Dinah Reddihough

Cerebral palsy in childhood

Background

Cerebral palsy is the most common cause of physical disability in childhood. While some children have only a motor disorder, others have a range of problems and associated health issues.

Objective

This article describes the known causes of cerebral palsy, the classification of motor disorders and associated disabilities, health maintenance, and the consequences of the motor disorder. The importance of multidisciplinary assessment and treatment in enabling children to achieve their optimal potential and independence is highlighted.

Discussion

General practitioners play an important role in the management of children with cerebral palsy. Disability is a life-long problem which impacts on the child, their parents and their siblings. After transition to adult services, the GP may be the only health professional that has known the young person over an extended period, providing important continuity of care.

Keywords: cerebral palsy; child; general practice

CPD

Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation.These disorders are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy and by secondary musculoskeletal problems.[1] This definition highlights the complexity of CP and the fact that it is not a single disorder, but a group of disorders with different causes.

Prevalence and causes

Cerebral palsy occurs in about 2.0–2.5 per 1000 live births.[2] Rates have remained fairly stable over the past 40 years.

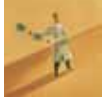
In many children, the cause of CP is unknown. Risk factors must be distinguished from causes. Known risk factors include low birth weight and prematurity, for example, infants of very low birth weight are 20–80 times more likely to have CP than infants of a birth weight of more than 2500 g.[3] It is likely that in a significant proportion of children, CP results from a series of events or 'causal pathways' that culminate in motor damage.[4]

Prenatal

Prenatal events are responsible for approximately 75% of all cases of CP and include brain malformations, maternal infections (the TORCH group of organisms – toxoplasmosis, rubella, cytomegalovirus and herpes simplex virus), vascular events such as middle cerebral artery occlusion, metabolic conditions and toxins.[4]

Perinatal

Perinatal causes are responsible for approximately 10–15% of cases and result from problems during labour and delivery such as antepartum haemorrhage or cord prolapse, compromising the fetus.[4] Significant perinatal events result in neonatal encephalopathy, a syndrome of disturbed neurologic function that is seen in the first days of life of a term infant, manifested by difficulty with initiating and maintaining respirations, depression of tone and reflexes, subnormal level of consciousness, and often, seizures. Minor perinatal events where the baby recovers rapidly do not result in CP.



Postneonatal

Postneonatal causes (occurring after 28 days of life) are responsible for about 10% of all cases of CP.[5] Causes include infections such as meningitis and accidental (eg. motor vehicle and near drowning accidents) and nonaccidental injuries.

A magnetic resonance imaging (MRI) brain scan should be undertaken where the cause is unknown.[6] Brain scans provide useful information about the timing of the brain injury, and there may be genetic implications. Common findings include periventricular white-matter injury; diffuse grey and white matter injury; and malformations.[7] In about 10% of children, the brain MRI scan is normal[6] and metabolic and genetic causes should be considered (see *Case study 1*).

Case study 1 – Jenny

Jenny, 5 years of age, has spastic quadriplegia, epilepsy and intellectual impairment. She is the third child of unrelated parents, born one Christmas Eve at term following a normal pregnancy. The second stage of delivery was quite rapid and her parents always felt that the CP was due to problems at birth. They were of the view that little care was given as many of the regular obstetric staff were on leave and those that were available seemed hurried and inexperienced. Jenny's Apgar scores were 7 and 10 and she developed transient tachypnoea of the newborn, requiring admission to a special care unit for a few hours of observation. No specific treatment was needed. Jenny's motor milestones were delayed and CP was diagnosed at 9 months of age. At 4 years of age, her parents asked their paediatrician about the risks of having further children with CP. A brain MRI was undertaken. This revealed a brain malformation, which provided a cause for Jenny's CP and timed the injury to 12–20 weeks gestation. The parents now understood that perinatal events were not responsible and were helped by this information.

Clinical assessment

Cerebral palsy is classified by:

- motor type
- topographical distribution, and
- severity of the motor disorder.

Spastic CP is the most common motor type with a prevalence of 70%. Dyskinetic CP (10–15%) is characterised by abnormalities of tone and various movement disorders including dystonia, athetosis or chorea. Ataxic cerebral palsy (less than 5%) is least common.

The terms 'diplegia', 'hemiplegia' and 'quadriplegia' are used to describe topographical distribution. In diplegia, the predominant problem is in the lower limbs but signs are usually also present in the upper limbs.

The Gross Motor Function Classification System[8] (GMFCS) provides information about the severity of the movement problems based on the child's motor abilities and their need for walking frames, wheelchairs and other mobility devices. There are five levels within the GMFCS: children in Level I and II walk independently; children in Level III need walking frames or elbow crutches (*Figure 1*); and children in Level IV and V require a wheelchair (*Figure 2*). Growth motor development curves for each of the GMFCS levels provide guidance regarding prognosis for motor development.[9] The Manual Abilities Classification System[10] (MACS) is a similar classification system that is used to describe how children with CP use their hands to handle objects in daily activities. It also has five classification levels.

Figure 1. Girl with walking frame

Diagnosis

Cerebral palsy presents with delayed motor milestones or asymmetric movement patterns. Children may also be identified when there is a history of prematurity with significant perinatal problems or neonatal encephalopathy.

Management

A team approach is essential, involving the general practitioner, other health professionals and teachers, and with input from the family. Caring for a child with CP involves an accurate diagnosis of cause (if possible), health maintenance, management of the associated disabilities, and the consequences of the motor disorder, referral to appropriate services and coordinated care and support for the family.

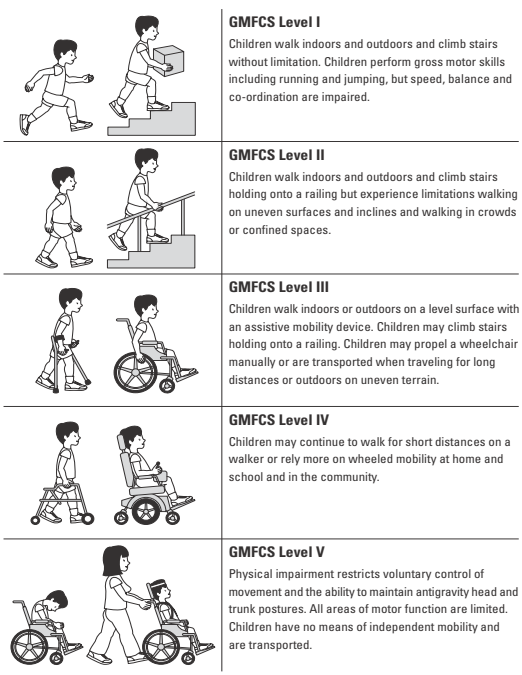
Health maintenance

Maintenance of good health is essential. The impact of intercurrent illness is often greater than in able-bodied children as food and fluid intake may deteriorate and recovery times are often longer.

- Growth should be monitored and dietary advice provided to ensure adequate nutrient and calorie intake. Obesity should be avoided. Failure to thrive is frequent due to eating difficulties resulting from oromotor dysfunction. Nasogastric or gastrostomy feeds should be considered if there is difficulty in achieving satisfactory weight gains, if the length of time taken to feed the child is excessive, or there is evidence of aspiration (see *Case study 2*)
- Gastro-oesophageal reflux is frequent and can result in

oesophagitis or gastritis, causing pain and poor appetite, and if severe, aspiration can result
- Constipation is common and dietary and laxative advice is important
- Up-to-date immunisations are important
- Chronic lung disease develops in some children due to aspiration from oromotor dysfunction or severe gastro-oesophageal reflux. Coughing or choking during meals, or wheeze during or after meals, may suggest aspiration. Barium videofluroscopy is a useful investigation
- Osteoporosis with pathological fractures may occur and can present with unexplained irritability
- Regular dental checks are important
- Skin problems are frequent. Buttock and perineal rashes are common, particularly in children who are incontinent; barrier creams may be helpful. Skin breakdown may also occur in body creases and around gastrostomy sites. Pressure areas over bony prominences may develop, particularly in children with poor nutrition. Pressure relieving devices and/or protective dressings should be considered with early involvement of nurses with specific expertise in continence and wound management
- Psychological and social difficulties need to be addressed.

GMFCS for children aged 6–12 years: Descriptors and illustrations



GMFCS Level I
Children walk indoors and outdoors and climb stairs without limitation. Children perform gross motor skills including running and jumping, but speed, balance and co-ordination are impaired.

GMFCS Level II
Children walk indoors and outdoors and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines and walking in crowds or confined spaces.

GMFCS Level III
Children walk indoors or outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto a railing. Children may propel a wheelchair manually or are transported when traveling for long distances or outdoors on uneven terrain.

GMFCS Level IV
Children may continue to walk for short distances on a walker or rely more on wheeled mobility at home and school and in the community.

GMFCS Level V
Physical impairment restricts voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Children have no means of independent mobility and are transported.

Footnote *GMFCS by Palisano et al.*
Palisano RJ, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol. 1997;45:113–120. Illustrated by Kerr Graham and Bill Reid, The Royal Children's Hospital, Melbourne.

Figure 2. Gross Motor Function Classification System
Reproduced with permission

Case study 2 – Craig

Craig, 12 years of age, is growing rapidly now he has entered puberty. He has severe CP; GMFCS Level V. Taking a meal has always been a slow task but his mother has not wished to embark on tube feeding, believing that it is preferable to be orally fed, even though it occupies several hours of her day. With rapid growth in length but virtually no weight gain, Craig appears very thin and develops a severe chest infection, requiring hospitalisation. During that admission, Craig's mother has the opportunity to talk to another parent whose child is partially fed with a gastrostomy tube. The parent of the other child explains that her daughter can still eat some foods orally but that she is no longer worried about daily intake. She now has more time to spend doing other activities that are enjoyable for the whole family. Craig's mother now agrees to gastrostomy placement with a good outcome for Craig and the family.

Management of associated disabilities

- Visual problems including strabismus, refractive errors and cortical visual impairment occur in children with CP, therefore referral to an ophthalmologist is important
- Hearing deficits are common, therefore all children with CP require a hearing test
- Communication disorders, including receptive and expressive language delays and articulation problems are frequent
- Epilepsy occurs in almost half of children with CP. Careful management is required
- Intellectual disability, learning problems and perceptual deficits are common. Children may benefit from a cognitive assessment.

Management of the consequences of the motor disorder

- Drooling (dribbling or poor saliva control) can be improved with speech pathology intervention, or by the use of anticholinergic medication or surgery
- Incontinence: bowel and bladder control may be achieved late due to cognitive deficits or difficulty in accessing the toilet because of physical disability and the inability to communicate. Detrusor overactivity may cause urgency and frequency. Advice about continence aids is useful
- Undescended testes usually require scrotal orchidopexy. The testes may be in the normal position at birth but ascend with time due to chronic spasm of cremaster muscle
- Orthopaedic problems: surgery is mainly undertaken on the lower limbs, but is occasionally helpful for the upper limbs. Physiotherapists are essential in the postoperative phase. Gait laboratories are useful in planning the surgical program for ambulant children
  - the hip: nonwalkers and those partially ambulant are at risk for hip dislocation, which causes pain and difficulty with perineal hygiene. Early detection is vital and hip X-rays should

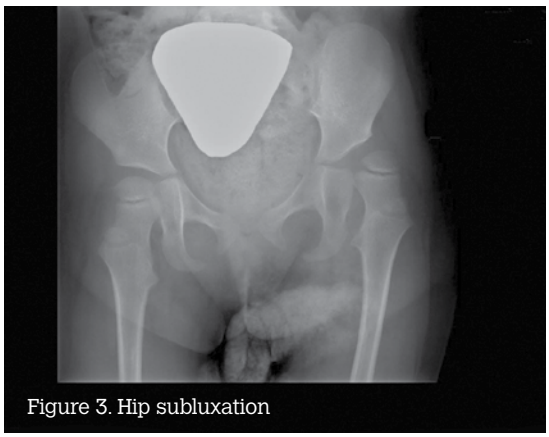


Figure 3. Hip subluxation

be performed at regular intervals according to hip surveillance guidelines.[11] If there is evidence of subluxation or dislocation (*Figure 3*), children should be referred for an orthopaedic opinion (see *Case study 3*)

- the knee: flexion contractures at the knee may require hamstring surgery to improve gait pattern, or the ability to stand for transfers
- the ankle: toe walking is treated conservatively in young children, with orthoses, inhibitory casts and botulinum toxin. Older children with equinus deformity require a definitive correction
- multilevel surgery: some children require surgery at several different levels, for example, hip, knee and ankle. This is called 'single event multilevel surgery' and is of most benefit to children who walk independently or with the assistance of crutches[12]
- scoliosis correction is sometimes necessary

- Spasticity management aims to improve function, comfort and care. Options include:
  - oral medications (eg. diazepam and baclofen)[13]
  - inhibitory casts to increase joint range, mainly used below the knee for equinus
  - botulinum toxin A to reduce localised spasticity[13]
  - intrathecal baclofen administered by an implanted pump (*Figure 4*); suitable for a small group of children with severe generalised spasticity and/or dystonia[14]
  - selective dorsal rhizotomy, an uncommon procedure whereby specific posterior spinal roots are sectioned to reduce spasticity.[15]

Case study 3 – Mark

Mark, 9 years of age, has presented to his family doctor on a number of occasions over the past 6 months. He has severe CP. His parents are concerned that he does not sleep and often seems uncomfortable during the day. Analgesics such as paracetamol have not been helpful. Physical examination demonstrates signs of quadriplegia. His left leg appears to be slightly shorter than the right leg. A pelvic X-ray demonstrates that the left hip is 60% subluxed. An urgent orthopaedic referral is made. Mark undergoes bilateral derotation osteotomies with relief of his pain and improvement in sleep patterns.

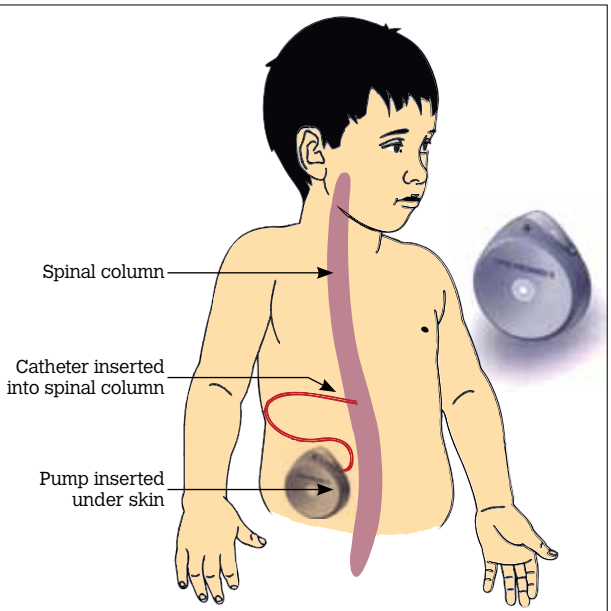


Figure 4. Baclofen pump in situ

Referral to appropriate services for the child and family

Therapy to address movement problems and to maximise the child's progress is incorporated into early childhood intervention and school programs. Physiotherapists, occupational therapists, speech pathologists, orthotists, dieticians, social workers, psychologists, special education teachers and nurses play an important role.

Services are best provided within local communities. Therapists and special education teachers work with children at home and later in childcare centres, kindergartens and schools. Most children attend mainstream preschools and schools but others benefit from centre based early childhood intervention programs and special schools. Parents should be made aware of all available options. Some hospitals provide allied health services.

Appropriate equipment can enhance communication, mobility, learning and socialisation. Examples include powered wheelchairs, electronic communication devices, and computers for educational and recreational purposes (see *Case study 4*).

Care of the child with CP involves developing a trusting, cooperative relationship with the parents. The child is part of a family, and parental concerns and questions must be addressed.[16] Parents may need practical support, such as provision of respite care, being made aware of financial allowances, and may find parental support groups helpful.

Case study 4 – Robert

Robert is aged 4 years. His parents are concerned that he is still not sitting independently and wish to discuss an alternative therapy, which claims to have good outcomes in helping children to walk. The GP explains that Robert is currently functioning at GMFCS Level V and the growth motor curves are shown to the parents. Following many discussions and substantial grief and disappointment,

they come to understand that Robert will never walk given his GMFCS level. They begin to concentrate more on his communication abilities and use the money that they had saved for alternative therapy to help purchase an electronic communication device.

Transition to adult care

Transition to adult care needs to be carefully planned and the GP has a vital role at this crucial stage: this is a time of life when not only is there transition of health services but also transition from school to employment or adult facility, and from living at home to living independently or in supported accommodation. In many parts of Australia there is a lack of tertiary health services for young adults with complex developmental disability, but transition clinics are slowly being established in some adult hospitals and rehabilitation physicians are providing input in many parts of the country.

Important practice points

- Search for the cause of CP as this is likely to be helpful for parents and may assist with genetic counselling.
- From the age of about 2 years it is possible to predict likely motor outcome. Advice can be given about the likelihood of independent walking or whether the child will be wheelchair-dependent.
- Ensuring an up-to-date immunisation schedule is essential. Yearly influenza vaccine is important for those with severe motor impairment.
- Regular hip surveillance is imperative to prevent the secondary problems of subluxation and dislocation.
- Consider the needs of the family, for example, has suitable lifting equipment been provided for a nonambulant child; has respite been discussed; do the needs of siblings require attention?
- When a child with CP presents with pain or irritability, consider the following possibilities:
  - intercurrent infection (particularly urinary tract infection)
  - dental disease
  - pathological fracture
  - gastro-oesophageal reflux and/or gastritis/oesophagitis
  - constipation
  - renal stones (increased risk if on topiramate)
  - subluxating and dislocated hips
  - sleep deprivation, which in turn may be caused by pain or spasm
  - increased intracranial pressure (many children have ventriculoperitoneal shunts)
  - nonaccidental injury (under-reported in this group)
  - changes in home environment, school or family that may be impacting on wellbeing.

Resources

- Cerebral Palsy Australia: www.cpaustralia.com.au/
- The Cerebral Palsy Institute: www.cpinstitute.com.au/
- Department of Developmental Medicine, The Royal Children's Hospital. Cerebral palsy booklet: www.rch.org.au/emplibrary/cdr/CPBooklet.pdf
- We Move Worldwide Education and Awareness for Movement Disorders: www.wemove.org/
- CanChild Centre for Childhood Disability Research: www.canchild.ca/en/.

Author

Dinah Reddihough MD, BSc, FRACP, FAFRM, is a paediatrician, Developmental Medicine, Royal Children's Hospital, Clinical Professor, University of Melbourne and group leader, Developmental Disability Research, Murdoch Childrens Research Institute, Melbourne, Victoria. dinah.reddihough@rch.org.au.

Conflict of interest: none declared.

References

1. Rosenbaum P, Paneth N, Leviton A, et al. A report: the definition and classification of cerebral palsy April 2006. Dev Med Child Neurol 2007;109(Suppl):8–14.
2. Odding E, Roebroeck ME, Stam HJ. The epidemiology of cerebral palsy: incidence, impairments and risk factors. Disabil Rehabil 2006;28:183–91.
3. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. Surveillance of Cerebral Palsy in Europe (SCPE). Dev Med Child Neurol 2000;42:816–24.
4. Stanley F, Blair E, Alberman E. Cerebral palsies: epidemiology and causal pathways. London: MacKeith Press, 2000.
5. Reid SM, Lanigan A, Reddihough DS. Post-neonatally acquired cerebral palsy in Victoria, Australia, 1970-1999. J Paediatr Child Health 2006;42(10):606–11.
6. Ashwal S, Russman BS, Blasco PA, et al. Practice parameter: diagnostic assessment of the child with cerebral palsy: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology 2004;62:851–63.
7. Korzeniewski SJ, Birbeck G, DeLano MC, et al. A systematic review of neuroimaging for cerebral palsy. J Child Neurol 2008;23:216–27.
8. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol 1997;39:214–23.
9. Rosenbaum PL, Walter SD, Hanna SE, et al. Prognosis for gross motor function in cerebral palsy: creation of motor development curves. JAMA 2002;288:1357–63.
10. Eliasson A-C, Krumlinde-Sundholm L, sblad B, Beckung E, Arner M, Ohrvall A-M, Rosenbaum P. The Manual Ability Classification System (MACS) for children with cerebral palsy: scale development and evidence of validity and reliability. Dev Med Child Neurol 2006;48:549–54.
11. The Australasian Academy of Cerebral Palsy and Developmental Medicine. Available at www.ausacpdm.org.au/__data/assets/pdf_file/0007/14569/consensus_statement.pdf.
12. Kerr Graham H, Selber P. Musculoskeletal aspects of cerebral palsy. J Bone Joint Surg Br 2003;85:157–66.
13. Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society, Delgado MR, Hirtz D, et al. Practice parameter: pharmacologic treatment of spasticity in children and adolescents with cerebral palsy (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology 2010;74:336–43.
14. Albright AL. Intrathecal baclofen for childhood hypertonia. Childs Nerv Syst 2007;23:971–9.
15. Farmer JP, Sabbagh AJ. Selective dorsal rhizotomies in the treatment of spasticity related to cerebral palsy. Childs Nerv Syst 2007;23:991–1002.
16. Rosenbaum P. Cerebral palsy: what parents and doctors want to know. BMJ 2003;326:970–4.

correspondence **afp@racgp.org.au**