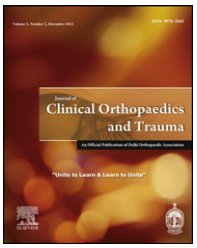
Available online at www.sciencedirect.com

SciVerse ScienceDirect

journal homepage: www.elsevier.com/locate/jcot



Review article

Cerebral palsy in children: An overview

Anil Agarwal [a,*], *Indreshwar Verma* [b,c]

[a] *Specialist (Ortho.), Department of Paediatric Orthopedics, CNBC, Geeta Colony, Delhi, India*
[b] *Senior Resident (Ortho.), Department of Paediatric Orthopedics, CNBC, Geeta Colony, Delhi, India*
[c] *Department of Orthopedics, CNBC, Geeta Colony, Delhi, India*

ARTICLE INFO

Article history:
Received 10 June 2012
Accepted 11 September 2012
Available online 18 September 2012

Keywords:
Cerebral palsy
Children
Management

ABSTRACT

A review article giving a brief synopsis of etiology, classification, diagnosis and management of cerebral in children.

1. Introduction

Cerebral palsy (CP) has been described as a group of disorders of the development of movement and posture that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behavior and/or a seizure disorder.[1] Diseases specific to the peripheral nerves of the spinal cord (e.g. spinal muscular atrophy, myelomeningocele) or to the muscles (e.g. muscular dystrophies), although causing early motor abnormalities, are not considered cerebral palsy.

2. Pathophysiology

The pathophysiology of cerebral palsy is not fully understood. The following events occurring during child's neural development occurring due to fetal, maternal, gestational or postnatal factors are may be responsible:

1. **Brain injury or abnormal brain development:** injury to the developing brain can occur anytime from gestation to early childhood. Contrary to popular belief, fewer than 10% of injuries occurring during the birth process result in cerebral palsy.
2. **Prematurity and postmaturity:** cohort studies have shown an increased risk of cerebral palsy in children born slightly preterm (37–38 weeks) or postterm (42 weeks) compared with children born at term (40 weeks).[2]
3. **Cerebral leukomalacia**
4. **Periventricular–intraventricular hemorrhage, hypoperfusion injuries** in the distribution of the middle cerebral artery, basal ganglia or other regions of brain.
5. **Cerebral infections or inflammations**

* *Corresponding author*. 4/103, East End Apartments, Mayur Vihar Ph-1 Ext, Delhi 110096, India.
E-mail address: rachna_anila@yahoo.co.in (A. Agarwal).

0976-5662/$ – see front matter
http://dx.doi.org/10.1016/j.jcot.2012.09.001

3. Epidemiology

In the developing world, the prevalence of cerebral palsy is not well established but estimates are 1.5–5.6 cases per 1000 live births.[3] The prevalence of this disorder among preterm and very preterm infants is substantially higher.[4,5] The incidence is higher in males than in females (ratio of 1.33:1).[3] Lower socioeconomic status may be a risk factor for cerebral palsy.[6] The approximate incidence of comorbidities in children with CP is mental retardation (IQ < 50): 31%; seizures: 21% and nonambulatory: 20%.[7]

4. Classification

1) Geographical classification (Table 1)
2) Physiological classification (Table 2)

Physiologically, cerebral palsy can be divided into a spastic type, which affects the corticospinal (pyramidal) tracts, and an extrapyramidal type, which affects the other regions of the developing brain. The extrapyramidal types of cerebral palsy include athetoid, choreiform, ataxic, rigid, and hypotonic.

3) Gross Motor Function Classification System (GMFC system)[8] (Table 3)

5. Diagnosis

5.1. *History*

The child with cerebral palsy can present with developmental delay or persistence of primitive reflexes. The history should include a detailed prenatal and perinatal history. Knowledge of normal motor developmental milestones and primitive reflexes allows identification of children who are delayed in their motor development. Important gross motor milestones of concern with cerebral palsy include head control at age 2 months, rolling at age 4 months, sitting at age 6 months, and walking at age 1 year.

Table 1 – Geographical classification of cerebral palsy.

Major types	Description
Monoplegia	• One extremity involved, usually lower
Hemiplegia (30%)	• Both extremities on same side involved • Usually upper extremity involved more than lower extremity
Paraplegia	• Both lower extremities equally involved
Diplegia (50%)	• Lower extremities more involved than upper extremities • Fine-motor/sensory abnormalities in upper extremity
Quadriplegia	• All extremities involved equally • Normal head/neck control
Double hemiplegia	• All extremities involved, upper more than lower
Total body	• All extremities severely involved • No head/neck control

Table 2 – Physiological classification of cerebral palsy.

Major types	Description
Spastic (80%)	• Velocity-dependent increase in muscle tone with passive stretch • Joint contractures are common
Athetoid	• Dyskinetic, purposeless movements • Joint contractures are uncommon • Dystonia or hypotonia can be associated
Choreiform	• Continual purposeless movements
Rigid	• Hypertonicity occurs in the absence of hyperreflexia, spasticity and clonus • "Cogwheel" or "lead pipe" muscle stiffness
Ataxic	• Disturbance of coordinated movement, most commonly walking • Normal head/neck control
Hypotonic	• Low muscle tone and normal deep tendon reflexes
Mixed	• Features of more than one type • No head/neck control

5.2. *Physical examination*

Classically, cerebral palsy becomes evident when the baby reaches the age of 6–9 months and initiates extremity mobilization, where preferential use of limbs, asymmetry or gross motor developmental delay is seen. Growth delay and persistent primitive reflexes may also be seen.

Before the formal physical examination, observation may reveal abnormal neck or truncal tone (floppy or very stiff); asymmetric posture, strength, or gait; or abnormal coordination.

In a bigger child, CP symptomatology is very diverse. In most common presentation, cerebral palsy is characterized by spasticities, spasms, involuntary movements, unsteady gait and problems with balance. There can be joint and bone deformities and contractures. Scissor walking and toe walking are common among people with CP who are able to walk.

5.3. *Associated conditions*

The most common associated conditions in patients with cerebral palsy are mental impairment or learning disability (40%); seizures (30%); complex movement disorders (20%); visual impairment (16%); malnutrition and related conditions, such as gastroesophageal reflux, obesity, and undernutrition (15%); and hydrocephalus (14%). Mental impairment and learning disability can range from very mild deficits to severe impairment and inability to live independently. Osteopenia

Table 3 – GMFC system of cerebral palsy and approximate percentage in each group.

I	Has nearly normal gross motor function – 35%
II	Walks independently, but has limitations with running and jumping – 16%
III	Uses assistive devices to walk and wheel chair for long distances – 14%
IV	Has ability to stand for transfers, but minimal walking ability; depends on wheel chair for mobility – 16%
V	Lacks head control, cannot sit independently, is dependent for all aspects of care – 18%

with increased risk of fracture also is common in severely affected children with cerebral palsy.[9–12]

5.4. *Investigations*

There are no definitive laboratory studies for diagnosing cerebral palsy, diagnosis being clinical.

If a diagnosis of a hereditary or neurodegenerative disorder is suspected, screening for an underlying metabolic or genetic disorder should be performed. Such studies may include thyroid function studies, chromosomal analysis etc. In addition, diagnostic testing for coagulation disorders is recommended if a cerebral infarction is seen.

Neuroimaging studies can help to evaluate brain damage and to identify children who are at risk for cerebral palsy. Cranial ultrasonography can be performed in the early neonatal period. Computed tomography (CT) scanning of the brain helps to identify congenital malformations, intracranial hemorrhage, and periventricular leukomalacia more clearly than ultrasonography. Magnetic resonance imaging (MRI) of the brain is most useful after 2–3 weeks of life and is the diagnostic neuroimaging study of choice for relatively older children. MRI also allows for the determination of appropriate myelination for a given age. However, patients who present clinically with cerebral palsy may have normal results from brain imaging studies and vice versa normal results from a neuroimaging studies do not exclude a clinical diagnosis of this disorder.

Electroencephalography (EEG) is useful in evaluating severe hypoxic-ischemic injury. This study is important in the diagnosis of seizure disorders.

5.5. *Gait analysis*

Before the development of computer-based gait analysis systems, careful clinical observation was the primary method of diagnosing gait disturbances in children with cerebral palsy. Clinical gait evaluation is still an essential component in CP examination. Modern quantitative gait analysis uses video cameras, retroreflective markers, and force platforms to measure the various components of gait. Kinematic data representing the three-dimensional motion of the joints during the gait cycle obtained are presented in a waveform pattern. Electromyography (EMG) is used to determine which muscles are firing in a normal/out of phase. Other components of quantitative gait analysis include pedobarography (foot pressure) and oxygen consumption measurement. Combined, these give an accurate representation of the complex interaction of all components of gait. Gait analysis frequently is used in preoperative planning before lower extremity surgery to delineate patient's gait deviations and plan the appropriate intervention.

5.6. *Treatment*

Because of the heterogeneous nature of cerebral palsy, an individualized approach to each patient is necessary. A multidisciplinary team approach comprising of physical, occupational, and speech therapy; orthotics; nutrition; social work; orthopedics; and pediatrics has been successful. Four basic treatment principles exist[9–12]:

1) Although the central nervous system injury, by definition, is nonprogressive, the deformities caused by abnormal muscle forces and contractures are progressive.
2) The treatments currently available correct the secondary deformities only and not the primary problem, which is the brain insult.
3) The deformities typically become worse during times of rapid growth as the bone lever and muscle length changes occur. For some patients, it may be beneficial to delay surgery until after a significant growth spurt to decrease the risk of recurrence.
4) Operative or nonoperative treatment should be done in consideration with patient's socioeconomic status and education.

5.7. *Nonoperative treatment*

Nonoperative modalities, such as medication, splinting and bracing, and physical therapy, are commonly used as primary treatment or in conjunction with other forms of treatment such as surgery.[9–12]

A wide variety of medications have been used to treat cerebral palsy. Diazepam and baclofen are centrally acting drugs. Baclofen mimics the action of g-aminobutyric acid, a powerful inhibitory neurotransmitter, whereas diazepam potentiates the activity of g-aminobutyric acid. These drugs have narrow therapeutic safety and careful monitoring is required to prevent overdosage. These medications pose systemic side effects including sedation, balance difficulties and cognitive dysfunction, which can have a detrimental effect on child's ambulation, learning and communication. Option of intrathecal baclofen pump is available and requires 1/30 the dose of oral baclofen to achieve a similar response. This method is usually recommended for patients where spasticity significantly interferes with self-care and quality of life.

Dantrolene, a peripheral acting drug, acts at the level of skeletal muscle and selectively decreases abnormal muscle stretch reflexes and tone. Dantrolene is used less frequently than other medications because some patients taking it develop profound weakness, and there is a risk of hepatotoxicity with its long-term use.

Botulinum toxin is a potent neurotoxin, produced by *Clostridium botulinum*. Botulinum toxin type A (BTX-A) has been used to weaken muscles selectively in patients with cerebral palsy. BTX-A injected directly into the muscle acts at the level of the motor end plate, blocking the release of the neurotransmitter acetylcholine and inhibiting muscle contraction. Because it can diffuse 2–3 cm in the tissues, it is easier to achieve the desired effect with BTX-A than with other agents, such as phenol or alcohol. It is also safer than these other agents because it binds selectively to the neuromuscular junction and not to other surrounding tissues. The effect begins approximately 24 h after injection and lasts upto 6 months. The most common side effects are local pain and irritation from the injection. The most common use of BTX-A is as an adjuvant to a bracing, casting, or physical therapy treatment program. It is beneficial in young patient in whom there is a need to delay surgery.

Physical therapy is an essential component in the treatment of patients with cerebral palsy. The therapist plays

a crucial role in all aspects of care of children with cerebral palsy. The parents should be encouraged from the beginning to take an active role in the child's therapy program.

5.8. *Operative treatment*

Operative treatment typically is indicated when contractures or deformities decrease function, cause pain, or interfere with activities of daily living. It is the only effective treatment when significant fixed contractures exist. Operative treatment of deformities related to cerebral palsy can be divided into several groups, including procedures to (1) correct static or dynamic deformity, (2) balance muscle power across a joint, (3) reduce spasticity (neurectomy), and (4) stabilize uncontrollable joints. Often, several procedures can be combined and children are offered multiple surgeries in one sitting.[9–12]

5.9. *Neurosurgical intervention*

The goal of selective dorsal rhizotomy is to surgically section the rootlets carrying excessive stimulatory information from the dorsal sensory fibers. The ideal patient for this procedure is a child 3–8 years old with spastic diplegia, voluntary motor and trunk control, pure spasticity, and no fixed contractures. Complications of selective dorsal root rhizotomy include hip subluxation and dislocation, lumbar hyperlordosis, spondylolysis, spondylolisthesis, and planovalgus foot deformities.

6. Regional interventions

6.1. *Hip*

6.1.1. *Adduction deformities*

Adduction is the most common deformity of the hip in children with cerebral palsy. Adduction contractures can cause various difficulties, including scissoring of the legs, hip subluxation, and in severely affected children, difficulty with perineal hygiene. For mild contractures, an adductor tenotomy usually is sufficient; more severe contractures often require additional release of the gracilis and the anterior half of the adductor brevis. Neurectomy of the anterior branch of the obturator nerve should be avoided to prevent iatrogenic hip abduction contracture.[9–12]

6.1.2. *Flexion deformities*

Excessive hip flexion brings the center of gravity anteriorly and is compensated for by increased lumbar lordosis, knee flexion, and ankle dorsiflexion. It is important to determine whether the increased hip flexion is the primary deformity or is compensatory to other deformities around the lower extremities, such as knee or ankle contractures. If an unrecognized knee flexion contracture is present, hip flexor release can weaken the hip further and increase hip flexion.

Hip flexion contractures of 15–30° are usually treated with psoas lengthening through an intramuscular recession over the pelvic brim. Contractures of more than 30° may require more extensive releases of the rectus femoris, sartorius, and tensor fasciae latae and the anterior fibers of the gluteus minimus and medius, in addition to the iliopsoas.

6.1.3. *Subluxation and dislocation*

Hip subluxation and dislocation occurs in 7% of ambulators and 60% of dependent sitters. The cause of this progressive deformity of hip is multifactorial and includes muscle imbalance, retained primitive reflexes, abnormal positioning, pelvic obliquity, acetabular dysplasia, excessive femoral anteversion, increased neck-shaft angle, and osteopenia.

Hip subluxation in patients with cerebral palsy can be difficult to detect clinically because of the presence of abnormal muscle forces and contractures, and because early hip subluxation typically is painless. Routine clinical and radiographic examinations should be done every 6 months, especially in rapidly growing children and patients with spastic quadriplegia. Clinically, hips with flexion contractures of more than 20° and abduction of less than 30° are at increased risk of progressive subluxation. Radiographically, a hip at risk has an increased neck-shaft angle and increased femoral anteversion. When a hip at risk is identified, a program of aggressive physical therapy and abduction splinting is started. If further progression continues, operative treatment consisting of soft-tissue release of contracted tendons is indicated. Operative correction of femoral valgus and anteversion and acetabular dysplasia is sometimes necessary at this stage. The treatment of an established dislocation is more controversial. A patient with a long-standing dislocation is not a good candidate for a relocation procedure because of the deformities of the proximal femur and acetabulum.

Resection arthroplasty, redirectional osteotomy, arthrodesis have been proposed for the treatment of a painful dislocated hip when a relocation procedure is impossible.

6.2. *Knee*

Deformities of the knee in patients with cerebral palsy are difficult to evaluate and rarely occur in isolation. Pelvic, hip, knee, ankle, and foot deformities are interrelated. Thus, a careful physical examination of the entire lower extremity is essential when evaluating the knee in patients with cerebral palsy. Important knee deformities are discussed below.

6.2.1. *Flexion deformity*

Flexion is the most common knee deformity in patients with cerebral palsy and frequently occurs in ambulatory children. Spastic hamstrings, weak quadriceps, or a combination of both can cause isolated knee flexion. Patients with spastic hip flexors or weak hip extensors or both develop compensatory knee flexion that results in a "jump gait," in which the hips, knees, and ankles are flexed. Patients with weakened gastrocnemius–soleus muscles from cerebral palsy or from Achilles tendon lengthenings, ambulate with knee flexion to accommodate for the relative overpull of the ankle dorsiflexors. The indications for hamstring lengthening are a straight leg raise of less than 70° or a popliteal angle of less than 135° in the absence of significant bony deformity. Care must be taken not to overlengthen the hamstrings because it can lead to excessive weakness and knee hyperextension gait.

6.2.2. *Stiff knee gait*

Stiff knee gait is common in patients with cerebral palsy. Cospasticity of the hamstrings and quadriceps causes a loss of

knee flexion that leads to decreased power and difficulties with foot clearance during the swing phase of gait. Gait analysis can highlight the spastic muscle. A transfer of the distal rectus femoral tendon to the semitendinosus medially or iliotibial band laterally is recommended, depending on the presence of malrotation.

6.2.3. *Recurvatum of the knee*

Recurvatum of the knee is caused by a relative imbalance between the quadriceps and the hamstrings owing to several factors, including (1) cospasticity of the quadriceps and hamstrings in which the quadriceps is stronger; (2) weakened hamstrings secondary to previous surgery, overlengthening, or transfer; (3) gastrocnemius-soleus weakness; and (4) ankle equinus. A significant isolated recurvatum should be treated with bilateral long leg braces with a pelvic band with the knees locked in 20° of flexion and ankle stops at 5° of dorsiflexion. When hip control is achieved, the pelvic band can be removed, but long leg braces often are used for years until a stable knee is obtained. Flexion osteotomy for this condition is not recommended.

6.3. *Foot*

Foot deformities are common in patients with cerebral palsy, with approximately 70%–90% of children affected. The most common deformity is ankle equinus, with equinovarus and equinovalgus deformities being equally common. A foot deformity can have significant effects on the patient's overall ambulatory level. Common foot deformities are described below.

6.3.1. *Equinus deformity*

Conservative treatment of eqiunus consists of stretching, bracing, and occasionally casting. Soleus stretching for 6 h a day can prevent equinus onset. Bracing, especially at night, to prevent the foot from going into the equinus position is essential. Surgery typically is indicated when the ankle cannot be brought into the neutral position in an ambulatory child and when it leads to difficulties with hygiene, shoe wear, and standing rehabilitation in a nonambulatory child.

6.3.2. *Varus or valgus deformity*

Varus and valgus deformities can occur in association with an equinus deformity. Valgus is more frequent than varus. It also is important to determine whether the deformity is flexible or rigid because flexible deformities are more likely to be successfully treated nonoperatively with orthotics, shoe modifications and operatively with soft-tissue procedures such as tendon lengthenings, releases, or transfers (usually of the abnormally active muscle). Patients with rigid varus and valgus deformities generally require bone procedures, such as calcaneal osteotomy, subtalar or triple fusions. Sometimes, the biomechanics of the hip, knee and tibia also influence the onset and outcome of a foot varus or valgus deformity and should be carefully evaluated. Other common foot deformities in children seen in cerebral palsy are forefoot adduction, hallux valgus and claw toes.

6.4. *Prognostic factors in cerebral palsy*

The persistence of primitive reflexes (asymmetrical tonic neck reflex, Moro reflex, extensor thrust on vertical suspension, absence of normal parachute reaction after 11 months, persistent neck-righting reflex) is associated with extensive and severe brain damage and a poor prognosis for independent ambulation, self-care, and activities of daily living. Approximately half of children who can sit independently by 2–4 years old eventually walk, and if a child cannot sit independently by 4 years, it is unlikely he or she will ever walk without assistance. Finally, if a child has not learned to walk by age 8 years, and he or she is not limited by severe contractures, it is unlikely he or she will ever walk at all. However, exceptions to above dictums are known and no child should be denied medical and rehabilitative treatment.

Conflicts of interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

REFERENCES

1. Bax M, Goldstein M, Rosenbaum P, et al. Proposed definition and classification of cerebral palsy. *Dev Med Child Neurol*. 2005;47:571–576.
2. Moster D, Wilcox AJ, Vollset SE, Markestad T, Lie RT. Cerebral palsy among term and postterm births. *JAMA*. 2010;304:976–982.
3. Stanley F, Blair E, Alberman E. *Cerebal Palsies: Epidemiology and Causal Pathways*. London MacKeith Press; 2000.
4. Vincer MJ, Allen AC, Joseph KS, Stinson Da, Scott H, Wood E. Increasing prevalence of cerebral palsy among very preterm infants: a population-based study. *Pediatrics*. 2006;118: 1621–1626.
5. Ancel PY, Livinec F, Larroque B, et al. Cerebral palsy among very preterm children in relation to gestational age and neonatal ultrasound abnormalities: the EPIPAGE cohort study. *Pediatrics*. 2006;117:828–835.
6. Dolk H, Pattenden S, Johnson A. Cerebral palsy, low birth weight and socio-economic deprivation: inequalities in a major cause of childhood disability. *Paediatr Perinat Epidemiol*. 2001;15:359–363.
7. Johnson A. Prevalence and characteristics of children with cerebral palsy in Europe. *Dev Med Child Neurol*. 2002;44: 633–640.
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;39:214–223.
9. Hoffer MM, Brink J. Orthopedic management of acquired cerebrospasticity in childhood. *Clin Orthop Relat Res*. 1975;110: 244–248.
10. Hoffer MM, Garrett A, Brink J, Perry J, Hale W, Nickel VL. The orthopaedic management of brain-injured children. *J Bone Joint Surg Am*. 1971;53:567–577.
11. Samilson RL. Current concepts of surgical management of deformities of the lower extremities in cerebral palsy. *Clin Orthop Relat Res*. 1981;158:99–107.
12. Sawyer JR. Cerebral palsy. In: Canale ST, Beaty JH, eds. *Campell's Operative Orthopaedics*. 11th ed. Philadelphia: Mosby Elsevier; 2008:1333–1399.