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MODULE III. 8

Cerebral palsy

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I. Introduction

PCI encompasses a heterogeneous group of syndromes that show persistent motor dysfunction affecting muscle tone, movement, and posture, due to injury to a developing brain. Although by definition it is a non-progressive disorder, its clinical expression varies with the age of the child and the appearance of various comorbidities that can condition their quality of life even more than neurological disorders (Bax et al, 2003).

PCI is attributed to disorders that occurred in the developing brain and whose diagnosis is established during the first 4-5 years of life. Therefore, it is the result of a brain alteration whose origin is multifactorial (Peláez-Cantero et al, 2021). **PCI can occur both in the prenatal, perinatal and postnatal stages** , highlighting the presence of hypoxic-ischemic lesions, intraventricular and periventricular hemorrhage, early defects in neuronal migration, cerebrovascular malformations and infections of the central nervous system. (Carrillo et al, 2018).

The presence of any of these alterations causes a disorder in tone and muscle coordination, generating an alteration of movement that can also affect speech. PCI is a heterogeneous picture, whose severity can vary widely, conditioning different clinical manifestations.

According to the predominant motor disorder, these are clinically classified into spastic, *dyskinetic and*, ataxic, PCI The most frequent cerebral palsy is the spastic hemiplegic form. PCI is not only characterized by the presence of motor alterations, it can also be accompanied by perceptual, cognitive, communication and behavioral disorders. Likewise, epilepsy and secondary musculoskeletal problems are common, encompassing all these disorders within the current definition of PCI (Guiu Antem et al, 2017., Pascual, 2011).

PCI is usually associated with periventricular **leukomalacia with intraventricular or periventricular** hemorrhage, and in fewer cases with **micropolygyria and porencephaly (**Carrillo de Albornoz et al, 2018).

II. Objectives

Know the main causes of cerebral palsy. Clinical and topographic classification based on brain damage.

Know the main disorders related to cerebral palsy, as well as their main characteristics.

III. Specific contents of the topic

3.1. Cerebral palsy (PCI)

Cerebral palsy is a clinical picture whose diagnosis requires the presence of **alterations in the development of posture, movement, and muscle tone**. The clinical picture is characterized by a disorder of movement and posture that causes an alteration in the child's ability to make voluntary use of his muscles.

Cerebral palsy is caused by an abnormality or disruption in brain development. The problem in brain development could arise at different times of development such as:

First trimester of gestation: Malformations, proliferation, cell migration, synaptic organization.

Second and third trimester of gestation: Brain injury. Prematurity

Brain injury of the newborn: injury of the premature child, white matter lesion (leukomalacia) Intraventricular hemorrhage, hemorrhagic infarction, cerebellar lesion. hypoxic-ischemic encephalopathy, metabolic disease.

Postnatal period: Trauma, infections.

With respect to the typology of PCI, the most frequent are *spastic PCI* where the lesion occurs in the **motor cortex or pyramidal bundle.** Increased muscle tone of greater or lesser intensity is observed, with decreased voluntary movement.

There is a predominance of flexor or extensor muscle groups that lead to shortening and deformities. (Carrillo de Albornoz et al, 2018). This type of paralysis can occur unilateral or bilateral, where muscle tone in both upper and lower limbs are increased.

On the other hand, *dyskinetic or dystonic PCI* arises as a consequence of injury to the basal ganglia or extrapyramidal bundle. Involuntary movements and sudden changes in tone appear. Download the presence hypokinesia and hypertonia being the type of dystonic paralysis and where hyperkinesia and hypotonia predominate is considered choreo-atheotosisal or dyskinetic paralysis.

With respect to *ataxic PCI,* it is caused by the lesion of the cerebellum. Characterized by the presence of hypotonia, incoordination and impaired balance.

If we consider the anatomical distribution of the motor disorder, CP is classified into diplegia (alteration of the movement of the four limbs, but with greater involvement of the lower limbs), *hemiplegia* (involvement of the upper and lower limbs of the same side) and *quadriplegia* (involvement of the four limbs).

Table 1. Classification of cerebral palsy. (Based on Carrillo et al, 2018).

Causes during pregnancy:
Prenatal: disorders of cortical development, intrauterine infections, toxic. **Perinatal**: hypoxia, prematurity, jaundice. **Postnatal:** infections, poisoning, accidents.
Compromised brain structure
Pyramidal pathway: spastic cerebral palsy. **Extrapyramidal pathway:** pdyskinetic cerebral palsy. Cerebellum: ataxic cerebral palsy.
The extent of affectation (topographic classification) Unilateral/Bilateral
Monoplegia (involvement of only one limb). **Hemiplegia:** The effect of the upper and lower extremity of a hemibody. **Diplegia:** greater involvement of the lower extremities than the upper extremities. **Tetraparesis:** involvement of all four limbs. **Tryparesia**: involvement of the lower extremities and a single upper limb.
According to the severity of the affectation
Functional classification (motor, manual, language, and functionality level) **Grade 0**: normal. **Grade 1:** slight anomalies with the possibility of voluntary correction. **Grade II:** obvious abnormalities that do not impede function. **Grade III:** limited function (slowness, tiredness and need for support). **Grade IV**: impossible function (no gear, no manual function or language)
Gross motor function classification system (GMFCS)
Level I: unrestricted march **Level II:** unaided walking, but with spatial constraints **Level III**: gait with support or orthosis **Level IV**: fairly limited independent skills. **Level V**: tally dependent for displacement.

3.2. Associated problems in cerebral palsy

Children with cerebral palsy, as indicated by authors such as Peláez-Cantero et al, 2021, require in most cases a multidisciplinary approach to treat the problems associated with this motor pathology, so it is common to find associated problems described below.

***Neurological problems*:** Epilepsy is present more frequently in those children who show pathological findings in neuroimaging tests and present greater motor alteration. Performing an EEG can be useful to establish neurophysiological parameters compatible with the presence of seizures of epileptic origin. (Peláez-Cantero et al, 2021).

***Intellectual disability**:* Between 40-70% of children with spastic and quadriplegic paralysis have intellectual disability and to a lesser extent with dyskinetic and hemiplegic paralysis. It is also associated with the presence of epilepsy and pathological neuroimaging study.

Language disorders: The language disorders most present in PCI are dysarthria (40%) followed by 25% showing absence of verbal language. They may also present difficulties in other areas of communication, such as the development of gestures and facial expression, acquisition of comprehensive and expressive language and the production of voice.

***Hearing problems**:* In children with CP, neonatal screening should consist of otoemissions and auditory evoked potentials, warning signs can range from poor response to auditory stimuli, abnormal behavioral responses, and impaired language development.

Sialorrhea: It is found in 10% and 58% of children with PCI so it is important to quantify frequency, severity, and impact on the quality of life of children and their caregivers.

***Neuropsychiatric problems**:* Present in more than 50% of children with CP, being the most frequent, emotional disorders, behavioral problems and social interaction, presence of hyperactivity and attention deficit, which added to all the problems present aggravated the school and adaptive problems of these children.

Spasticity: Appears in 85% of children with CP and causes functional problems in ADLs (gait, feeding, clothing and hygiene). This alteration usually causes muscle pain, spasms, and dystonic postures.

Orthopedic problems: Caused largely by spasticity, which causes fixed muscle contractures that cause osteoarticular deformities (thumb included, wrist and elbow flexo, scoliosis, hip displacement / dislocation, clubfoot, which worsen the clinical situation of the child needing in certain cases, a surgical approach.

Digestive problems: Present in 80-90% of cases related to nutrition and growth and the presence of dysphagia, gastrointestinal reflux, and constipation.

Bone health problems: These children are at risk for low bone density and osteoporosis. Pathological fractures may be present in up to 20% of children with CP and most often affect the distal femur. Their recognition is important since in most cases they occur asymptomatically (80% of vertebral fractures).

Oral health problems: Children with CP more often have cavities, malocclusions, and periodontal disease (90%)

Respiratory problems: *One of* the main causes of morbidity and mortality in children affected with CP. Respiratory symptoms vary with the age of the child, with infants with feeding difficulties, aspiration or apparent life-threatening episode, persistent cough, noisy breathing and repeated respiratory infections being more frequent. There is also a risk of sleep apnea-hypopnea syndrome.

Visual problems: Between 40-75% of children have some type of visual disability, nystagmus, absence of reflex visual response, endless eye movements and lack of attention and visual curiosity may also appear.

Urological problems: 60% of these children have voiding dysfunction, enuresis, voiding urgency, incontinence, or neurogenic bladder. The warning signs are continuous drip incontinence, need for abdominal pressure for the onset of urination or weak voiding stream or polydipsia.

Sleep problems: The presence of sleep disorders is present in 25% of children with CP. The main sleep disorders in these children are difficulty initiating and maintaining nighttime sleep, difficult morning awakening, nightmares, and sleep anxiety.

Pain: Frequent symptom in PCI, the most frequent mechanisms that generate pain include both nocioceptive pain: somatic (spasticity, hip subluxation, fracture, etc.) and visceral (constipation, GER, gastric ulcer) neuropathic pain, and pain secondary to treatments: physiotherapy, infiltration of botulinum toxin.

3.3. Assessment of the functional ability of the child with cerebral palsy

Given all the alterations, it is clear that cerebral palsy is not just a motor disorder, but the sum of many alterations presents to a greater or lesser extent. To assess the functional capacity and degree of dependence of the child with CP, there are currently five scales that evaluate, motor aspects, manual function, communicative, feeding, and visual aspects.

With respect to the GMFCS (*Gross motor function Classification System*) it is the most widely used system currently to classify motor gravity. It establishes five levels of severity and allows to assess the natural history of PCI, which is different in the different levels of involvement and also to assess the usefulness of treatments (Palisano et al, 1997) (Table 1)

Regarding the *assessment of manual function,* evaluated by the Manual Skill Classification System (MACS) describes how children with cerebral palsy (CP) use their hands to manipulate objects in daily activities. This system describes five levels.

The levels are based on the child's ability to self-initiate the ability to manipulate objects and his need for assistance or adaptation to perform manual activities in daily life. (Eliasson et al, 2006).

The classification proposed by the CFCS is to *assess the child's performance in daily communication.* This classification focuses on levels of activity and participation, as described in the World Health Organization (WHO) International Classification of Functioning Disability and Health (ICF). It establishes five levels of communicative effectiveness. CFCS is analogous to and complementary to the Gross Motor Function Classification System (GMFCS-ER), the Manual Skill Classification System (MACS), and the Eating and Drinking Ability Classification System (EDACS).

With respect *to the* Eating and Drinking Ability Classification System. (EDACS) (Sellers et al, 2013). It aims to classify and describe the ways in which people with CP eat and drink. It proposes five levels of skills that evaluate the functional activities of eating, such as sucking, biting, chewing, as well as the adaptation of food consistencies, feeding pathway and level of independence.

The difference between the levels is established based on safety and efficiency at the time of eating. Safety is understood as the risk of choking and bronchoaspiration that is associated with eating and drinking. While efficiency refers to the time and effort required to feed. It also presents a decision algorithm as a graphic tool to determine the level at which the child with cerebral palsy is.

And, finally, the classification system of visual functions (VFCS) (Baranello et al, 2020) that allows to classify in five levels the visual abilities of these children and how these capacities are used by these children in daily life.

3.4. Multidisciplinary approach in the treatment of cerebral palsy

As seen throughout this chapter, children with CP present a state of fragility where there are periods of symptomatic stability, the disease itself makes these children more likely to present episodes of decompensation due to intercurrent processes that can worsen their baseline situation. Therefore, at certain times and throughout their development, the child with CP will require a multidisciplinary treatment that helps them recover their previous baseline situation and contribute to improving their quality of life and in which, in addition to the professionals, the family and the environment of the child with cerebral palsy must be included. (Peláez-Cantero et al, 2021).

Summary

Cerebral palsy (CPI) is one of the most common major disabilities in child development. It causes significant physical disability in childhood. The clinical manifestations of this disease will depend on the extent and location of the brain injury, as well as the ability of the brain to adapt to it. PCI is classified according to the clinical manifestations of the motor disorder, the brain structures involved, the period in which the injuries occur, and the severity of the type of PCI.

The approach of these children often requires attention by a multidisciplinary team since these children have complex medical care needs.

Glossary

Athetosis: Lesions originating in the extrapyramidal system manifested in slow, involuntary, uncontrolled, and objectless movements.

Ballism: Involuntary, very abrupt, and wide movements that occur while the person is conscious and are caused by an injury to the subthalamic nucleus of the brain or its connections**.**

Korea: Involuntary movements of the limbs, trunk, neck, or face. They are fast, abrupt, arrhythmic movements that pass from one body region to another irregularly.

Clonus: involuntary, rhythmic contractions that occur in a muscle group when a sudden and passive extension of the tendons is performed in a sustained manner**.**

Dystonia: A movement disorder that causes involuntary contractions of the muscles. These contractions result in twisting and repetitive motions. Sometimes they are painful. Dystonia can affect only one muscle, one muscle group, or all muscles.

Spasticity: Increased muscle tone in lower limbs that affects mobility and causes serious complications: pain, joint limitation, contractures, and pressure ulcers, which lead to a significant impairment of the individual's functionality and quality of life.

Pyramidal bundle: set of nerve fibers that allow the transfer of orders from the brain to the nerve cells contained in the spinal cord

Ventricular leukomalacia: Damage or softening of the white matter that transmits information between nerve cells and the spinal cord, as well as from one part of the brain to another. Damage that is located around or near the ventricles that contain CSF.

Micropolygyria: Decrease in the size of cerebral grooves and convolutions with an increase in their number.

Porencephaly: A congenital neural tube closure defect extending from the surface of the cerebral hemisphere to the underlying ventricle, including uni- or bilateral cystic cavities resulting from vascular lesions. related to intrauterine vascular accidents (trauma), neonatal hypoxia, and hypertensive disease of pregnancy.

Sialorrhea: oral disorder characterized by excessive accumulation of saliva. This abundant segregation generates an involuntary loss of saliva, causing difficulty in controlling oral secretions**.**

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Web

Spanish Confederation of Federations and Associations of Care for People with Cerebral Palsy and Related (ASPACE Confederation) http://www.aspace.org/

Eating and Drinking Ability Classification System. (EDACS) (https://www.sussexcommunity.nhs.uk).

NIPACE Foundation for children with cerebral palsy. https://www.fundacionnipace.org/

Gross motor function classification System: https://www.abclawcenters.com/cerebral-palsy/gross-motor-function-