

MANAGEMENT OF CHILDREN WITH CEREBRAL PALSY

The term cerebral palsy refers to motor impairments that result from an insult to the immature motor cortex and/or its motor pathways.



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I would like to dedicate this article to Dr Leila J Arens, whose passion for managing children with cerebral palsy spans more than 4 decades, and has inspired so many of us to work in this field.

The neurological lesion causing cerebral palsy (CP) is static, but the clinical condition changes over time. CP has a wide clinical spectrum with a number of aetiologies. It is not sufficient just to make the diagnosis of CP; the term needs further qualification by a more detailed description of the clinical condition. There are different types of CP, ranging in severity and associated with a variety of problems. Doctors at primary and secondary level often feel inadequate, due to the complex and diverse nature of the CPs and their associated problems. There is also no magic cure, but rather intervention to allow the patient to reach maximum function and potential and to prevent further complications. This article aims to give a logical and sound approach to the assessment and management of the common problems and complications in patients with CP. These children may then be referred when a need for specialised intervention is identified, if they are deteriorating on current management, or if there is doubt about the diagnosis or management plan.

ASSESSMENT OF CHILDREN WITH CEREBRAL PALSY

Careful assessment of a patient with CP (Table I) will elicit problems specific to the individual child and allow the practitioner to plan the appropriate management. The following questions should be asked:

Is it cerebral palsy?

There must be evidence of an upper motor neuron (UMN) lesion, with brisk reflexes and possibly clonus; there should also be increased tone or persistent primitive reflexes. Hypotonic CP has many mimics and this diagnosis is best made at specialist centres.

What could mimic CP?

A number of CNS disorders mimic CP.¹ These include metabolic and genetic conditions. Be very careful of making the diagnosis of CP if:

- the history is not clearly compatible with the common causes of CP
- the child's development is regressing
- new clinical signs appear
- the clinical signs fluctuate during the day
- there is abnormal posturing
- there is a 'family history' of CP.

Table I. **Assessment of a child with CP**

- Confirmation of the diagnosis of CP
- Excluding the mimics
- Defining the aetiology
- The type of cerebral palsy
- The severity
- Associated medical problems
- Level of functioning
- Short- and long-term goals

What was the aetiology?

There is often a clear history of a CNS insult or high-risk situation like premature birth, but frequently no identifiable cause can be found.

What type is it?

CP is divided into different types according to the clinical presentation: spastic, hypotonic, dyskinetic, ataxic or mixed. It is further qualified according to the distribution of the limbs and the extent of involvement, e.g. quadriplegia, hemiplegia or diplegia, or the type of movement. The section on CP in Coovadia and Wittenberg² has a good description of the different types. The type of CP is not always pure and should be reviewed regularly, as the clinical condition may change over time as the brain matures (e.g. an infant with spastic quadriplegic CP may develop dystonic posturing later; children with hypotonia may evolve into the ataxic type later, and athetoid movements usually appear later).

How severe is the condition?

This is a clinical description based on the physical ability of the child. The traditional categories are: minimal (motor signs present but no functional impairment), mild (limited impairment), moderate (obvious impairment, usually requires assistive devices for ambulation), severe (little purposeful voluntary action).³

What are the associated problems?

Table II lists the common problem associated with CP that should be addressed during history taking and examination. In some cases the associated problem may be more problematic than the CP.

What is the child's level of functioning?

The description should include other deficits, and the level of functioning with and without assistive devices, e.g. patient is educable, able to walk using ankle-foot orthoses and holding onto the wall, otherwise crawls; moderate hearing loss corrected with hearing aids; speech is understandable, but slow and deliberate.

Table II. **Common associated problems/complications**

- Intellectual disability
- Epilepsy or seizures
- Visual impairment
- Hearing impairment
- Feeding/swallowing problems
- Gastro-oesophageal reflux
- Failure to thrive
- Constipation
- Dental caries
- Orthopaedic complications
- Financial
- Mental health of patients and carers

What are the short- and long-term goals for this patient?

Decisions should be made by a multi-disciplinary team, which should include the children and their family.

MANAGEMENT OF CEREBRAL PALSY

Management is aimed at minimising the progressive deformity, improving the child's functional outcome and managing the associated problems.

Physical rehabilitation

Physiotherapy

Physiotherapy is the mainstay of treatment for the majority of children with CP. The role of physiotherapy in improving the outcome (e.g. improving a spastic quadriplegic's mobility to enable them to walk) is controversial. However, there is clearly no doubt about the role of physiotherapy in maintaining the current function of the child and reducing the incidence and severity of further complications like joint contractures, deformities and dislocations. The success of medical and surgical interventions for CP is also highly dependent on rehabilitative exercises to optimise results.⁴

Physiotherapy is not just 'stretching exercises'. It is orientated towards activities of daily living by improving important components of movement, e.g. improving trunk control to improve a child's walking ability.

Neurodevelopmental treatment-trained (NDT) therapists have been trained in the complexities of movement at the different gross motor milestones and are invaluable in the management of these patients. Where resources are limited, teaching the caregiver to practise these techniques on a daily basis is the primary goal, with monthly visits to the therapist if possible.

Important aspects of physiotherapy

Hemiplegia

An infant using only one hand needs urgent referral to a therapist before the infant is upright and has learned to compensate totally with the unaffected side. Weight bearing on the affected arm is possible while the infant is learning to come up to sitting and crawling. The prognosis is good for the affected arm to weight bear and act as an assisting arm, even if the child is unable to fasten a button.

Severe spastic quadriplegia

Supine lying should be avoided if possible as this reinforces the abnormal extended position, with the child staring at the ceiling. Windswept position in supine should also be avoided to prevent scoliosis and dislocated hip.

Encourage the caregiver to place the child on its side, lying with a pillow between the legs. This will also bring the hands to the midline to play with a toy.

Prone lying over a longitudinal pillow with weight bearing on the forearms should alternate with side lying.

Never lose the feet. Always teach a caregiver to maintain 90 degrees of ankle dorsiflexion so that even a severely affected child can be placed in the standing position, if only for transfer from bed to chair. Lifting a heavy child is difficult.

Maintaining passive joint range is important for washing, especially hip abduction in side lying for girls during menstruation.

Speech therapy

General movement difficulties, as well as involvement of the oral muscles, may contribute to the failure to develop communicative intent, and cause speech delay and difficult feeding. Intervention should start early. The basic principles are to introduce a therapeutic feeding programme that includes positioning of the child, and techniques to facilitate mouth closure and appropriate chewing movements, plus blowing, sucking, licking etc. Receptive language development is often much better than expressive language development and these children's cognitive function should not be assessed on expressive language alone. Communication may be assisted by augmentative alternative communication like communication boards or home-made pictures to point at.

Occupational therapy

This should be goal-orientated to assist with activities of daily living, to improve or refine grasp and skilled fine motor movement and is especially useful for hemiplegic and dystonic CP patients.

MEDICATION

Diazepam (Valium)

This is useful only in severe spastic quadriplegia and severe disability, but it may cause cognitive slowing. The recommended dose is 0.12 - 0.8 mg/kg/day.⁵ Start with a single dose at night (quarter of half a 2 mg tablet) and if there is no problem with sedation, a 12-hourly dose may be given. A crushed tablet is better tolerated than syrup. Patients may have an idiosyncratic response and become alert, in which case the medication should be given in the morning. Tolerance may develop and the dose may need to be increased over time. Diazepam should be discontinued by slowly tapering the dose, as stopping abruptly may cause a withdrawal syndrome.

Clonazepam (Rivotril)

This drug is particularly useful in patients who also have myoclonic epilepsy. The recommended dose is 0.05 - 0.3 mg/kg/day divided 12 or 8 hourly. It is available as oral drops

and may increase oral secretions and care ought to be taken in patients with swallowing problems.

Baclofen (Lioresal) (10 mg tablet)

Sedation is less than with diazepam and this drug is useful in higher functioning patients. It has variable results but is worth a trial. Start at 2.5 - 5 mg per day and increase every 4 - 7 days to maximum doses of 30 mg (children 2 - 7 years of age) to 60 mg (children 8 years or older) per day in 3 divided doses. It is very poorly absorbed orally and high doses are usually needed. It may cause sedation, ataxia, hypotension and paraesthesia. Some patients may have increased seizures. Abrupt discontinuation can produce a withdrawal syndrome.⁵ Baclofen may also be given intrathecally as a continuous infusion into the subarachnoid space around the spinal cord. It is used for severe generalised spasticity with minimal CNS effects. It is not available in South Africa.

Intramuscular botulinum toxin type A

This causes temporary partial paralysis of spastic muscle and assists the antagonist muscles to strengthen, decreases shortening and positions the joint into a functional position. The effect lasts 3 - 6 months with minimal adverse effects. It will not reverse contractures. It must be used in conjunction with physiotherapy and most often with splinting or serial casting.

Assistive devices

Orthoses are used during activity to maintain joints in a functional position, or at rest to delay the onset of contractures and deformities. Their use facilitates skill development and improves gait efficiency. Hand and wrist splints and ankle-foot orthoses are the most common. They should fit properly.

Standing frames can be used for the non-ambulatory child, to improve musculoskeletal alignment, promote bone mineralisation and stimulate acetabular growth.

Correct supportive **seating** can maintain good posture and positioning and assist with improved function. Buggies are most commonly used for young children.

Surgical intervention

Orthopaedic surgery

Specialised paediatric orthopaedic surgeons are an essential part of the CP management team. Their knowledge of the dynamics of multilevel joint involvement in functional mobility is invaluable in determining the most appropriate management. Once there are contractures or joint abnormalities, surgical intervention is the only treatment available. Indications for surgery include improving the quality of movement and preventing subluxation or dislocation of joints. Orthopaedic interventions can be performed at any age and should be considered whenever an anatomical structure is at risk as a consequence of spasticity.⁶

Neurosurgery

Selective dorsal rhizotomy involves severing specific afferent nerve rootlets to decrease the reflex spastic arc. This is indicated for spastic diplegia with good trunk and hip girdle control, and severe spastic quadriplegia with hygiene problems. Patients should have regular physiotherapy.

Other

Hyperbaric oxygen

This is no longer considered as a treatment option following the results of a randomised controlled trial which showed no advantage compared with controls.⁷

MANAGEMENT OF COMMON PROBLEMS

Intellectual disability

Up to 10% of CP children may be profoundly intellectually disabled, including being blind and deaf. Assessing a patient's intellectual level may be very difficult in severe CP with associated problems. Children with athetoid CP are often labelled as having severe global delay even though they may

have normal intelligence. Beware of labelling a child with CP as being intellectually impaired when using assessment methods for normally developed children. It is the doctor's role to advocate for specific educational needs.

Epilepsy

Epilepsy is more common in children with CP than in the general population, especially in those with associated intellectual disability.⁸ Seizures should be treated according to the guidelines for epilepsy.

Vision

Visual deficits may impact negatively on the function of the child with CP. The clinical signs may be subtle, e.g. abnormal posture or position of head. If there is any doubt, visual acuity and visual fields should be formally assessed. Where vision is difficult to assess, visual evoked responses may be useful.

Hearing

Normal hearing is essential for speech development. Children with CP may react to loud sounds, but cannot hear the speech range. In children who already have a physical disability, and possibly a visual deficit, correction of a hearing deficit will have a major impact on their functional abilities. If there is any doubt, a formal hearing assessment should be performed. Behavioural testing is the gold standard, but the child must be able to sit unsupported and turn towards the sound. If not, a brainstem auditory evoked response is recommended.

Feeding problems

There are two distinct problems, which may occur separately or together:

- problems with sucking, chewing and swallowing
- gastro-oesophageal reflux.

Labelling a child as having 'feeding problems' is insufficient. The problem must be clearly identified, as the management of these two conditions differs.

Feeding problems are caused by:

- poor mouth closure or lip seal
- inco-ordinate swallowing

- tongue thrusting
- poor positioning while feeding (as this exacerbates all of the above)
- gastro-oesophageal reflux, with or without aspiration
- drooling (oral incompetence)
- dystonic posturing due to cold food or incorrect positioning.

The diagnosis is made by a careful assessment of the patient's feeding. One should ask about coughing during or after feeding, the time taken to finish a bottle or an average amount of porridge, tongue thrusting or spitting food out. Observation may reveal drooling of saliva, coughing, refluxing, or dystonic posturing. Dystonic posturing may occur in response to cold temperatures like being put down onto a cold surface, or being fed with a cold spoon, or due to pain from reflux oesophagitis.

The first step in management is to assess the positioning of the child while being fed. If the head/neck is hanging backward in extension it will be very difficult to swallow. Holding the child in the best physiological position will overcome many feeding problems (Fig. 1).

Gastro-oesophageal reflux may be managed by thickening liquid feeds (with maizena, custard, porridge) or by giving smaller amounts more frequently. Inco-ordinate swallowing may require feeding via a gastrostomy tube if a trial of feeding therapy fails. Patients with recurrent respiratory tract infections, failure to thrive, or taking an excessive time to feed should be referred to a specialist centre.

Drooling (oral incompetence)

Pseudobulbar palsy, inco-ordinate mouthing movements and poor swallowing mechanism contribute to excessive drooling in some children. Many may improve on a therapeutic feeding programme supplied by speech therapists. For those who do not improve, e.g. patients with pseudobulbar palsies, surgically transplanting salivary ducts into the posterior pharynx may help.

Dental hygiene

In severe CP, the discrete skilled movements of the tongue to clean the teeth are very poor. These children are



Fig. 1. The best position for feeding children with CP: A mother finger-feeding her infant to promote chewing.

often mouth breathers, which increases the formation of dental caries. Extra care and attention should be paid to oral hygiene; this should start before dental caries develop. If the child is oversensitive to the stimulation of a toothbrush, toothpaste etc., oral hygiene can be effectively introduced by wrapping a small piece of gauze or similar soft cloth over the finger, dipping it in a solution of salt and bicarbonate or a commercial mouth rinse, and rubbing firmly and swiftly over the child's gums.

Failure to thrive/nutrition

Spastic and hyperkinetic CP increases energy expenditure and therefore increases the daily calorie requirements. The child's calorie intake should be increased accordingly. Patients with severe CNS insults, especially if there are associated feeding problems, are also known to have slower growth rates with less muscle bulk. Nutrition staff should be educated appropriately about what to expect rather than blaming parents for their child's poor weight gain. The best advice is to give 'high energy and protein per volume feeds' e.g. mix 5 - 10 ml of oil or margarine into every feed, use peanut butter liberally, use eggs regularly, give high-energy drinks like milk, yoghurt and fruit juice instead of rooibos tea or water.

Constipation

Due to decreased movement and decreased fluid and fibre intake, many children with CP are constipated. This should be managed early and not left until the patient has severe faecal loading and discomfort. Routine management should include dietary changes like pureed dried fruit, but most patients require daily lactulose or sorbitol. Glycerine suppositories or fleet enemas are also useful.

Orthopaedic complications

Long-bone fractures are common in children in home-care facilities, especially if they are on anticonvulsants. These have been shown to be due to vitamin D deficiency.⁹ These patients should receive a multivitamin syrup with vitamin D. Patients with spastic quadriplegic CP are at high risk for hip dislocation. They should have hip X-rays at least

yearly and should be referred for a specialist orthopaedic opinion if there is evidence of subluxation of the hip joint (Fig. 2). Scoliosis may be minimised by maintaining correct positioning.



Fig. 2. Subluxation of the hip joint. A: Hilgenreiner's line: horizontal line through the triradiate cartilages. B: Perkins' line: Verticals from the outer edge of acetabulum at 90° to A. Subluxation is present if > 1/3 of the visible femoral head is lateral to Perkins' line.¹⁰ Note subluxation of left hip with superior displacement of the left femoral head. This X-ray also shows faecal loading.

Infectious diseases

All immunisations should be given as per schedule.

Mental health

Secondary health problems like behaviour problems, depression and anxiety are common in CP. One needs to be aware of this to identify them. The mental health of parents and caregivers should also be considered and counselling should be offered on an ongoing basis.

Financial problems

Visits to doctors, hospital pharmacies and therapists often drain the family's resources. Caregivers of children with severe CP are often unable to find employment, as their children require full-time care. These families may qualify for a care dependency grant and this should be offered to them.

CONCLUSION

Many children with CP can be managed at primary and secondary level as long as the practitioners have a logical approach to their management. It is not acceptable to say, 'Sorry, there is nothing

we can do for this child'. It may not be possible to correct the brain damage, but we can provide a life that is free from pain and discomfort, by anticipating problems and managing them appropriately and by preventing complications. There are interventions that may improve a child's function and quality of life. Hopefully this approach will assist the health practitioner to focus on the appropriate systems and allow for a more efficient consultation.

ACKNOWLEDGEMENT

I would like to acknowledge the invaluable input from Mrs Vivien Magasiner and Mrs Diana Novotny from the Western Cape Cerebral Palsy Association.

References available on request.

IN A NUTSHELL

Cerebral palsy is an umbrella term. It is not sufficient just to make the diagnosis of CP; the term needs to be further qualified by a more detailed description of the clinical condition.

The clinical signs may change over time.

The diagnosis and initial assessments should be made by professionals experienced in the field where possible.

If a sound and logical approach is used, patients with CP can be managed at primary and secondary level for a number of their problems.

Intervention is aimed at reaching maximum function and potential and preventing further complications.

Physiotherapy is the mainstay of physical treatment and early referral, good positioning and passive movements are paramount.

There are a number of associated problems that also need to be recognised and managed appropriately.

An experienced orthopaedic surgeon is an essential part of the team.