Cerebral palsy: classification and etiology

Beyin felci: Sınıflama ve etyoloji

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Cerebral palsy (CP), a common condition of abnormalities in the brain, arises early in life. Since the term was first introduced in 1843, many authors have tried to define and classify CP. The most recent definition was released by the American Academy for Cerebral Palsy and Developmental Medicine (AACPDM) in 2005. This article summarizes the latest and familiar classifications of, and etiologies associated with CP.

Key words: Cerebral palsy/classification; child; motor skills disorders; severity of illness index.

Definition
The first definition of cerebral palsy (CP) was described by Bax in 1964 as “a disorder of posture and movement due to a defect or lesion of the immature brain.”[1] In 2005, a committee of the American Academy for Cerebral Palsy and Developmental Medicine (AACPDM), led by Peter Rosenbaum, defined CP as “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 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 by a seizure disorder.”

They emphasized that motor dysfunction resulting from a progressive brain disorder or neurological disabilities that does not affect movement and postures cannot be considered as CP.[2] This definition has been widely accepted among health professionals all over the world, and provides a good basis for future research and clinical work.

The incidence of CP is considered to be 2 to 2.5/1000 live births.[3] The prevalence of CP in the developing countries tends to be in a similar range.[4-6]

Classification
Most known classifications are based on anatomic or topographic considerations, and on movement abnormality. Rosenbaum et al.[6] proposed a classification based on several components at the international working group in 2005, including (i) motor abnormalities, (ii) accompanying impairments, (iii) anatomical and neuro-imaging findings, (iv) cause and timing of the disorder.

(i) Motor abnormalities are assessed based on the following:

a) Nature and typology of the motor disorder such as spasticity, dyskinesia, and ataxia. Spasticity, the most common type of motor dysfunction, re-
fers to a velocity-dependent increase in the muscle tone (resistance to stretch).[7] Spasticity is usually associated with involvement of pyramidal tracts, the basal ganglia, or reticular formation, with upper motor neuron signs. Dyskinesia, an extra pyramidal involvement, may be either dystonic which includes hypertonia and reduced activity, or associated with choreoathetosis which includes irregular spasmatic, involuntary movements of the limbs or facial muscles. Ataxia refers to loss of orderly muscular coordination,[8] usually caused by a cerebellar deficit.

b) Functional motor abilities. This aspect of CP should be assessed using objective functional scales, and includes the extremities and oromotor function. The most commonly used system is the one developed by Palisano et al.,[9] the Gross Motor Function Classification System (GMFCS), based on disability and functional limitation. This system is used by many clinicians in adjacent to the above-mentioned classification. The GMFCS define five levels of function for four age groups, before 2 years, 2-4 years, 4-6 years, 6-12 years, with recent addition of 12-18 years. There is a strong correlation between this classification and the World Health Organization International Classification of Impairments, Disabilities and Handicaps,[10] and is also easy to use. Another less known classification is the Bimanual Fine Motor Function Classification for the upper extremity in CP.[11] This classification corresponds to the GMFCS. It includes five levels, where in level 1 both hands function with no limitation in fine motor skills, and in level 5, both hands have only ability to hold or worse. In the levels between these two, the hands have varying degrees of limitation in fine motor skills.

(ii) Accompanying impairments include the presence of epilepsy, and decreased IQ, hearing, and vision.

(iii) Anatomical and neuro-imaging findings.

a) The anatomic distribution is based on the involvement of limbs, trunk, and oropharynx. Commonly used terms are quadriplegia, diplegia, and hemiplegia. Quadriplegia is the most severe form. It involves all four limbs with the trunk, and the upper extremities are involved more than the lower extremities. The spectrum of severity is wide, from no sitting ability or head control, to being able to walk independently. This type of CP is commonly associated with hypoxia, and cystic degeneration of the brain.[12] Most children have pseudobulbar signs with swallowing and drooling problems. Mental retardation is common in many children.[13]

Diplegia is the most common anatomical type, in which the lower limbs are more severely affected than the upper limbs, and is associated with prematurity and low birth rate. Often there is periventricular leukomalacia.[14] Gait problems and seizures are common, nistagmus and strabismus may be present.

Hemiplegia is defined as involvement of ipsilateral upper and lower limbs, with the upper limb more severely affected than the lower limb, hand function being most affected. A focal lesion is likely to be the cause of hemiplegia. Seizure disorders are most common in this type of CP probably because of the focal brain lesion.[15] Sensory abnormalities on the involved side are common.[16]

Monoplegia and triplegia are relatively uncommon.

The authors recommend that the anatomical terms mentioned above, be abandoned until a more precise terminology is available, due to the large range of possibilities included in each term.[17]

b) Neuro-imaging findings. At the present time, there is insufficient information to recommend a classification for neuro-imaging findings.

(iv) Cause and timing. Cerebral palsy is associated with multiple risk factors, and in many cases, no identifiable cause can be found. We believe that it is not practical at this time to classify CP by causes.

Timing should only be used if firm evidence is present showing that CP was caused during a specific time-window such as a documented brain hemorrhage during delivery.

Etiology

Most patients with CP have no known cause for the disorder, but multiple risk factors can be found. However, in over 30% of the patients, no risk factor could be identified.[18]

The injury of the developing brain can be prenatal, perinatal and postnatal.
A history of prenatal cause is found in 75% to 80% of the patients. Only 10% to 15% are associated with hypoxia or birth trauma. Sixty percent of the children affected by CP are born at full term, and thus, prematurity is not the only cause for CP; nevertheless, low birth weight (less than 1500 g) and prematurity are well-known risk factors for CP. Other prenatal risk factors include infection and maternal drug or alcohol abuse, maternal epilepsy, mental retardation, hyperthyroidism, severe toxemia, and third trimester bleeding. Chorioamnionitis was found to be a risk factor for CP, in as many as 28% of premature infants. Cystic periventricular leukomalacia, a congenital brain malformation, may play a causative role.

Perinatal risk factors include multiple pregnancies, with significantly increased risk for CP. Twin pregnancies result in a child with CP about 12 times more than a single pregnancy, probably related to a low birth rate.

Brain hemorrhage during delivery, other types of birth trauma, kernicterus, vaginal bleeding on admission, placental complications, hypoxia, and anoxia were all associated with increased rate of CP.

Postnatal causes include head trauma, meningitis, encephalitis, and brain infarcts.

Genetic causes that are known to be a risk factor for CP involve a gene on chromosome 19.

Conclusion

Reviewing the literature, one can see that there is still much to be discovered about the impairments and neurological findings of CP. With more advanced technology and better understanding of the nature of CP, this might be possible in the future.

References
