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Cerebral palsy in prominence with ataxic cerebral palsy-A review

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Abstract
Motor function impairment, found in children affecting the movement, cognitive and sensory functions, refers to the cerebral palsy(1).This is a neuro developmental condition that severely affects the child's development.(1). The impairments have early onset starting from feeding, irritability and sleep patterns. (1). the predominant form of abnormality is the spasticity; other forms include dyskinesia and ataxic cerebral palsy. The diagnosis is often based on the observations and the parent reports and less comparison to the laboratory reports and neuro imagings. (2). in most of the cases, the cerebral palsy is followed by muscoskeletal dysfunction, epilepsy, sensational disturbances, perceptions, communication, cognitive and behavioural disturbances. (2).

Keywords: cerebral palsy, ataxic cerebral palsy, neuro developmental, spasticity, cognitive, dyskinesia

Introduction

Cerebral palsy, physical impairment affecting the movement, or a common physical disability rightly found in children [1].
Cerebral palsy, being the most prevalent cause for motor function impairment, include dyskinetic (dystonia & chareoathetosis) and ataxic cerebral palsy [2]. Cerebral palsies, severely affects the Childs development and is a neuro developmental condition [2].
In case of ataxic cerebral palsy, children are found to be of normal birth weight with prenatal origin of condition [3].
Being an heterogenic group, it confines to permanent abnormality in movement and maintaining posture [4].
The motor abnormalities associated with cerebral palsy include disturbances in sensation, cognition, perception, communication, epilepsy, behaviour and also mainly associated to muscosekeletal problems [4]
Here, ataxia also refers to the lack of movement, coordination, which is associated with the dysfunction of the brain part, cerebellum. This mostly affects children, since, they are still at a stage of learning and developing motor coordination [5]
Along with the motor coordination, the patients also exhibit sensory and cognitive impairments and nutritional deficiencies too in case of cerebral palsy.
Accordingly, abnormalities in dorsal prefrontal cortex, somato sensory cortex, dorsal anterior ingulate Grus and cerebellum is also observed in the neuroimaging studies [6].
The advances in the research is leading a way for opportunities in primary prevention and in specific intervention strategies [1].

Epidemiology

From around the world, the population based studies estimates the prevalence of cerebral palsy ranges from 1.5 – 4 in 1000 live births or children of specific age (10-14) [6].
The datas represented majorly by WHO on childhood ataxia is from Europe and it accounted to 52% of the studies done [5].
In the study done in 962 family members with 36 patients, 8% were found with neuro developmental disorder and 3% population with congenital malformation [7]
The prevalence in Western Australia gives around 3-2% to 4-2% births with major congenital malformation [4]
And in another population based study done in Al kharga District of Egypt, it was found like, out of a geographic population of 62583, the identified causes of cerebellar ataxia were found to be 17 of which 8 were vascular, 6 were subtype ataxic cerebral palsy and 3 were post encephalitic ataxica.
The prevalence rate for ataxia here is 27.16 in 100000 (95% confidence interval (CI), 14.3 – 40.1) [8]
In another study conducted in Winnipeg children hospital in 1991-2008, 184 patients were identified with ataxia. The incidence rate here was found to be 5.77 in 10000 and the prevalence rate was 6.59 in 100000 and mortality rate was 0.446 in 10000. [9]
The prevalence of cerebral palsy in Turkish children (2-16yrs) was like 186 out of 41861 children selected were identified with cerebral palsy and the prevalence was determined to be 4.4 in 1000 live births. [10]

Pathophysiology

Patients with cerebral palsy, that may be congenital or acquired, have a damage to the brain (cerebellum) in the early childhood [11]. The cerebellum gives the motor commands that is initiated by the descending pathways in the brain, giving accurate and adaptive movements [12]. Th case of older children. There is a difficulty in understanding congenital ataxic syndromes and the heredodegenerative ataxias. In most cases, congenital ataxias have a prenatal origin that is mostly genetic, with acquired haemorrhagic cerebellar lesions [13].

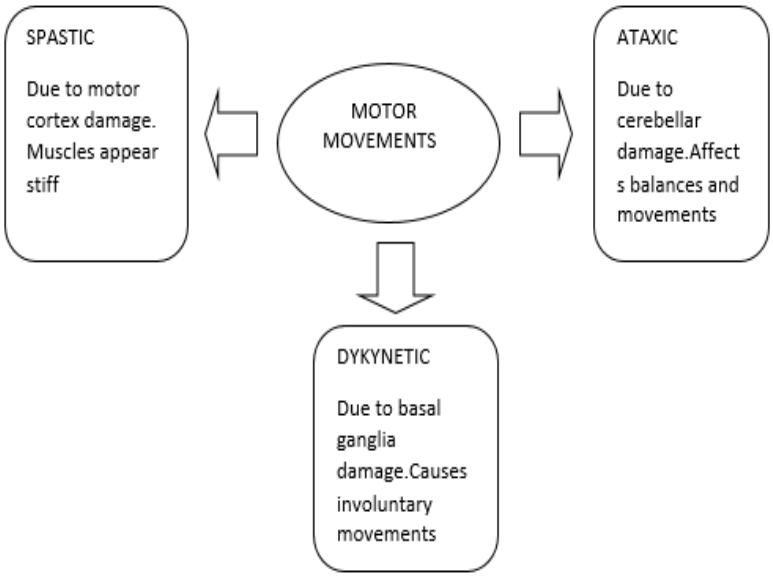

Fig 1

The mechanisms leading to cerebral palsy include:

1. Intrauterine exposure that leads to infection that evokes a fetal inflammatory response syndrome and causes fetal white matter damage, growth restriction in fetus, placental vascular disorders that provokes vascular injuries in the fetal brain and congenital anomalies.
2. Intrapartum events that effects the fetus during the labour and delivery which include chorioamnionitis, birth asphyxia, placental abruption which are associated with implication on the fetal brain.
3. Postpartum exposure leads to prematurity complications leading to cerebral palsy [7].

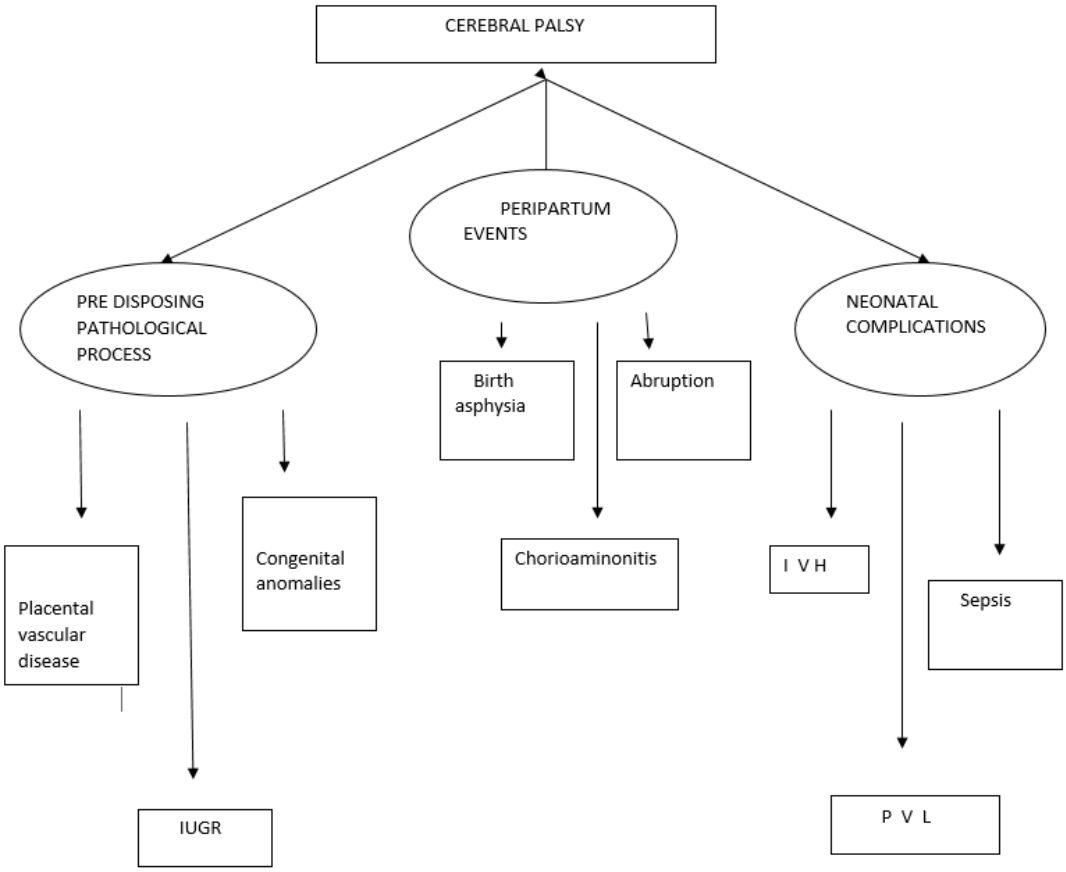

Fig 2

Clinical features

Childhood ataxia is characterised by lose of balance and impaired coordination. Ataxia does not occur due to muscle weakness rather due to the malfunction of any of the following:

- Sensory pathway
- Vestibular system
- Cerebellum

Symptoms of ataxia include poor coordination in voluntary movement like speech, swallowing and gait [15].

Patients with ataxic cerebral palsy is also observed with Dystonia which means hypertonia and reduced activity, choreathetosis, involuntary movements of limbs and muscles.

In case of ataxic cerebral palsy, loss of muscular coordination is observed with abnormal force, rhythm and accuracy [2].

Diagnosis

In most cases, the diagnosis is based on observations or parent reports on motor instabilities, such as sitting, standing, walking, posture evaluation, muscle tone, response to reflexes. (2) Conventional magnetic resonance imaging (MRI) of the brain, CT scan, X ray are also used to find the genetic aetiologies. Chromosomal microarray (CMA) is done which is a high resolution method of chromosome analysis [16, 17].

Classification

Quadriplegia, hemiplegia, Diplegia, Monoplegia, Triplegia, Ataxic Cerebral Palsy, Dyskinesia, Mixed cerebral palsy are the most commomnly found cerebral palsy classifications.(11,17) Monoplegia and Triplegia are the least common form of cerebral palsy and the most common form is Diplegia (30-40%) followed by Hemiplegia(20-30%) [11].

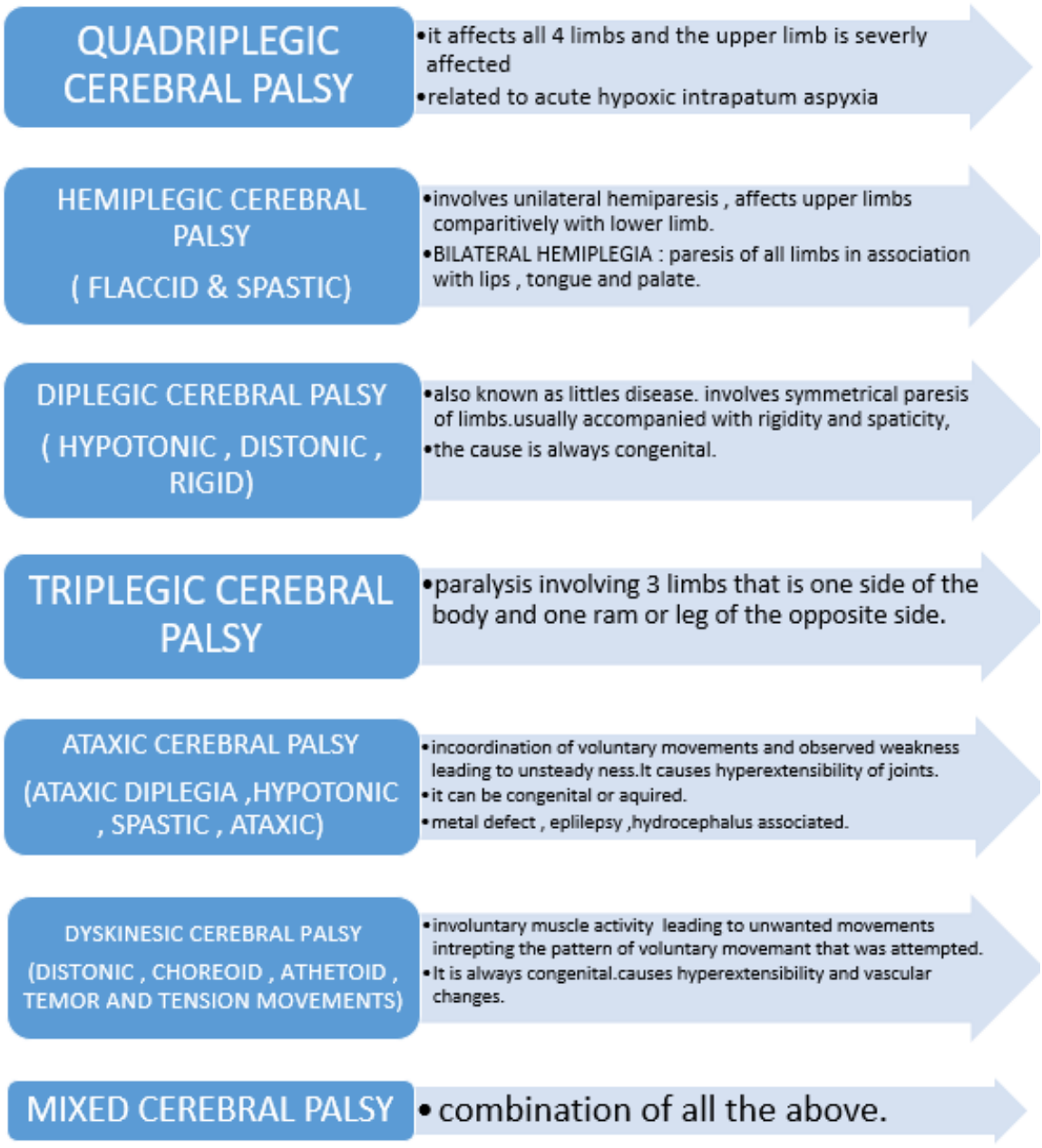

Fig 3

These can be termed as the Topographic classification.
Apart from these classifications. The division is also based upon:

1. Physiologic classification: this can be further classified into pyramidal (cases in which spaticity is prominent) and extra pyramidal (cases of chorea, athetosis, dystonia, ataxia.)
2. Etiologic classification: this classification is not well established and have not been found successful in addressing prevention.
3. Neuro pathologic classification: this was proposed to reflect and correlate the inability of the brain structure to that of the brain function.
4. Supplemental classification & associated conditions: this classification describes the conditions and abnormalities associated in children with cerebral palsy and trials to connect them to physiologic and topographic classification.
5. Functional and therapeutic classification: in functional classification understands the degree of severity of the patient condition based on the activity. The therapeutic classification deals with 4 categories – non treatment, modest interventions. Need for a treatment team, pervasive support [18].

Treatment

The treatment of the patients with cerebral palsy leads through the evaluation that includes complete patient history, from birth to present time, physical examination, radiography, other consultations and gait analysis, which provides understanding on the current stage of the disease.
After the recognition of primary deformities, the orthopaedic surgeon will assimilate the data and formulate the treatment plan of surgical or nonsurgical interventions [19].
The therapy can be differentiated into rehabilitative, medical and surgical components.
Rehabilitative components involves therapeutic approaches, orthotic bracing, strength exercises, hypnotherapy, Aqua therapy, Communication and power mobility [20].
The physiotherapy programmes are the milestone in treatment for young children with cerebral palsy [13].
The physiotherapy includes the principle of motor learning, fitness training and strength [20].
The major goals of physiotherapy is to provide sufficient strength to maintain the body, weight support, to provide active range of motion with less spasticity and dystonia [21].
Physical activity also contributes to positivity to emotional and mental function [21].
The medical management to cerebral palsy patient often include physiatrists, neurologists, paediatricians and orthopaedists [21].
Complementary and alternative medicines (CAM) is also recommended in cerebral palsy cases which consists of acupuncture, homeopathic remedies, massage, magnetic therapy, herbal treatments. But these has to be taken care when interactions with the prescribed medications [21].
The most commonly used oral medications in cerebral palsy include baclofen, diazepam, levodopa, trihexyphenidyl and botulinum toxin [20].
Injection botulinum toxin improve the kinematics of lower limbs in children with spastic cerebral palsy [22]. The use of BTX caused changes in the lower limb inter segmental coordination and found to show increased trunk movements in transverse plane [22].
In case of pre term infants, caffeine is the only treatment that decreases the risk of cerebral palsy [2].

Orthopaedic surgery

Orthopaedic surgery involves the treatment of improvising the muscoskeletal functions and ambulation that is often confined as spasticity, joint dislocation, contractures and bone deformities. The orthopaedic surgery is determined according to the CNS maturation, rate at which contractures and lever arm dysfunction (LAD) and ambulation potential [23].
At present the muscoskeletol deformities are fixed with single event multilevel surgery (SEMLS).
Goals of orthopaedic surgery,

- To decrease spasticity
- To correct LAD and dislocated joints
- To correct contractures.

Apart from this, it is found that orthopaedic surgery cannot cure the abnormalities with motor control, balance and strength which are the prominent symptoms in ataxic cerebral palsy [23].

Conclusion

Chronic non progressive disorders of motor function due to the damage caused in the brain, can be stated as a simple definition for cerebral palsy [24]. In all the cerebral palsies, ataxic cerebral palsy accounts for 5-10%. 50% of all ataxic cerebral palsy cases are due to the single gene defect [25].
There are studies were the children with ataxic cerebral palsy, found to have de novo mutation in gene such as KCNC3 (encoding voltage gated potassium channels), ITPRI (encodes receptor for inositol 1, 4, 5 triphosphate) and SPTBN2 (encodes beta3 spectrin [26].
The present day findings provides a new approach for the treatment of spasticity and contracture palsy which may give rise to promising avenues for better understanding of disorders such as cerebral palsy [24].

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