Review Article

Medical and Dental Implications of Cerebral Palsy. Part 1 General and Medical Characteristics: A Review

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Abstract

Cerebral palsy (CP) is a group of neuromuscular disorders that affects the development of movement and posture, causing activity limitations. CP is classified into three main groups: spastic, dyskinetic and ataxic. Population-based studies from around the world report estimates of CP prevalence ranging from 1.5 to more than 4 per 1,000 live births. The commonest cause of CP remains unknown in 50% of the cases; prematurity remains the common asterisk factor. CP children suffer from numerous problems and potential disabilities such as mental retardation, epilepsy, feeding difficulties, and ophthalmologic and hearing impairments. This paper reviews and critically discusses the definition, epidemiology, aetiology, classifications, treatments and associated manifestation and complications of CP.

INTRODUCTION

Cerebral palsy (CP) is a group of neuromuscular disorders, which affect the development of movement and posture, causing activity limitations. These limitations are attributable to non-progressive disturbances, which have occurred in the developing infant brain. Such disturbances include infection, hypoxia, trauma, and hyper bilirubin anaemia; biochemical and genetic factors may also be involved [1].

The motor disorders of CP are often accompanied by disturbances of sensation, cognition, communication, perception, and/or by seizure disorders [2].

CP may be classified into three main groups [3].

1) Spastic, characterized by increased muscle tone;
2) Dyskinetic, characterized by hypotonic, slow writhing movements (athetotic), abnormal postural control movements, swallowing difficulties, problems of speech and coordination; 3) Ataxic, characterized by involuntary movement, lack of balance and depth perception.
CP is the most common motor disability which occurs during childhood [4]. Population-based studies from around the world report estimates of CP prevalence ranging from 1.5 to more than 4 per 1,000 live births of children of a defined age range [5-9]. In the industrialized nations, the prevalence of cerebral palsy is approximately 2 per 1000 live births [10]. In the United States, approximately 10,000 infants and babies are diagnosed with CP each year, and a further 1200-1500 are diagnosed at preschool age [10].

The prevalence of CP in the developing world is not well established, but estimated to be 1.5-5.6 cases per 1000 live births [11].

**LITERATURE REVIEW**

**Definition**

Sir John Little was the first to define cerebral palsy in his famous work of 1862, though he did not apply that term at the time. William Osler, in his book “The cerebral palsy of children” (1889), defined the clinical features, which presented 151 CP children, grouping them according to their expected aetiology, in order to interpret the physiopathological mechanisms of the cerebral lesion (damage location). Sigmund Freud, in his “Die infantile Cerebralähmung” (1897) inspected the causes of these motor disorders, emphasizing the significance of pre-term birth and intrauterine development disorders, in contrast to Little [12].

The American Academy for Cerebral Palsy (AAPC) was founded in 1947 and is considered a multidisciplinary professional association, aimed at sponsoring research in the field of infant disability.

In 1992, Mutch et al. defined CP as “an umbrella term covering a number of syndromes with motor deficiency, non-progressive, but often changing, secondary to brain lesions or anomalies appearing in the early stages of brain development” [12].

Another definition was then introduced in 2007 by Rosenbaum et al. as follows: “Cerebral palsy (CP) describes a group of disorders of the development of movement and posture, causing activity limitation that are attributed to non-progressive disturbances that occurred in the developing foetus or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behaviour, and/or by a seizure disorder” [12].

**Prevalence**

Cerebral palsy (CP) is the most common motor disability occurring during childhood [4]. Population-based studies from around the world report prevalence estimates of CP ranging from 1.5 to more than 4 per 1,000 live births or children of a defined age range [5-9]. In the industrialized world, the prevalence of cerebral palsy is about 2 per 1000 live births [10]. In the United States, approximately 10,000 infants and babies are diagnosed with CP each year, and a further 1200-1500 are diagnosed at preschool age [10]. The prevalence of CP in the developing world is not well established, however, it is estimated to be 1.5-5.6 cases per 1000 live births [11]. In Saudi Arabia, a prevalence ratio of 5.3 in every 1000 individuals was reported among the Saudi population [13]. While there are no available studies which indicate the prevalence of the CP in the UAE, or Dubai in particular, experts in the field indicate that it is similar in proportion to that of Western populations [14].

**Aetiology**

**Congenital aetiology:** CP patients have distinctive deformities which result from defects occurring in normal development, and follow outlines based on failures of normal formation [15]. The earliest acknowledged deformities which lead to survival with motor defects are defects of the neural tube closure, such as Meningomyelocele (the most common neural tube defect occurring in the spine). On the other hand, the neural tube defects in the brain known as encephalocele may be anterior, with main mid face or nasal defects [16]. Children with significant encephaloceles have significant motor impairments, including quadriplegic patterns associated with hypotonia, rather than hypertonia. Segmental defects in the brain known as schizencephaly refer to a cleft in the brain; such schizencephalies vary greatly in their effects, from causing minimal disability to causing very severe quadriplegic pattern involvement, and usually present with spasticity and mental retardation [17].

Primary proliferation defects of the brain result in microencephaly. Regardless, there are numerous other causes of microencephaly, most of which relate to toxins or infection [15]. Megalencephaly is caused by cellular hyper-proliferation, commonly in syndromes such as sebaceous nerves syndrome, where as macrocephaly is most often due to hydrocephalus. Throughout development neurons transfer toward the periphery of the brain, a defect in this migration pattern leads to lissencephaly, which means “smooth brain”; in other words, a child with decreased cerebral gyri. This defect usually leads to severe spastic quadriplegic pattern involvement. There is a broad-spectrum understanding that significant seizure activity in a young child may preclude synaptic remodelling through excitotoxic injury, which may lead to CP.

**Neonatal aetiology:** Neonatal and prenatal causes of CP are chiefly linked to prematurity and birthing problems, which lead to numerous patterns [15]. The general trend is that premature infants with more severe bleeds in the brain (in ventricles and the periventricular white matter areas) have a worse prognosis for survival and are at a higher risk of developing CP; nonetheless, there are no restrictions which may accurately predict the risk of developing CP, much less predict the severity of CP in an individual child. In full-term infants, Hypoxic events occurring surrounding delivery usually lead to disability; these events have been called hypoxic-ischemic encephalopathies (HIE) [16].

Sub cortical cyst formations develop in severe cases of HIE and are called multi cysticencephalomalacia. When this cystic pattern forms, the prognosis for good function is poor, with most children developing severe quadriplegic pattern involvement, with severe mental retardation. Further, some of these children develop cysts in the thalamus and basal ganglia, which may lead to dystonia [16].

In the preterm or full-term infant, neonatal stroke usually affects the middle cerebral artery and presents as a wedge-shaped defect in one hemisphere. If such a wedge-shaped defect is small, the child may not be affected; similarly, a significant defect, especially with a cyst, usually presents as hemiplegic pattern.
CP. Despite a large cyst, these child's overall, and especially cognitive, functions may be quite good [15].

**Postnatal causes of cerebral palsy**

Postnatal causes of CP may overlap to some extent with the prenatal and neonatal group; however, metabolic encephalopathy, postnatal trauma, infections, and toxicities are considered as aetiologies in this group [18]. Child abuse, falls, or motor vehicle accidents can result in Blunt head trauma and skull fractures, which comprise not only the direct injury, but also the secondary injury from brain swelling. However, children with blunt trauma may recover and have no resultant motor defects [19]. Nevertheless, if there is a unilateral bleed in the brain, affected children are frequently left with hemiplegic pattern motor disabilities. The more severely affected children are frequently left with severe quadriplegic pattern involvement and often do not become functional community ambulators.

Many children with motor damages from closed head injuries suffer from ataxia as a major impairment. A wide variety of infections leave children with permanent neurologic deficits. Prenatal and neonatal viral infections are the most common infectious cause of CP. Cytomegalovirus (CMV) leaves 90% of children with mental retardation and deafness, while only 50% develop CP or motor defects. Children who develop congenital rubella infections will commonly suffer from mental retardation; however, only 15% develop CP [20]. Neonatal herpes simplex infection shave high mortality rates, while 30-60% of survivors have some neurologic sequelae; with regard to such infections, resultant CP is not common.

Infections with human immunodeficiency virus (HIV) may cause neurologic sequelae; affected children usually develop a progressive encephalopathy, and should be treated with anticipated short life expectancy. Neonatal bacterial meningitis may be caused by many organisms the effects of which may be very severe; as many as 30-50% of survivors suffer from CP [20].

Many toxic agents can cause temporary neurologic deficits, alcohol being the most commonly encountered. Children with prolonged anoxic events, such as near drowning, near hanging, or near asphyxiation can make outstanding recoveries. However, when these children do not make a complete recovery, they are usually left with exceedingly severe neurologic deficits.

**CLASSIFICATION**

**Classification based on severity level**

Cerebral palsy is frequently classified by severity levels mild, moderate, severe, or no CP [21].

1) Mild - A child is able to move without assistance; their daily activities are not restricted; 2) Moderate - A child will need medication, braces, and adaptive technology in order to achieve daily activities; 3) Severe - A child will need a wheelchair and will have substantial challenges in achieving daily activities and 4) No CP - The child has CP signs, but the damage was acquired after the completion of brain development, and is consequently classified under the incident that caused CP, for example: traumatic brain injury or encephalopathy.

**Classification based on topographical distribution**

Topographical classifications describe the affected parts of the body [3,21,22]. This is useful in establishing treatment protocols as follows: paresis (weakened), plegia/plegia (paralyzed), monoplegia/monoparesis (refers to when only one limb is affected) and Diplegia/diparesis (when the lower body is principally affected; legs are affected more frequently than arms). When one side of the body is affected (arm and leg) this is referred to as hemiplegia/hemiparesis; when the lower half of the body (both legs) are affected it is called Paraplegia/paraparesis. When three limbs are affected this condition known as Triplegia/triparesis. Lastly, when all four limbs are involved it is classified as quadriplegia/quadriparesis.

**Classification based on motor function**

CP is caused by brain injuries which affect motor function, as well as the ability to control the body in a normal manner [3,21,23]. There are two main groups, spastic and non-spastic. Each group has multiple characteristics and it is possible to have a mixture of both types. Spastic cerebral palsy is characterized by increased muscle tone while non-spastic cerebral palsy exhibits decreased or fluctuating muscle tone.

**Pyramidal or spastic cerebral palsy**

Pyramidal CP means that the pyramidal tract is affected by upper motor neuron damage, and is either not functioning properly or is damaged. Spastic CP is hypertonic and accounts for 70% to 80% of cases. It creates stress on the body, which result in interconnected conditions such as scoliosis, hip dislocation, limb deformities, and contracture (painful joint deformities) [3,21].

**Extrapyramidal or non-spastic cerebral palsy**

Non-spastic CP manifests through decreased or fluctuating muscle tone, and is characterized by involuntary movement, which is worsened by stress and eliminated by sleep. Non-spastic CP has multiple types, which are characterized by their specific damages. Non-spastic cerebral palsy is divided into ataxic and dyskinetic groups, which make up 20% of CP cases (with dyskinetic comprising 15%, and ataxic 5%) [3,21].

Ataxic CP affects coordinated movements that involve balance, posture; walking gait is frequently very wide and sometimes irregular. In addition, fine motor skills requiring coordination of the eyes and hands, such as writing, are difficult. Dyskinetic CP is divided into two different groups: athetoid CP - which consists of cases with involuntary movements, principally in the arms, legs, and hands and dystonia/dystonic CP - which includes cases affecting trunk muscles more than limbs and results in a fixed, twisted posture.

**MIXED CEREBRAL PALSY**

Mixed CP refers to impairments which fall into both spastic and non-spastic categories. 60% of mixed CP cases have their age of onset in an individual’s thirties or later [3,21].

**Classification based on the gross motor function classification system (gmfs)**

GMFS is composed of a five-level system that is related to the individual’s extent of ability, and the limitations of their impairment(s) [24]. A higher number signifies a higher degree of severity. Each level is determined by age range, and a set of activities the child may achieve on their own.
GMFCS classification levels:
GMFCS Level I – “walks without limitations.”
GMFCS Level II – “walks with limitations.”
GMFCS Level III – “walks with adaptive equipment assistance.”
GMFCS Level IV – “self-mobility with use of powered mobility assistance.”
GMFCS Level V – “severe head and trunk control limitations.”

DIAGNOSIS
CP is not frequently diagnosed at birth, with most children diagnosed between the ages of six months and 2 years. Infants with severe CP may have recognizable signs at birth, such as abnormal muscle tone. In addition, delayed development is usually the first sign observed in a child with CP. These infants may have abnormal muscle tone or some abnormal movements [25]. Children with normal development will usually be able to sit unaided at 6 months of age, crawl by 8 months, pull to stand by 12 months, and walk by 15 months [25].

There is some variety, and some children meet these developmental milestones later. Nevertheless, a child who is late in meeting these milestones should generally be evaluated for CP. Diagnosis can be made by a child specialist evaluating the symptoms, and signs of delay in development [25].

ESTS AND SCANS
To rule out other problems with similar symptoms to CP, such as muscular dystrophy (a group of inherited conditions that gradually weaken the muscles), additional tests may be suggested [26].

Further testing – including Magnetic Resonance Imaging (MRI) scan, Ultrasound Scan, Computerized Tomography (CT) scan, Electroencephalogram (EEG), Electromyogram (EMG), and blood tests – may be able to confirm cerebral palsy diagnoses, as the condition can cause changes to the brain’s structure [26].

TREATMENT
The treatment for children with CP may include physical therapy, occupational therapy, speech therapy, medication, surgery, communication aids and assistive technology, vision and hearing aids, orthotic devices, and the use of other equipment [27].

PHYSICAL THERAPY
For CP children, physical therapy is a broadly used intervention, as it is aimed at encouraging motor skills, in addition to developmental skills and functional independence [28,29]. Physical therapy includes exercise, muscle training, as well as the use of orthotics or braces and other equipment [30,31].

SPEECH THERAPY
Many children with CP face problems with drooling and dysarthria (difficulty in articulating words caused by impairment of the speech muscles). Therefore, speech therapy for those children may help with issues related to speech, along with related feeding and swallowing problems [32]. Furthermore, speech therapists may make use of augmentative communication devices or sign language as instructional devices to provide additional communication services.

DEVICES AND GADGETS
Assistive technology, consisting of grab bars, magnifiers, rails, Velcro grips for eating utensils and writing implements, along with voice communication devices, computer software programs, customized wheelchairs, and positioning equipment (for the correction of posture), are devices for treatments of symptoms associated with CP [33-35].

SURGICAL AND MEDICAL INTERVENTIONS
The most common surgical procedures for patients with CP are performed for the treatment of scoliosis, hip dislocation, and severe contractures, along with deformities in tendons, bones and joints. Tendon lengthening, or transfer, and osteomy to realign a limb, are also common procedures. Further, intraoral insertion of a baclofen pump has been shown to be suitable to decrease spasticity in the lower extremities and trunk. In this procedure, an intra spinal catheter is placed and connected to a reservoir under the skin of the abdomen [27]. Another relatively common surgical procedure is dorsal rhizotomy, “a procedure in which the surgeon cuts a portion of the spinal sensory roots that provides input to spastic leg muscles.”

GENERAL CLINICAL MANIFESTATIONS AND COMPLICATIONS
Mental retardation
Not all children with CP are cognitively impaired. Indeed, the most common type of CP (spastic diplegic) is characterized by normal cognition, as the lesion is in the peri ventricular white matter. Nevertheless, there is some relationship between the severity of CP and mental retardation, as in the case of spastic quadriplegic CP patients. In addition to other factors which increase cognitive impairment (such as epilepsy, and cortical abnormalities on neuro imaging) individuals with spastic quadriplegic CP have a greater degree of mental retardation than those with spastic hemiplegia [36,37].

Epilepsy
Epilepsy can be a sign of the severity of neurological injury (quadriplegic CP) or cortical insult (hemiplegic CP). Up to 36% of children with CP have epilepsy, with 70% of cases presenting with onset in the first year of life [38]. However, Children with spastic diplegic CP are at a lower risk for epilepsy, as their pathology predominantly involves the periventricular white matter. Several new antiepileptic drugs have enhanced the ability to control the seizures in these children [39].

Feeding, nutrition, and growth
In children with severe CP, the most common concerns relate to feeding, nutrition, and growth. About 30% of CP children are undernourished below the third percentile. The leading cause for growth delay appears to be poor nutrition secondary to pseudo bulbar palsy, an upper motor neuron disorder resulting in poor coordination in sucking, chewing, and swallowing [40].
In addition, gastroesophageal (GE) reflux results in regurgitation, vomiting, and possible infections of the difficult-to-feed child. Early nasogastric (NG) or gastrostomy tube (GT) feedings can resolve these problems, with improved growth and greater family satisfaction [40]. NG tube feeding can be used for short-term nutritional support, though this method is generally not acceptable as it can be associated with nasal discomfort, sinuses, and irritation of the larynx, as well as recurrent tube blockage or displacement on a long-term basis. Surgically or endoscopically placed GT can afford long-term solution to feeding disorders, in combination with treating the associated GE reflux [40].

Recently Penagini et al. [41], reported that feeding problems are frequently secondary to oropharyngeal dysphagia, which usually correlates with the severity of motor impairment and presents in around 90% of preschool children with cerebral palsy (CP) during the first year of life. Early identification and intervention of nutritional issues of NI children with a multidisciplinary approach is crucial to improve the overall health and quality of life of these complex children [41].

**Bladder dysfunction**

Children with CP are at increased risk for urinary incontinence, urgency, and infections [42]. It has been reported that up to 23% of these children had Primary incontinence, which shows a relationship between lower cognition and severe motor deficits [43]. However, the attainment of continence can be influenced by communication skills, along with physical ability to get to the bathroom and manage clothing. Spastic CP can be associated with spasticity of the destructor muscles, resulting in small frequent voids, and a low capacity irritable bladder. In addition, adapted toilet seats, handrails, and clothing modifications can improve the number of toileting successes.

**Bowel dysfunction**

In children with CP, Constipation is common and results from various factors including immobility, poor feeding, and reduced water intake. For Initiating bowel evacuation, a combination of laxatives and suppositories are recommended. Further, softening agents with dietary modifications can give rise to more regular and softer bowel movements [36].

**Sleep disturbances**

In CP children Sleep disorders are common and occur in up to 50% of cases, namely in those with visual impairment [44]. The manifestations of these disorders include disturbed sleep patterns, fragmented sleep, and frequent nocturnal awakenings, which can be extremely distressing to parents. Medications which improve the sleep-wake cycle may also decrease spasticity and improve daytime behaviour [44,45]. Hypnotics are generally effective for short periods, but tolerance against them is built in the span of a few days.

**Hearing loss**

Hearing loss, if not diagnosed and treated early, can hinder developmental progress and rehabilitation in children with CP, thereby contributing to further developmental delays [46]. Certain aetiologies can increase the risk for hearing loss such as congenital rubella, kernicterus and post-meningitis. Screening and hearing assessment is recommended routinely for any child with developmental delay, including children with CP. These screenings may include behavioural audiometry, auditory-evoked brainstem responses (ABR), or transient evoked oto acoustic emissions [46].

**Visual abnormalities**

Children with CP are at an increased risk for visual impairment including retinopathy, amblyopia. Strabismus, glaucoma, and myopia, which can interfere with developmental progress and rehabilitation if not diagnosed and managed early on. Accordingly, screening and serial ophthalmologic assessments are recommended routinely for any child with global developmental delays (such as CP), principally if there is suspected vision loss [47].

**Orthopaedics abnormalities**

Developing bones grow in the direction of the forces placed upon them, thus, progressive joint contractures, shortened muscles, and hip or foot deformities can arise from spasticity. Other orthopaedic complications, which must be observed, include scoliosis and fractures due to osteoporosis or osteomalacia. These symptoms are more common with severe motor disability and immobility, such as quadriplegia [36].

**CONCLUSION**

Children with CP suffer from multiple problems and potential disabilities such as mental retardation, epilepsy, feeding difficulties, vision, and hearing impairments screening for these conditions should be part of the initial assessment. The child with CP is best cared for with an individualized treatment plan that provides a combination of interventions. Management if provided optimally can improve the quality of life of these children and their families.

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**REFERENCES**


