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Cerebral palsy

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Abstract

Cerebral palsy (CP) is a neurodevelopmental disorder characterized by abnormalities of muscle tone, movement and motor skills, and is attributed to injury to the developing brain. The clinical features of this entity evolve over time and the specific CP syndrome may be recognizable only after 3-5 y of age; although suggestive signs and symptoms may be present at an earlier age. The management involves neurological rehabilitation (addressing muscle tonal abnormalities, and devising physical and occupational therapies) and diagnosis and management of co-morbidities (including epilepsy, impairment of cognition, vision, hearing, and disturbances of growth and gastrointestinal function). The management, therefore, is multidisciplinary involving the treating physician working with a team of rehabilitation-, orthopedic-, psychologic-, and social care- providers.

Introduction

Cerebral palsy (CP) is primarily a neuromotor disorder that affects the development of movement, muscle tone and posture [1][3]The underlying pathophysiology is an injury to the developing brain in the prenatal through neonatal period [1][3].Although the initial neuropathologic lesion is non-progressive, children with CP may develop a range of secondary conditions over time that will variably affect their functional abilities [4][5]

Based on an international consensus, a generally agreed upon definition of CP is as follows:

CP describes a group of permanent disorders of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or immature brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems [1].

CP is characterized by heterogeneity in risk factors, underlying specific etiology, clinical features, severity of functional limitations, associated and secondary conditions, treatment options, and evolution of the condition over the lifespan of the individual [6][8], has explored the argument for a consideration to view CP as a spectrum disorder rather than a discrete unitary clinical condition [9].

The prevalence of CP for all live births ranges from 1.5 to 3 per 1,000 live births, with variation between high income and low to middle income countries and geographic region [2][5][7][10] Because, in many infants and children, abnormal neuromotor findings tend to resolve within the first few years, especially during the first 2–5 years, of life, the reported prevalence of CP tends to be higher during infancy. Although prematurity and low birthweight are main risk factors for CP, multiple other factors are also associated with or potentially increase the risk for CP [2][5][7][13][15]Multiple epidemiological studies report that half of the children who develop CP were born at term without any identified risk factor [3][8][15] Although in most cases, CP is a result of an injury to the fetal or neonatal brain, post-neonatal onset CP has been recognized. Postneonatal CP results from an injury to the brain after neonatal period and before 5 years of age[5][15]The most common causes of postneonatal CP are traumatic brain injury, near-drowning, and meningitis [15]

Epidemiology

Cerebral palsy occurs in about 2.1 per 1000 live births.[16] In those born at term rates are lower at 1 per 1000 live births.[17] Rates appear to be similar in both the developing and developed world.[17] Within a population it may occur more often in poorer people. The rate is higher in males than in females; in Europe it is 1.3 times more common in males. There was a "moderate, but significant" rise in the prevalence of CP between the 1970s and 1990s. This is thought to be due to a rise in low birth weight of infants and the increased survival rate of these infants. The increased survival rate of infants with CP in the 1970s and 80s may be indirectly due to the disability rights movement challenging perspectives around the worth of infants with disability, as well as the Baby Doe Law.[18]

As of 2005, advances in the care of pregnant mothers and their babies has not resulted in a noticeable decrease in CP. This is generally attributed to medical advances in areas related to the care of premature babies (which results in a greater survival rate). Only the introduction of quality medical care to locations with less than adequate medical care has shown any decreases. The incidence of CP increases with premature or very low weight babies regardless of the quality of care.[19] As of 2016, there is a suggestion that both incidence and severity are

slightly decreasing more research is needed to find out if this is significant, and if so, which interventions are effective.[20]
Prevalence of cerebral palsy is best calculated around the school entry age of about 6 years, the prevalence in the U.S. is estimated to be 2.4 out of 1000 children.[21]

Etiology

Cerebral palsy is caused by a problem with the brain that happens before, during or soon after birth.[22]
The brain can either being damaged or not develop normally, although the exact cause is not always clear.

Problems before birth

Cerebral palsy is usually caused by a problem that affects the development of a baby's brain while it's growing in the womb.[7]

These include:

- damage to part of the brain called white matter, possibly as a result of a reduced blood or oxygen supply – this is known as periventricular leukomalacia (PVL)[12]
- an infection caught by the mother – such as cytomegalovirus, rubella, chickenpox or toxoplasmosis
- a stroke – where there's bleeding in the baby's brain or the blood supply to their brain is cut off
- an injury to the unborn baby's head

Problems during or after birth

Cerebral palsy can also sometimes be caused by damage to a baby's brain during or shortly after birth.

For example, it can be due to:

- the brain temporarily not getting enough oxygen (asphyxiation) during a difficult birth
- an infection of the brain, such as meningitis
- a serious head injury
- choking or nearly drowning, resulting in the brain not getting enough oxygen
- a very low blood sugar level
- a stroke[13]

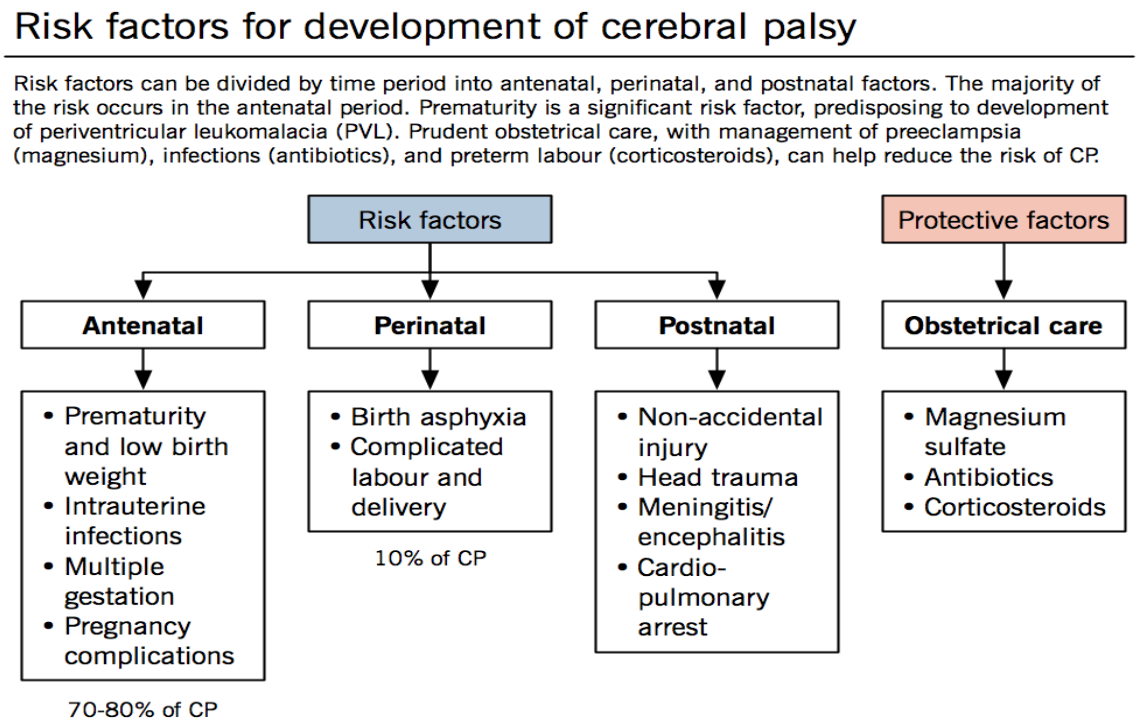

Figure.1risk factors for development cerebral palsy

Increased risk

Some things can increase a baby's risk of being born with cerebral palsy including:

- being born prematurely (before the 37th week of pregnancy) – babies born at 32 weeks or earlier are at a particularly high risk
- having a low birthweight
- being part of a multiple birth, such as a twin or triplet
- the mother smoking, drinking a lot of alcohol, or taking drugs such as cocaine, during pregnancy

recommend regular check-ups to look for symptoms of cerebral palsy during the first 2 years of their life if they have an increased risk of developing the condition[15]

Types of cerebral palsy

The type of movement issues seen in a person with cerebral palsy depends on how severely a brain injury has affected muscle tone. Muscle tone is defined as the strength and tension of the muscles.[23] There are two common terms used to describe how cerebral palsy affects muscle tone hypotonia and hypertonia. These are useful to better understand the types of cerebral palsy.

Hypotonia: Low muscle tone that causes a loss of strength and firmness, resulting in floppy muscles

Hypertonia: High muscle tone that causes rigidity and spasmodic movement, resulting in stiff muscles.[24]

Different Types of Cerebral Palsy

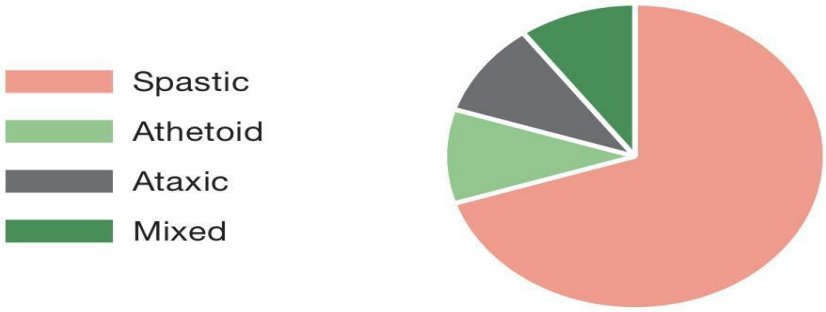

Figure.2 prevalence and classification of cerebral palsy

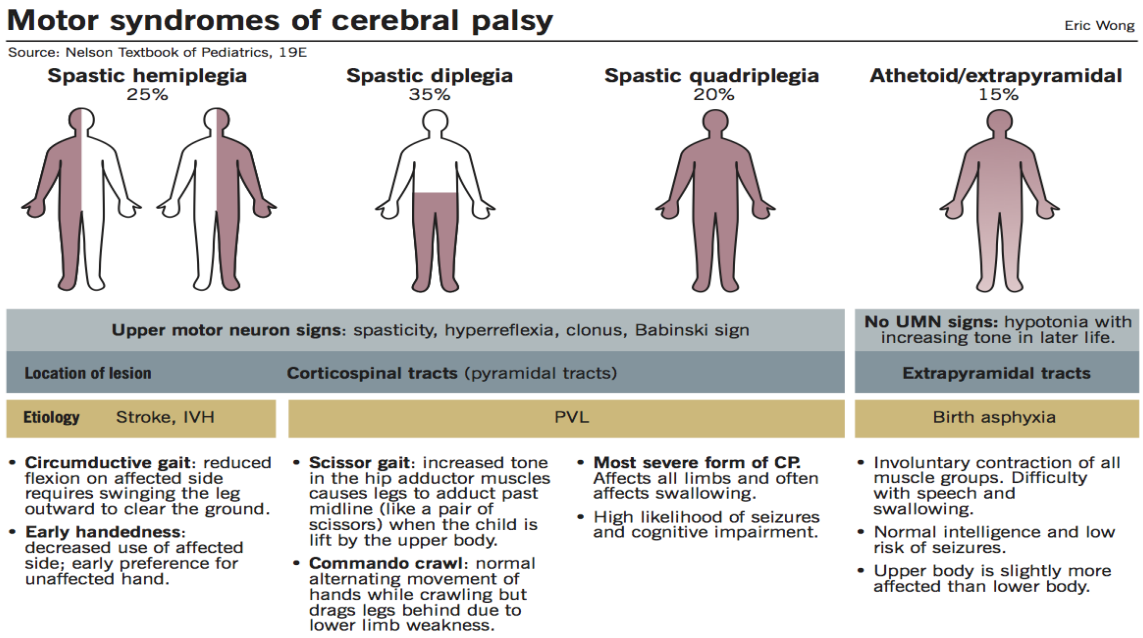

Figure.3 motor syndrome of cerebral palsy.

Spastic Cerebral Palsy

Spastic cerebral palsy is one of the most common types of cerebral palsy, making up 70%-80% of cases. People with this type often experience spasticity and exaggerated or jerky movements (hypertonia).[25]

Spastic cerebral palsy is caused by damage to the brain's motor cortex, which controls voluntary movement. It is also caused by damage to the

pyramidal tracts, which help relay signals to the muscles. This is why spastic cerebral palsy is sometimes referred to as “pyramidal”.
The motor cortex is found on both sides of the brain, and the pyramidal tracts connect each side of the motor cortex. Damage to the right side of the motor cortex causes movement problems on the left side of the body, and vice versa.[26[

Common signs and symptoms of spastic cerebral palsy include:[25]

- Abnormal walking
- Awkward reflexes
- Contractures (permanently tightened muscles or joints)
- Stiffness in one part of the body

Athetoid Cerebral Palsy

About 10% of children with the condition are diagnosed with athetoid cerebral palsy, or non-spastic cerebral palsy. This palsy type is also called dyskinetic cerebral palsy.[12]
This form is characterized by a mixture of hypotonia and hypertonia, which causes muscle tone to fluctuate. The main trait of athetoid cerebral palsy is involuntary movement in the face, torso, and limbs.
This type of cerebral palsy is caused by damage to the brain’s basal ganglia and/or cerebellum. The basal ganglia regulates voluntary motor function and eye movement. The cerebellum controls balance and coordination.
Athetoid cerebral palsy is considered extrapyramidal. The extrapyramidal tracts in the brain regulate involuntary reflexes and movement signaled by the basal ganglia and cerebellum.[27]

Common symptoms associated with athetoid cerebral palsy include:

- Floppiness in the limbs
- Issues feeding
- Problems with posture
- Stiff or rigid body

Figure.4 child with athetoid cerebral palsy

Ataxic Cerebral Palsy

Ataxia is a type of cerebral palsy that causes problems with balance and coordination. Ataxic cerebral palsy makes up a small percentage of cases. Those with ataxic CP typically have issues surrounding voluntary movement.

Ataxic cerebral palsy differs from other types of cerebral palsy because it is caused by damage to the cerebellum. The cerebellum is the part of the brain that controls balance and coordination. People with ataxic cerebral palsy often experience tremors and a reduction in muscle tone.[12]

Common symptoms of ataxic cerebral palsy include:

- Difficulty speaking
- Problems with depth perception
- Shakiness and tremors
- Spreading feet apart when walking

Mixed Cerebral Palsy

Sometimes damage to the developing brain is not confined to one location. It is possible for a child to develop different types of cerebral palsy caused by several areas of brain damage.

When a child is showing symptoms of several types of cerebral palsy, it is considered to be mixed cerebral palsy. This diagnosis makes up less than 10% of all cerebral palsy cases.

The most common mixed cerebral palsy diagnosis is a combination of spastic and athetoid cerebral palsy. Parents and caregivers are encouraged to seek out cerebral palsy specialists if they suspect more than one type of cerebral palsy is present.[25]

Location of Movement Problems

The location of movement problems is related to the location of a brain injury and can determine which type of cerebral palsy your child has.

- Monoplegia occurs when only one limb's movement is affected. It usually occurs in the arm or leg. This type of movement problem is very rare.
- Diplegia affects two limbs, which are usually the legs. Oftentimes, those with diplegia have mild movement problems in the upper body as well. In those with cerebral palsy, diplegia is commonly the result of premature birth.
- Hemiplegia affects one entire side of the body. The arm is usually more impacted than other limbs, distinguished by a rigidly flexed wrist or elbow. Some people with hemiplegia may not be able to use the affected hand. Prenatal bleeding in the brain can cause hemiplegia.
- Triplegia occurs when three limbs are affected. This may occur if both legs and one arm have impacted movement.
- Quadriplegia impacts all four limbs, but the legs are generally affected worse than the arms. There may be limited control over facial muscles as well.
- Double hemiplegia occurs when all four limbs are affected, but one side is impacted more than the other side.[23]

Diagnosis

Signs and symptoms of cerebral palsy can become more apparent over time, so a diagnosis might not be made until a few months after birth.[26] If family doctor or pediatrician suspects that the child has cerebral palsy, he or she will evaluate child's signs and symptoms, monitor growth and development, review l child's medical history, and conduct a physical exam[27]. refer to a specialist trained in treating children with brain and nervous system conditions (pediatric neurologist, pediatric physical medicine and rehabilitation specialist, or child developmental specialist). also order a series of tests to make a diagnosis and rule out other possible causes.[28]

Brain scans

Brain-imaging technologies can reveal areas of damage or abnormal development in the brain.[26]

These tests might include the following:

MRI. An MRI scan uses radio waves and a magnetic field to produce detailed 3D or cross-sectional images of child's brain. An MRI can often identify lesions or abnormalities in child's brain.[12]

This test is painless, but it's noisy and can take up to an hour to complete. child will likely receive a sedative or light general anesthesia beforehand.[29]

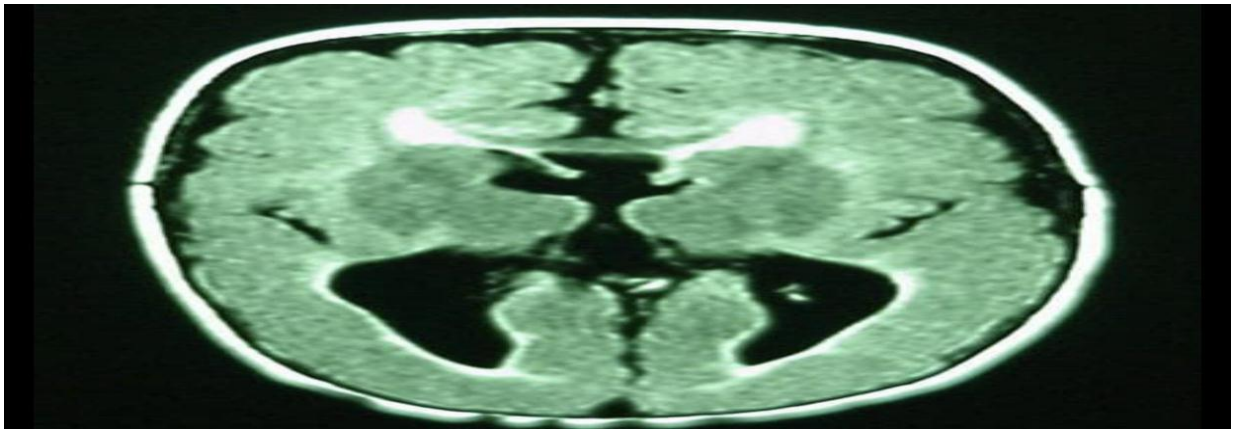

figure 5 . magnetic resonance image (MRI) of 1-years old boy who was born at gestational week 27.

Cranial ultrasound.

This can be performed during infancy. A cranial ultrasound uses high-frequency sound waves to produce images of the brain. An ultrasound doesn't produce a detailed image, but it may be used because it's quick and inexpensive, and it can provide a valuable preliminary assessment of the brain.[28]

Electroencephalogram (EEG)

If child is suspected of having seizures, an EEG can evaluate the condition further. Seizures can develop in a child with epilepsy. In an EEG test, a series of electrodes are attached to your child's scalp.

The EEG records the electrical activity of your child's brain. It's common for there to be changes in normal brain wave patterns in epilepsy.[28]

Laboratory tests

Tests on the blood, urine or skin might be used to screen for genetic or metabolic problems.[26]

Additional test

If child is diagnosed with cerebral palsy, you'll likely be referred to specialists to test for other conditions often associated with the disorder. These tests can identify problems with.[29]

- Vision
- Hearing
- Speech
- Intellect
- Development
- Movement

MANAGEMENT

The goal of management of cerebral palsy is not to cure or to achieve normalcy but to increase functionality, improve capabilities, and sustain health in terms of locomotion, cognitive development, social interaction, and independence. The best clinical outcomes result from early, intensive management. Optimal treatment in children requires a team approach.[27] A modern team approach focuses on total patient development, not just on improvement of a single symptom. Treatmentprograms encompass physical and behavioural therapy, pharmacologic and surgical treatments, mechanical aids, and management of associated medical conditions. In physical, occupational, speech, and behavioural therapies, the goals include enhancing patient and caregiver inter- actions while providing family support.[27] Management of spasticity is a major challenge to treatment team. Various forms of therapy are available to people living with cerebral palsy as well as caregivers and parents caring for someone with this disability. They can all be useful at all stages of this disability and are vital in a CP person’s ability to function and live more effectively[26]

Oral Medications

Oral medications are a systemic, rather than focal, treat- ment for spasticity in children with cerebral palsy. Oral medications commonly used in children are baclofen, diazepam, clonazepam, dantrolene and tizanidine.[30]

Intrathecal Baclofen

Intrathecal baclofen (ITB) was approved for the treat- ment of spasticity of cerebral origin in 1996. ITB is a surgically implanted system used to control spasticity by infusing baclofen directly into the spinal canal and

around the spinal cord.26Baclofen inhibits spasticity by blocking excitatory neurotransmitters in the spinal dorsal horn. ITB maximizes the dose delivered to spinal recep- tors and minimizes the side effects associated with oral baclofen.[33][34]

Botulinum Toxin

Botulinum toxin (BT) injection is now an established first-line treatment for focal spasticity[35]. Botulinum toxin type A produces dose-related weakness of skeletal muscle by impairing the release of acetylcholine at the neuro- muscular junction. This partially interrupts muscle contraction making the muscle temporarily weaker. Muscles commonly treated with BT include the gastroc- nemius-soleus complex, hamstrings, hip adductors and flexor synergy muscles of the upper extremity[36][37]. Intramus- cular injections can be localized by surface landmarks, electromyography stimulation, and/or ultrasound[38]. Following injection, muscle relaxation is evident within 48 to 72 hr and persists for a period of 3 to 6 months. Botox injection can help improve a child's ability to walk or use hands and allow for a better fitting orthotics by reducing spasticity. Therapists can take advantage of the time when an overly powerful muscle is weakened to work on strengthening the muscle on the opposite side of the joint (antagonist). Sometimes, casting of the involved extremity is done after the injection to increase the stretch of the tight muscle.[35][36][40]

SURGERY

Orthopedic surgery

Surgery is mainly undertaken on the lower limb, but occasionally in the upper limb. Some children require surgery for scoliosis. Physiotherapy is an essential part of post-operative management. Gait laboratories are useful in planning the surgical program for children who can walk independently or with sticks or walking frames.

• The hip: soft tissue surgery is often effective for children when the hip problems are detected at an early stage (hence the importance of regular X-rays). Lengthening of the adductor muscles may be all that is required in younger children. However, if the problem progresses, and especially if it is neglected, more extensive surgery to the hip bones is required for a significant number of children. For most children surgery to keep the hips in joint, or to put the hips back in joint, is preferable to

leaving the child with a dislocated hip which is frequently painful in later life.

• The knee: lengthening of the hamstrings can help the knee straighten and so improve the walking pattern. Sometimes transferring a muscle from the front to the back of the knee can also help by reducing stiffness around the knee.

• The ankle and foot: This is the commonest area where orthopedic surgery is required. Sometimes children require orthopedic surgery in several different areas (for example, hip, knee and ankle). Frequently this now involves a single hospitalization and is called ‘multilevel surgery’. Multilevel surgery is of most benefit to children whowalk independently or with the assistance of crutches.

The best age is usually between 8 and 12 years old although it can occasionally be helpful for older or younger children.[26][31][40]

TREATMENT FOR THE ASSOCIATED MEDICAL PROBLEMS

1. Epilepsy

Knowledge of epilepsy has increased substantially in the past few years. There are many types of epilepsy, and medication is often prescribed following a careful diagnosis of the type of seizures and their cause. The most commonly used anticonvulsants are: Carbamazepine, Sodium valproate, Lamotrigine, Phenytoin etc.

2. Saliva control

The speech pathologist plays a central role and can provide strategies to improve dribbling problems. When these strategies are not effective, medication isoccasionally used, particularly in children over the age of six years. These medications are as follows:

• Benzhexol hydrochloride (‘Artane’) reduces salivary secretions. Occasionally irritability may occur. Blurring of vision, constipation and difficulty with urination are also potential side effects.

• lycopyrrolate (‘Robinul’) is like benzhexol hydro- chloride but seems to produce fewer side effects.

3. Constipation

Children with cerebral palsy often have problems with constipation. A high fiber diet and increased fluid intake can help with this problem. This may not be easily achieved in some children with cerebral palsy. Careful use of laxatives is preferable to severe constipation.

4. Nutrition

A dietitian can provide useful advice about adequate nutrition. Excessive weight gain can be very disadvanta- geous for children learning to walk. Under nutrition and failure to make adequate weight gains may be related to feeding difficulties. In a small proportion of children, the use of tube feeding can be helpful.[40]

Prognosis

The use of the GMFCS (in the Ontario Motor Growth Study) has been shown to be an effective tool in assessing outcomes for individuals with CP.[12]

Motor assessments have been used alongside growth charts to characterize gross motor development over time.

These trends can be divided into 5 distinct motor development curves which children can be categorized into to assist with providing further prognostic information for parents.

Prognosis for motor function depends on the type and severity of motor impairment.[12][40]

Individuals with CP on average have a life expectancy that is 44% of normal (this can be applied to countries with varying life expectancy rates).

Mortality risk increases with increasing number of impairments (e.g., intellectual, hearing, vision).

Research has shown that the strongest predictors of early mortality are immobility and impaired feeding ability (i.e., the need for tube feeding).

Shortest life expectancy is associated with individuals who are unable to lift their head in prone position.[41]

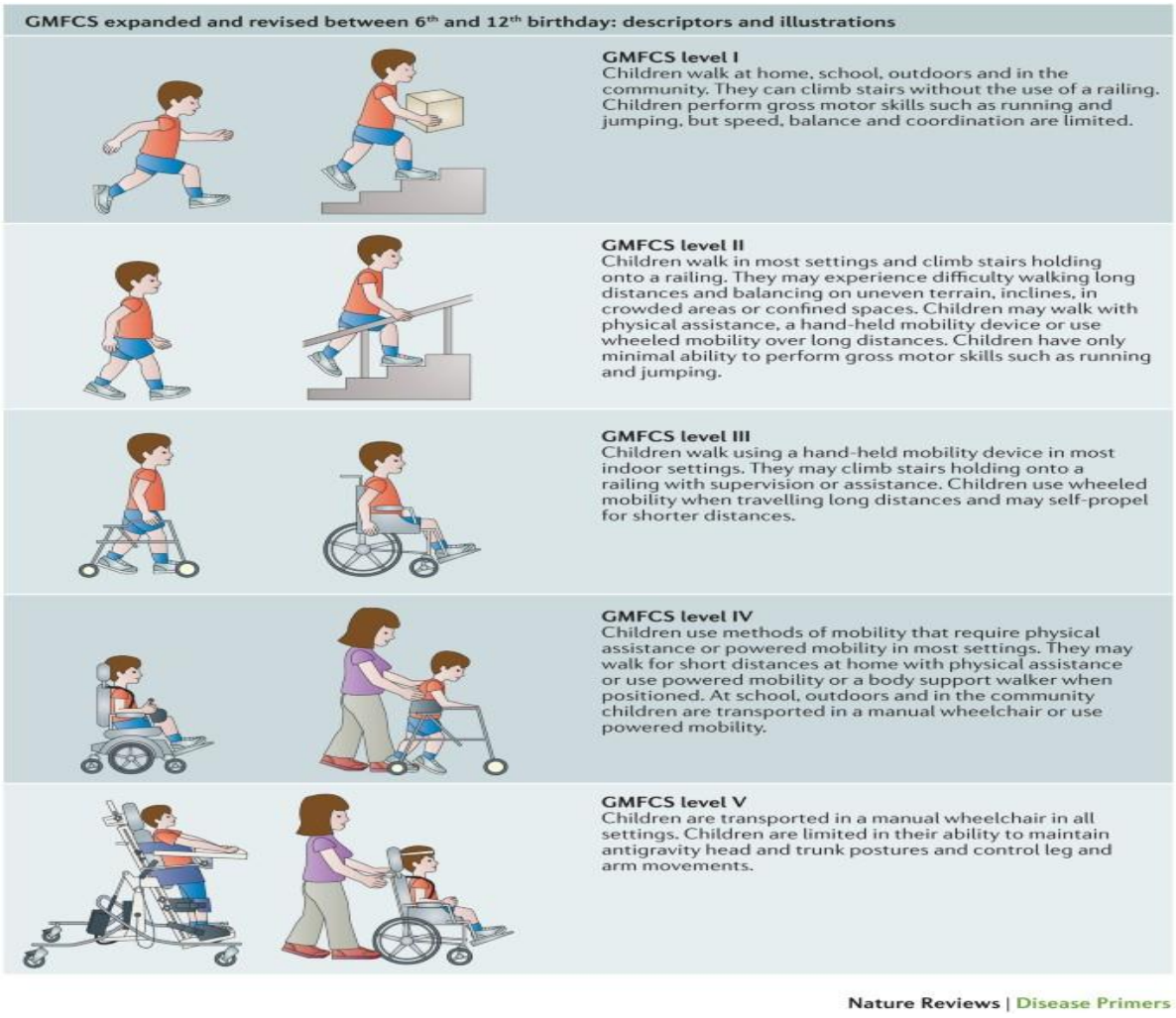

Figure.6 Gross Motor Function Classification System expanded and revised for children with cerebral palsy, 6–12・years of age.

Conclusion

Cerebral Palsy involves an insult to a fetal or infant brain that results in disordered and limited movement and posture. Although the brain damage is non-progressive, the secondary changes that occur to the infant's musculoskeletal and nervous systems are progressive, and can be influenced by early experiences. Early physiotherapy interventions can provide the infant with the opportunity to practice functional movements that would otherwise not be possible due to muscle weakness and poor motor control. By practicing meaningful and effective skills in varied environments, the child will have the opportunity to acquire skills that will allow them to be more independent with ADLs and increase their level of participation in society.

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