able population, whereas posture, feeding, sleep and pain management are important to those more severely involved. For more severely involved patients with limited communication ability, clinicians should adopt a screening approach (box 4).

Novak *et al*[4] systematically reviewed the evidence for interventions in cerebral palsy; of the 64 discrete interventions, 24% were effective, 70% had uncertain effects with outcomes requiring close measurement and 6% were ineffective. Importantly, the effective interventions had evidence of benefit only at the body structure or activity levels of the International Classification of Functioning, Disability and Health, and not in participation. Clinicians should remember that, first, the lack of evidence for an intervention in this context does not necessarily mean lack of benefit

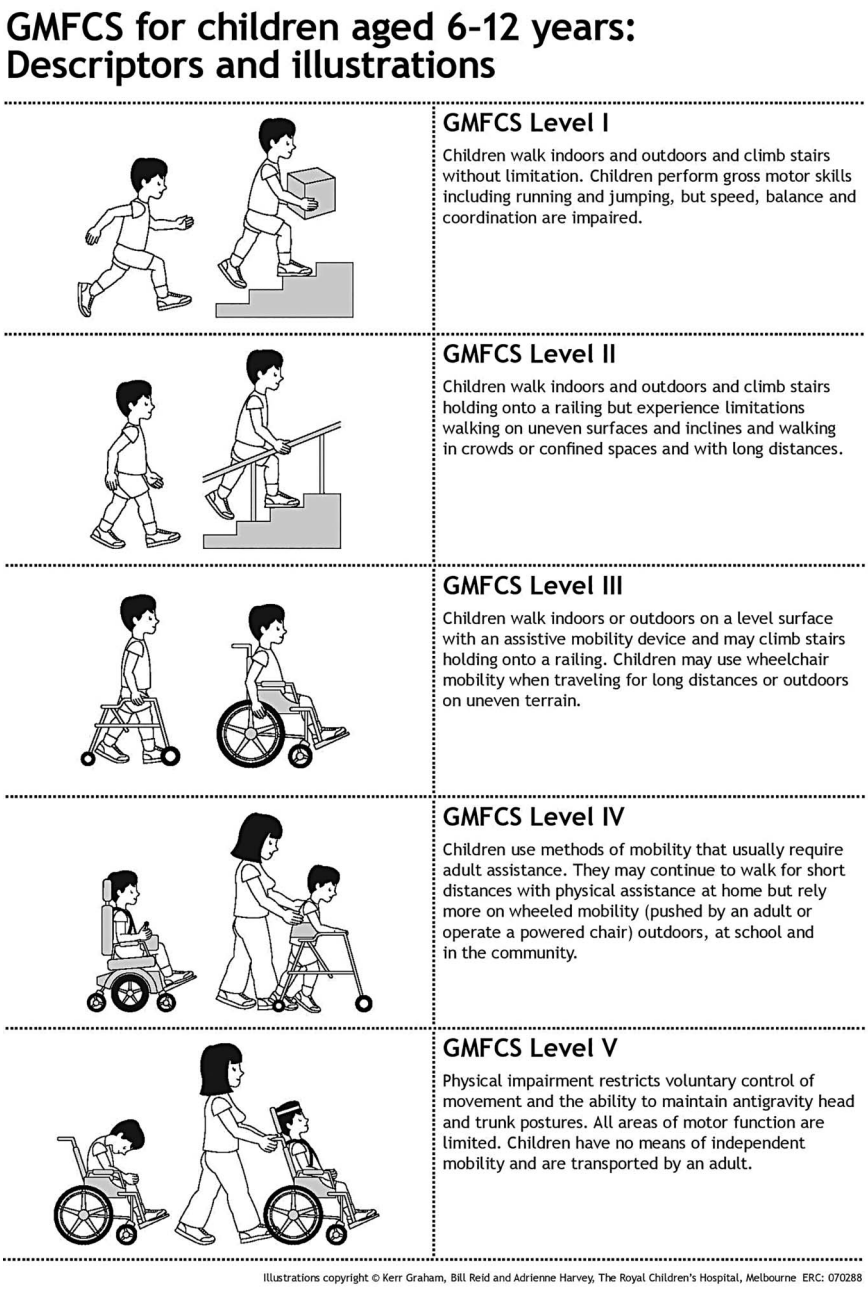

Figure 5 Gross motor function classification system to classify how a child with cerebral palsy mobilises and are classified according to age.[19]

and second, activity, participation and quality of life are the ultimate goals for any intervention.

The UK's National Institute of Health and Care Excellence (NICE) is currently developing guidelines for cerebral palsy management and already has guidance for spasticity management in childhood static brain injury.[25]

The treatment strategies for paediatric spasticity and dystonia are similar to adult practice but with notable nuances (box 5). In adult practice, if generalised or lower limb spasticity persists despite oral treatments, clinicians might consider intrathecal baclofen. However, in paediatric practice, this is usually reserved for severely affected patients with GMFCS of 4 or 5. Selective dorsal rhizotomy may be better for managing leg spasticity in bilateral spastic cerebral palsy (GMFCS level 2–4); this involves surgically dividing 50%–60% of the afferent sensory rootlets between L2 and S1. Park and Johnston have developed a newer minimally invasive technique for selective dorsal rhizotomy which has many fewer spinal orthopaedic complications and problems of long-term sensory or bowel and bladder disturbance.[26]

In 2014, NHS England commissioned five centres to provide selective dorsal rhizotomy in a 2-year evaluation project. We are collecting prospective data on spasticity, function and quality of life, before and after the procedure. International data suggest that

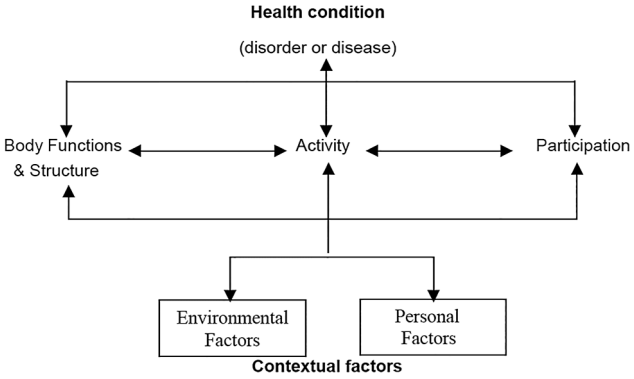

Figure 6 WHO. *International classification of impairment, activity, and participation*.[2] Does the intervention at the body/structure level affect activity and participation?.

selective dorsal rhizotomy successfully reduces spasticity in the long term and improves gross motor function in children (GMFCS level 2 and level 3) in both the short and longer term.[27] It also reduces the orthopaedic needs of children with cerebral palsy.[27]

PRESENTING PROBLEMS FOR THE ADULT NEUROLOGIST

Is the diagnosis correct and has there been a deterioration in function?

Although transition to adult services is improving, many young people with cerebral palsy unfortunately lose touch with health professionals during this time, sometimes due to lack of available specialist services and due to the pressures of normal adolescence and adult life. People with cerebral palsy therefore may not transition to adult services as expected when leaving paediatric services but may present to adult physicians only much later, often when things are going wrong.

The initial diagnosis may have been made at a very young age, often without neuroimaging; thus, the clinician should always reassess the diagnosis when meeting a person with cerebral palsy for the first time. Box 6 outlines potential clues to alternative diagnosis.

Even though cerebral palsy results from a non-progressive disturbance to the developing brain, the clinical signs and level of disability may change over time. This is not surprising considering the consequence of long-standing hypertonia, abnormal skeletal development and contracture formation.

Box 4 Screening areas for people with severe cerebral palsy and significant learning or communication impairment

1. Mobility and posture—standing, seating and lying tolerance, including equipment and orthotics—specific questioning regarding hip subluxation, kyphoscoliosis, hoisting and need for orthopaedic involvement
2. Dressing and hygiene—range of movement in legs and arms
3. Pain—dental, spasms, bone and joint-related
4. Respiratory status and drooling
5. Sleep pattern, quantity and quality
6. Gastrointestinal—gastro-oesophageal reflux, constipation
7. Feeding—nutritional adequacy, aspiration risk, fluid intake, bone health
8. Caregivers burden and concerns—stress, environment (adaptations), access to equipment including mobility aids and respite
9. Occupation—activity and participation of the individual
10. Adequate tone management—spasticity, dystonia, oral medications, botulinum toxin and intrathecal baclofen.

Box 5 Hypertonia management in childhood

Oral medications
- Baclofen (s/d)
- Diazepam (s/d)
- Tizanidine (s)
- Dantrolene (s) (rarely used in paediatrics due to adverse events—liver)
- L-dopa (d) (not very effective in paediatric secondary ds)
- Trihexyphenidyl (d) (first line for secondary d)
- Gabapentin (d) (increasingly used in difficult to control d)
- Tetrabenazine (d) (rarely used in paediatrics due to adverse events—mood)

Injectable (focal treatments)
- Botulinum toxin A (s/d) (intramuscular injection—useful for all muscles)
- Ethanol (s/d) (intramuscular injection—used in larger muscles)
- Phenol (s/d) (nerve block (ablation)—effective for larger muscle groups for example, adductors; obturator nerve)
- In the UK, botulinum toxin and intrathecal baclofen have superseded ethanol and phenol

Surgical treatments
- Orthopaedic (s/d) (muscle and tendon lengthening, transfers, disconnection)
- Intrathecal baclofen (s/d) (d needs higher doses—may exacerbate hyperkinesia)
- Selective dorsal rhizotomy (s) (only effective for s in legs)
- Deep brain stimulation (d) (more effective in paediatric primary ds than in secondary ds)
- d, dystonia; s, spasticity

Box 6 Features suggesting an alternative diagnosis to cerebral palsy

1. The history does not suggest a cause. For example, born at term but spastic diplegic pattern; no definite antenatal, perinatal or postnatal history of brain injury.
2. Pure signs, that is, pure dystonia or pure spasticity. For example, spinal cord lesions, hereditary spastic paraparesis, metabolic disorders; glutaric aciduria and mitochondrial disorders. Cerebral palsy often has a dominant motor type but usually there is a mixture of motor signs and often subtle arm involvement in leg dominant patterns. This is due to the 'crude' cause of the original brain injury.
3. Deteriorating or fluctuant course. For example, dopa-responsive dystonia; Segawa disease, primary dystonias, neurometabolic conditions; Wilson's disease, glucose transport protein 1 deficiency syndrome, neurodegeneration with brain iron accumulation; pantothenate kinase-associated neurodegeneration.
4. Diurnal variation in motor symptoms. For example, Segawa disease.
5. Strong family history. For example, primary dystonias: DYT1, hereditary spastic paraparesis, neurometabolic and mitochondrial disorders.
6. Dysmorphic features. For example, genetic (Angelman syndrome, Allan–Herndon–Dudley syndrome); or metabolic (molybdenum cofactor deficiency (lens dislocation), Lesch–Nyhan (self-mutilation), storage disorders (lysosomal)).
7. Functional level not in keeping with motor classification and distribution may imply more cognitive impairment than physical impairment.
8. Neuroimaging not consistent with clinical history or examination. For example, normal brain or spine MR scan, specific patterns for example, pontocerebellar hypoplasia, iron deposition in basal ganglia, progressive MR changes; Pelizaeus–Merzbacher disease.

Musculoskeletal deformity leads to biomechanical disadvantage and more energy inefficient gait patterns, such as crouch gait. These changes, combined with increase in height, weight and the natural age-related decline in muscle power, further reduce mobility (figure 7).

This decline begins as early as age 8.5 years of age in children with GMFCS level 3–5.[28] Despite the decline in motor function, the person generally stays at the same GMFCS level: any absolute change in GMFCS level may indicate an alternative progressive diagnosis.

In the more severe subtypes of cerebral palsy, especially those with feeding difficulties, low bone density can become problematic with advancing age. Low-impact fractures may even arise from normal handling and care. Low bone density develops through a combination of factors, including poor nutrition, inadequate sunlight exposure, antiepileptic medication and immobility with poor weight bearing in standing. Patients need their bone health monitored with biochemical markers such as serum vitamin D concentrations and, especially following fractures, bone density measurements. Patients may need bisphosphonates if low bone density has led to a fracture.

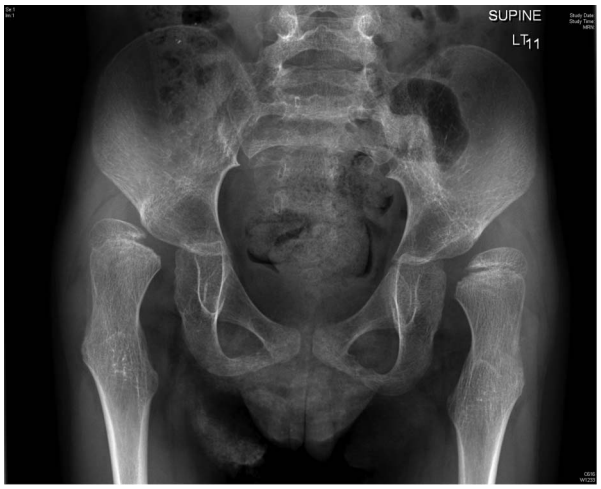

Figure 7 Progressive hip subluxation in a child with bilateral spastic cerebral palsy gross motor function classification system level 4 resulting in progressive reduction in mobility and pain (children with cerebral palsy have regular hip surveillance X-rays[25]).

Accurate diagnosis and classification, combined with an understanding of the natural history, allow a clinician to differentiate cerebral palsy from genuine progressive conditions.

If there is any decline in domains such as cognition, vision, communication or new motor patterns, then the person should be reassessed for a possible all-encompassing neurodegenerative condition or a secondary diagnosis.

Cervical myelopathy in dyskinetic (dystonic or choreoathetoid) cerebral palsy is something of which the adult physician should be particularly aware. Due to constant movement, the cervical cord is susceptible to injury: the person may present with worsening spasticity, bowel or bladder dysfunction, paraesthesias or weakness. These patients need cervical cord imaging and possible referral to a neurosurgeon for a stabilisation procedure.[29] Due to the pre-existing movement disorder and sometimes cognitive and communication difficulties in this population, there is often a delay in diagnosis.

Tone management in adult cerebral palsy

Although the fundamental principles of tone management in children and adults are similar, the treatment goals may differ. The focus in early childhood may be

to minimise orthopaedic intervention and to promote normal development of movement and posture. Input is often intensive with repeated treatments of botulinum toxin, splinting or selective dorsal rhizotomy. Severely affected children may be helped by deep-brain stimulation or intrathecal baclofen. In adults, the shift is more to long-term self-management with sporadic therapy and medical intervention. Goals are function-based and not aimed at treating the spasticity per se.

It is rare in adults to use widespread treatments with botulinum toxin to more than one limb, due to the number and size of muscles involved. In this situation, intrathecal baclofen is preferred and not considered a last resort; in adult practice, there is a move to use this in lower levels of disability specifically to improve or maintain walking.[30]

Deep-brain stimulation is used in childhood cerebral palsy but less commonly in adults; experience in adults suggests that it can give small improvements in the level of dystonia but with little impact on functional abilities. This contrasts with the success of deep-brain stimulation in other conditions, for example, Parkinson's disease or primary dystonia.[31]

Life expectancy

Life expectancy is clearly very important for people with more severe cerebral palsy. It is often discussed with families around the time of diagnosis but not again, until transition to adult services. Data on life expectancy vary, depending on the country, but the statistics are similar among developed countries.[32 33] What does differ is the method used and the adjustments for temporal effects, such as improved mortality for the general population or the availability of specific interventions such as gastrostomy.[34]

Life expectancy relates to the severity of mental, manual, ambulatory and visual impairment. It is only marginally reduced in patients with mild-to-moderate impairments.[5 9] Colver *et al*[9] reported that, in the UK, the survival of people with cerebral palsy alive at age 2 years with four severe impairments (IQ <50, non-ambulant, partially sighted and poor manual function) is 72% at 10 years, 44% at 20 years, 34% at 30 years and 27% at 40 years.

Apart from a recent review of Californian data, there is no evidence that life expectancy in cerebral palsy is improving.[9 32 33] This may partly relate to improved survival of very severely impaired children who previously might have died before having an established diagnosis. The Californian data suggested that over the past 30 years there has been improved survival of children with very severe disability and improved life expectancy in adults who are tube-fed.[32 33] The changes were more modest in among adults. This study stratified life expectancy according to motor ability and augmented feeding, both in a predictive manner from 4 years of age and in terms of additional years for adolescents and adults with cerebral palsy. Table 1 shows an example of additional life years for adults with cerebral palsy (male results are provided without parentheses, and female results are provided within parentheses in the table).[32 33]

As with all epidemiological studies on survival, these figures need adjusting for individual risk factors, such as scoliosis, hydrocephalus, seizures, cardiorespiratory status or endocrine insufficiency. It can be difficult to establish the actual cause of death in cerebral palsy as surveillance studies often note that the cause of death recorded on the death certificate is 'severe cerebral palsy'.

Management of seizures

Epilepsy is more frequent in children and adults with cerebral palsy than in the general population. Data from a meta-analysis involving 1918 children (Ashwal *et al*[14]) found that 43% (range 35%–62%) of children with cerebral palsy develop seizures, usually in the first year of life. In general terms, the more severe and global the cerebral palsy, the higher is the likelihood of epilepsy. Children may have generalised-onset or partial-onset seizures that do not correlate with their cerebral palsy phenotype. Control of seizures is often difficult in those with cerebral palsy, who may have more prolonged seizures and often require multiple antiepileptic drugs.[35]

The aims of seizure management, as in anyone with epilepsy, are to improve quality of life, reduce seizures and their associated risks, while avoiding or minimising medication adverse effects. Pragmatically, clinicians must often balance these, accepting that seizure

Table 1

	Additional life expectancy years			
	Cannot lift head			
Current age (years)	Tube-fed	Fed by others	Walks unaided	General population
15	14 years (14 years)	18 years (18 years)	52 years (55 years)	61.4 years (66.2 years)
30	14 years (14 years)	19 years (19 years)	39 years (43 years)	47.4 years (51.6 years)
45	12 years (12 years)	14 years (14 years)	25 years (29 years)	33.5 years (37.4 years)
60	7 years (7 years)	10 years (10 years)	15 years (19 years)	21.1 years (24.1 years)

freedom is not always possible without intolerable adverse effects.[36]

There are published guidelines to assist in the antiepileptic medication choice in childhood and adult epilepsy.[37] Cognitive and behavioural effects are the main issue with antiepileptic medication; avoiding polytherapy, slow titration and using the lowest effective dose can minimise these. It is extremely important to monitor such effects, along with seizure frequency and quality of life. In children with cerebral palsy who have failed on two or more antiepileptic medications, it might be worth considering a ketogenic diet.[36]

DISCUSSION

Finally, it is worth considering whether the name 'cerebral palsy' is appropriate nowadays. Sir William Osler coined the term in 'The Cerebral Palsies of Children' (1889). The root meaning of 'palsy' is 'paralysis' which clearly does not describe the impairment in the vast majority of people with cerebral palsy. Some also question whether the name 'cerebral palsy' has a similar stigma to previously used terms such as referring to a person with cerebral palsy as 'a spastic'; indeed the UK cerebral palsy charity 'Scope', formerly called 'The Spastics Society' changed its name in 1994.

As we gain further insights into the causes of cerebral palsy, it might be possible to replace this generic term for a label that defines its genetic, metabolic or other causes. However, for now we generally accept that this term is useful for patients, families and health professionals. The diagnostic term cerebral palsy must, however, be used accurately, including a description of the motor type, the motor distribution, the functional level (GMFCS) and comorbidities, to enable the medical, functional and social needs of this patient group to be properly addressed, through the use of guidance such as that from NICE.

Key points

- Cerebral palsy is caused by an injury to the developing brain; the timing of injury determines its type and associated comorbidities.
- About 80% of cerebral palsy is caused by an *in utero* brain injury; only 10% occurs around the time of birth and 10% occurs in early childhood.
- Injuries in early gestation lead to cortical malformations and the more severe phenotypes; mid-trimester injuries (24–32 weeks gestation) may result in a bilateral spastic leg dominant pattern (spastic diplegia) and injuries at term can cause a dyskinetic pattern.
- Cerebral palsy is classified according to its motor type, distribution and functional level (gross motor function classification system (GMFCS)) to facilitate appropriate intervention and prognostication.
- Clinicians should expect some motor deterioration in cerebral palsy at GMFCS levels 3, 4 and 5; however, if the GMFCS level changes or the pattern of deterioration is unexpected, consider an alternative diagnosis and repeat neuroimaging.
- There is no cure for cerebral palsy and any intervention should be decided jointly with the patient and carers, and be directed at improving activity and participation; do not underestimate the issues around communication, pain and sleep.

Contributors NW is the main contributor and author (researcher) of the paper. VLS was the primary author (researcher) on the sections on tone management in adult cerebral palsy and epilepsy management. VLS proofread and commented on the rest of the document, which was authored by NW. Both NW and VLS have had no funding assistance with the production of this paper. NW and VLS have no competing interests. Both are advisors on the NICE Cerebral Palsy guidelines group who are currently developing guidelines for cerebral palsy management. Production of this paper has been disclosed to the NICE guidelines group.

Competing interests None declared.

Provenance and peer review Not commissioned; externally peer reviewed. This paper was reviewed by Johann te Water Naudé, Cardiff, UK.

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Pract Neurol 2016 16: 184-194 originally published online February 2, 2016
doi: 10.1136/practneurol-2015-001184

Notes