Management of cerebral palsy: the neurologist's view

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Cerebral palsy is a complex disorder which compromises motor abilities. Other systems are often involved and its effects on the child and their family may be profound. This article examines clinical aspects of cerebral palsy and discusses the approach to comprehensive management, with particular reference to the role of the neurologist.

Cerebral palsy (CP) is a diverse and complex condition. It is an umbrella term describing a persistent disorder of movement and posture caused by non-progressive pathology of the immature brain (Bax, 1964). In the developed world the incidence of CP has barely changed over 30 years and remains between 2 and 2.5/1000 live births, thus affecting around 1 in 400 children (Hagberg et al, 2001). However, understanding of the natural history of the condition has advanced considerably, as have the interventions available. While the pathology itself is static the manifestations of CP evolve over time. The motor disorder itself may lead to permanent muscle and joint changes, resulting in contractures, joint dislocation and scoliosis. In addition to the motor disorder, there are often difficulties in other areas such as communication, sensation and cognition (*Table 1*). The functional impact of these associated difficulties may ultimately be greater than the motor disorder itself and such children generally require a multidisciplinary input for effective and comprehensive management.

TABLE 1.
Non-motor features associated with cerebral palsy

Disorders of vision (especially squint) and hearing
Sensory impairment
Feeding difficulties
Dysarthria
Drooling
Bladder dysfunction and incontinence
Epilepsy (approximately 30%)
Hydrocephalus
Learning difficulties
Behavioural problems

This article will discuss the neurologist's role in the management of the motor and non-motor features of CP. Currently there are less than 60 paediatric neurologists in the UK, most working in tertiary centres. The majority of children with CP will never see a neurologist. Generally the medical management will be coordinated by a community paediatrician working as part of a larger child development team. Numerous professionals (some of whom are listed in *Table 2*) may be involved in the overall care of a child with CP, particularly the more severely affected child. In this situation a key worker is often needed to coordinate care and ensure effective communication between the team, child and family.

There are a number of areas in which the neurologist may work with the multidisciplinary team in meeting the needs of the child. These include:

- Investigation and confirmation of diagnosis
- Counselling and advice (aetiology, prognosis and future genetic risk)
- Assessment and management in specialist clinics, e.g. neuro-orthopaedic, dysphagia, epilepsy
- Periodic re-evaluation of diagnosis, management and priorities
- Assisting with transition to adult care.

For effective management, a holistic approach is needed with consideration of the broader medical and social issues. A comprehensive assessment, with accurate description of the medical features, will establish a baseline of abilities and difficulties against which any

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change can be measured. Problems can then be prioritized, goals set and appropriate treatment regimens agreed. The accuracy of the diagnosis of CP, as a result of static underlying pathology, should also be evaluated. The aetiology and differential diagnosis is wide and there are many children with 'CP' who have never been investigated. As yet there are no agreed guidelines for the investigation of CP, however, the majority of children warrant neuroimaging as a minimum. A number of features should alert the clinician and prompt further investigation: these include unexplained CP in the absence of risk factors, a positive family history, and unusual clinical features such as episodes of encephalopathy or regression. *Table 3* lists some of these 'warning signs'.

An understanding of aetiology confirms the diagnosis and while investigations may not alter management or treatment, they are most important for genetic advice and often valuable for the child and parents in understanding the condition. In a number of children 'CP' is a misdiagnosis; a small number will have a treatable condition, such as a dopa-responsive dystonia or spinal cord tumours. Others may have a metabolic or degenerative condition such as mitochondrial disease or spinocerebellar ataxia. Spastic diplegia may be hard to distinguish from familial hereditary spastic paraplegia or, initially, from arginase deficiency. The genetic and management implications of an alternative diagnosis may be profound.

In 'classifying' CP, both the 'geographical' pattern of involvement and the neurological features of tone and movement are considered, e.g. dystonic hemiplegia or spastic diplegia. The underlying neuropathology is very variable and at present is not used in the classification. A clear understanding of the type of CP allows better prognostic advice and also alerts the clinician to likely associations. These are outlined in *Table*

TABLE 2.
Members of the multidisciplinary team

Primary members	Developmental paediatrician	
	Physiotherapist	
	Occupational therapist	
	Speech and language therapist	
	Psychologist	
	Specialist health visitor	
	Close links with education and social services	Special educational needs coordinator
		Peripatetic teachers
		Social worker
Secondary members (determined by specific needs)	Paediatric subspecialties	Neurology
		Neurosurgery
		Orthopaedics
		Psychiatry
		Audiology
		Ophthalmology
	Orthotist	
	Dietician	

TABLE 3.
Features that should prompt further investigation

A positive family history
Dysmorphic features
An absence of risk factors
Appearance or progression of neurological signs
Marked functional fluctuation – episodes of encephalopathy or regression

4. Assessment may also highlight atypical clinical features which would prompt further diagnostic investigation, as discussed above.

The second section of this article will discuss some specific aspects of management; namely the assessment and management of tone, and some broader management issues including epilepsy, dysphagia, drooling and incontinence.

MANAGEMENT AND TREATMENT

The general aim of management in the child with CP is to optimize his/her functional outcome: assets should be promoted, 'handicap' reduced and the family supported. Management plans therefore need to consider the specific and individual needs of the child and his/her family. These will change with development and growth so that management plans will need re-evaluating on a regular basis. Before instigating any specific intervention all aspects of the child's management need to be considered and the priorities agreed. Important areas include:

- Motor function, mobility and posture
- Communication and feeding
- Other medical problems
- Education
- Social and emotional support.

MEDICAL MANAGEMENT OF TONE

By definition muscle tone will be abnormal in the child with CP. However, this is not in itself a reason for treatment; hypertonicity may aid standing and walking if there is underlying weakness. The clinician needs to determine if the abnormal tone is causing specific problems for the child, his/her carers and therapists, such as limiting function or compromising nursing care. The aims of any intervention then need to be agreed and, once instituted, regularly reviewed.

The assessment of tone is largely subjective, although the Tardieu and Modified Ashworth scales do attempt to quantify spasticity in a standardized way (Tardieu et al, 1954; Bohannon and Smith, 1987). Evaluating a child's tone in the clinic is often not representative of their normal state; tone is variable and influenced by many factors, including:

- Immediate factors such as anxiety and fatigue
- Medical conditions, e.g. shunt malfunction, epilepsy, gastro-oesophageal reflux or constipation
- Pain, e.g. from poorly fitting orthotics or skin trauma
- Medication.

Formal examination of joint ranges and video recordings provide a useful baseline. There are also standardized and validated measures of movement and function, in particular the Gross Motor Function Measure (Russell et al, 1989), the Pediatric Evaluation of Disability (Haley et al, 1992) and the Functional Independence Measure for Children (Msall et al, 1990) which can indicate meaningful change after any intervention. When treating tone a number of management strategies may be used, including:

- Physical management (physiotherapy and orthotics)

TABLE 4.
Classification of cerebral palsy

Type (frequency)	Motor features	Specific points
Spastic cerebral palsy (70%)	Pyramidal signs with distal weakness, dynamic hypertonus, hyperreflexia. (Reflects cortical damage)	Spastic diplegia (0.9/1000 live births). Pattern of involvement – lower limbs more involved than upper limbs resulting in delayed motor milestones and abnormal gait >10% non-ambulant. May show bulbar involvement affecting oromotor function. Non-motor associations: especially vision (squint) and hydrocephalus. Epilepsy and learning difficulties are found in the minority.
		Spastic hemiplegia (0.8/1000 live births). Pattern of involvement – unilateral, the infant often shows early hand preference, 50% walk late. Non-motor associations: especially sensory disturbance. Visual field defects, facial weakness, epilepsy and learning difficulties are found in the minority.
		Four limb cerebral palsy (tetraplegia/quadriplegia/total body involvement) 0.2/1000 live births. Pattern of involvement - all four limbs. Non-motor associations: the majority have severe learning difficulties, with epilepsy and microcephaly. Pseudobulbar palsy, visual impairment, incontinence and orthopaedic problems are common.
Dyskinetic cerebral palsy (dystonic or choreo-athetoid) (15%)	Extrapyramidal signs with variations in muscle tone, abnormal postures and involuntary movements. (Reflects basal ganglia damage)	0.3/1000 live births. Pattern of involvement – often hypotonic initially. Abnormal movements or postures evolve later. Non-motor associations: dysarthria, feeding difficulties and hearing loss
Ataxic cerebral palsy (5%)	Cerebellar signs with axial hypotonia, truncal oscillations and intention tremor. Scanning speech may be a feature	Particularly important to consider alternative diagnoses such as hydrocephalus, posterior fossa tumours and progressive ataxias
Mixed cerebral palsy (spastic/dystonic) is common		

Compiled from: Aicardi and Bax (1998); Uvebrant (1988); Veelken et al (1983)

- Pharmacological treatment (systemic or focal; *Table 5*)
- Surgery (orthopaedic surgery or selective dorsal rhizotomy)
- Alternative therapies.

For effective management it is important to work in a multidisciplinary setting with a physiotherapist, orthopaedic surgeon and an orthotist available. Physiotherapy remains the mainstay of management, as discussed in the accompanying article. An adjunctive trial of medication may also be indicated, either systemic or focal. The most commonly used are listed in *Table 5*.

In the UK baclofen is the most commonly used antispastic agent. A gamma aminobutyric acid ($GABA_B$) agonist, it reduces the release of glutamate and aspartate so increasing presynaptic inhibition. It is less likely to lead to tolerance and is less sedating than diazepam (Roussan et al, 1985). However, with oral administration baclofen has poor penetration across the blood–brain barrier and side effects often limit its use. Direct intrathecal administration into the CSF has theoretical advantages. In the USA there is extensive experience of the use of intrathecal baclofen in CP (see Albright and Neville, 2000). The procedure involves the insertion of an intrathecal lumbar catheter and an intra-abdominal pump, which can be refilled and programmed to allow dose modifications. This is, however, a costly and invasive procedure with a high complication rate and is not widely available in the UK.

Intramuscular botulinum toxin A injections are increasingly used for more focal spasticity. It is derived from *Clostridium botulinum* and irreversibly blocks acetylcholine release at the neuromuscular junction so reducing muscle contraction. The effects last 2–4 months until the neuromuscular junction is re-established. Botulinum toxin is licensed for use in treating dynamic equinus in ambulatory children over 2 years of age with CP. However, it is more widely used in both upper and lower limbs. It may also have a role in the management of drooling. If effective, injections can be repeated. Minor side effects are seen in around 10% of children and include bruising, excessive weakness, increased falls and incontinence. Antibody formation is uncommon (Bakheit et al, 2001).

Dystonia may respond to anticholinergics such as benzhexol, or a dopamine agonist such as L-dopa. If after investigation dystonia remains unexplained, particularly if it shows a diurnal variation, a 3–6-month trial of L-dopa is mandatory. The clinical spectrum of dopa-responsive dystonias is broadening and is sometimes only be confirmed by treatment (Furukawa and Kish, 1999).

OTHER MEDICAL ISSUES IN THE MANAGEMENT OF CP

Epilepsy

Children with CP have an increased risk of symptomatic epilepsy. The absolute risks are closely linked to the extent of cortical abnormality and therefore to the type and severity of CP. Overall the given risks range from around 25% in dyskinetic CP, rising in spastic CP to around 16–27% in diplegia, 30–50% in hemiplegia and 50–95% in tetraplegia (Wallace, 2001). Where CP co-exists with learning difficulties the incidence of epilepsy is much higher (71% in those with IQ<50 in the series of Hadjipanayis et al, 1997). In the general paediatric population the prevalence of epilepsy is between 3 and 6/1000.

TABLE 5.
Drugs commonly used in the systemic treatment of spasticity and dystonia

Condition	Drug	Mode of action	Dose	Side effects
Spasticity	Diazepam	$GABA_A$ agonist at central and spinal cord level	Child <10 kg 2 mg twice daily >20 kg 10 mg twice daily	Sedation, ataxia, weakness, tolerance and dependence
	Baclofen	$GABA_B$ agonist at spinal cord level. Increases inhibition	0.2–1 mg/kg three times daily	Sedation, ataxia (central hypotonia), weakness, altered seizure control
	Dantrolene*†	Inhibits calcium release from sarcoplasmic reticulum	0.5–3 mg/kg four times daily	Drowsiness, hepatotoxicity
	Tizanidine*	$\alpha 2$ adrenergic receptor agonist, inhibits polysynaptic reflex	Adults 2 mg three or four times daily	Drowsiness, dizziness, dry mouth, occasional hepatoxicity, hypotension
Dystonia	Benzhexol*	Central and peripheral anticholinergic action	Initially 1 mg twice daily Increase as tolerated by 1 mg every 4 days (often 2–3 mg/kg/day)	Dry mouth, pupil dilatation, urinary retention, constipation
	L-dopa*	Dopamine agonists may require 8–10 mg/kg/day	Initially 0.5 mg/kg twice daily	Nausea, agitation

*not licensed for use in children. †Regular check of liver function tests recommended. GABA = gamma aminobutyric acid

Neonatal seizures may be a manifestation of the cause of CP itself, in particular cortical dysplasia, or the result of hypoxic ischaemic encephalopathy or infection. In these cases the subsequent risk of CP and ongoing epilepsy is high. Many seizure types and epilepsy syndromes are described in CP (Wallace, 2001) and clinically it may be difficult to recognize seizures in the context of other involuntary movements. Neurophysiology is often helpful in diagnosis and management although the electroencephalogram (EEG) recordings may be difficult to interpret in the context of diffuse cortical damage. The significance of subclinical seizure activity is not known in children with CP, but is recognized as adversely affecting behaviour and cognition in other children. As with all paroxysmal episodes, a clear account and if possible a video of the event is helpful.

Treatment is principally with antiepileptic drugs. The choice of anticonvulsant should be guided by the seizure type and there are a number of helpful published reviews (Wallace, 2000). Sodium valproate is still the most widely used anticonvulsant for generalized seizures although lamotrigine, carbamazepine and topiramate can be highly effective. With topiramate anorexia (reported in up to 10% of adult patients) can limit its use in children with CP. The place of the newer anticonvulsants is still to be established. Vigabatrin remains the drug of choice in infantile spasms but more general use has been severely curtailed by the finding of irreversible concentric visual field loss in up to 50% of adult patients with long-term use (Miller et al, 1999). There remains a place for dietary intervention (ketogenic diet) in some cases. For intractable seizures where there is focal cortical pathology, epilepsy surgery may have an important role.

Dysphagia

The high incidence of feeding difficulties in CP has only recently been recognized (Motion et al, 2002). Difficulties are particularly prevalent in children in children with four limb involvement (Shapiro et al, 1986). While CP itself is likely to affect growth to some extent, as evidenced by hemiplegia, it is now recognized that in a significant number of children with CP, poor growth also results from inadequate intake of food (Stallings et al, 1993). The importance of adequate nutrition has now been demonstrated, not only for effective immune function and optimal skin condition, but also in cognition and mood (Rosenbloom and Sullivan, 1996).

The child with more severe CP is likely to be entirely dependent for feeding, so has limited autonomy over the timing, content and pace of mealtimes. Difficulties with the oral and pharyngeal stages of chewing and swallowing are often compounded by associated postural difficulties. Furthermore gastro-oesophageal reflux is common in this group of children. Reflux may cause pain, aversive feeding and behavioural changes and on occasions dystonic movements and spasms. Refluxed material may also result in pulmonary aspiration.

Clinical assessment requires a doctor, speech therapist, occupational therapist and dietician, often with support from psychology. It should include observation of a meal. Subsequently oesophageal pH monitoring, videofluoroscopy and chest X-ray may be indicated. Once posture and seating have been optimized, modifying utensils and altering the calorific content and texture of feeds may suffice. Significant gastro-oesphageal reflux warrants a trial of treatment. generally with antacids, pro-kinetics and an H_2 antagonist or proton pump inhibitor. If feeding is unsafe, because of aspiration, or is failing to meet the child's nutritional needs, non-oral feeding is indicated. Short-term nasogastric feeding may be instituted but ultimately gastrostomy placement is often required. In the presence of gastro-oesophageal reflux a Nissen's fundoplication may also be necessary.

Constipation

This is often distressing to the child and his/her carers. It may directly affect appetite and compromise bladder function and muscle tone (Clayden, 1996). Management strategies include dietary and pharmacological treatment.

Drooling

Drooling may have a serious impact on quality of life and be socially isolating. It may improve with postural management and medical treatment with anticholinergics, particularly hyoscine patches. Botulinum toxin may also have a role in the management of drooling. If persistent, salivary duct excision or diversion may be indicated (Blasco, 1996).

Incontinence

Persisting incontinence may largely be a reflection of cognitive impairment, particularly in more severely involved children (Roijen et al, 2001). However, bladder dysfunction is almost certainly under-recognized. In a population of children with CP referred to a specialist clinic, with incontinence, urgency and frequency,

investigation revealed a high incidence of urodynamic abnormalities, many of which were amenable to therapeutic intervention (Reid and Borzyskowski, 1993).

Hydrocephalus

The premature infant is at particular risk of developing obstructive hydrocephalus after intraventricular haemorrhage. The classical clinical signs are of a rapidly expanding head circumference and ultimately the evolution of clinical signs. This is usually detected while still in the special care unit and a neurosurgeon involved if shunting is required. Shunt malfunction must be considered if there is a change in neurological signs or function.

CONCLUSION

CP is a diverse and complex condition. It is important that the aetiology is understood. Each child has different individual needs. The array of treatments in CP can be bewildering for parents and professionals alike. Selecting the 'best treatment' is difficult and there is a dearth of objective evidence. For effective management, the value of the multidisciplinary team cannot be overstated and within this team the specific expertise of a paediatric neurologist may be required. A 'needs led' and goal-orientated approach is important, with regular and comprehensive reassessment. The common objective should be for the best possible outcome, preparing the child for adult life and maximal independence. HM

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KEY POINTS

- Cerebral palsy may affect motor, sensory and cognitive functions.
- A child with unexplained cerebral palsy needs investigation.
- To identify and prioritize the specific needs and goals of each child, comprehensive multidisciplinary assessment is necessary, with regular review.
- There should be a holistic approach to management with the involvement of a multidisciplinary team, working closely with social and educational services.
- Preplanning is needed for a smooth transition into adult services.