

# 1. What is Cerebral Palsy?

## Introduction

Cerebral palsy refers to a group of disorders that affect movement. It is a permanent, but not unchanging, physical disability caused by an injury to the developing brain, usually before birth.

Cerebral palsy may only be mild and cause only a slight disruption to a person's daily life. It can also be more severe, affecting the whole body and may significantly impact how a person participates in daily activities.

In India, there are approximately **33,000** people with cerebral palsy. Worldwide, the incidence of cerebral palsy is **1 in 500 births**. For most people with cerebral palsy, the cause is unknown. There is no known cure.

## Types

There are three predominant types of cerebral palsy and each are characterised by different movement patterns. Movements can be uncontrolled or unpredictable, muscles can be stiff or tight and in some cases people have shaky movements or tremors.

## Associated Impairments

In addition to their motor disability, people with cerebral palsy may have other associated impairments including epilepsy, and disorders of speech, vision, hearing and/or intellect.

## Diagnosis

Cerebral palsy, except in its mildest forms, can be seen in the first 12 months of life. Doctors may suspect cerebral palsy if a baby has slow motor development (does not reach movement milestones) has tight or floppy muscle tone, or displays unusual postures. Babies most at risk of cerebral palsy are those born prematurely or with low birth weight. Multiple births (e.g. twins or triplets) are also associated with higher rates of cerebral palsy.

## Life Expectancy

Most people with cerebral palsy have a normal life expectancy. The damage to the brain that causes cerebral palsy does not progress (get worse) as the person gets older.

## 2. What Causes Cerebral Palsy?

### Causes

There is rarely one single cause of cerebral palsy. For most babies born with cerebral palsy, the cause remains unknown.

For a long time, it was believed that cerebral palsy was due to complications at birth, including asphyxia (lack of oxygen). Researchers now understand that this contributes to only a very small percentage of cases of cerebral palsy.

Now, it is generally accepted that cerebral palsy usually arises from a series of 'causal pathways', i.e. a combination of events that can lead to an injury in a baby's developing brain.

In 13 out of 14 cases of cerebral palsy in India, the brain injury leading to cerebral palsy occurs either in the uterus (while the mother is pregnant) or before 1 month of age. At present, the cause is not well understood for most of these babies.

Stroke is the most common cause in babies who acquire cerebral palsy after 1 month of age. The stroke may occur spontaneously or arise from surgical or heart complications.

### Risk Factors

Some risk factors for cerebral palsy have been identified. These include:

- **premature birth**
- **low birth weight** (small for gestational age)
- **blood clotting problems** (thrombophilia)
- an inability of the placenta to provide the developing foetus with oxygen and nutrients
- **RH or A-B-O blood type** incompatibility between mother and baby
- **infection** of the mother with German measles or other **viral diseases** in early pregnancy
- **bacterial infection** of the mother, foetus or baby that directly or indirectly attacks the infant's central nervous system
- **prolonged loss of oxygen** during the pregnancy or birthing process, or **severe jaundice** shortly after birth.

It is important to understand that even if a mother or infant does have any of these risk factors, it doesn't mean that this will definitely result in cerebral palsy. It just means that the chance of having a child with cerebral palsy is increased.

## Who is at Greatest Risk?

The Indian Cerebral Palsy Register Report 2009 identified four groups that, statistically, have a greater risk of cerebral palsy.

**1. Males** - Males are at greater risk of having cerebral palsy.

**2. Premature babies** - Prematurity is associated with higher rates of cerebral palsy. 42% of children with cerebral palsy are born prematurely, compared to 8% of the Indian population.

**3. Small babies** - Low birth weight is associated with higher rates of cerebral palsy. This may be a result of prematurity or slow intrauterine growth. 43% of children with cerebral palsy had low birth weight, compared to just over 6% of the Indian population.

**4. Twins, triplets and higher multiple births** - Multiple births are associated with higher rates of cerebral palsy. 11% of children with cerebral palsy were from a multiple birth, whereas the rates of multiple births are only 1.7% in the Indian population.

## Is There A Genetic Link?

For many years, the common understanding has been that cerebral palsy cannot be inherited, and this is basically true. Familial cerebral palsy is uncommon; approximately 1% of people with cerebral palsy will have a sibling with the condition. It is even uncommon in twins – when one twin has cerebral palsy, 90% of co-twins will not have cerebral palsy. However small these statistics, they are enough to suggest that there might be some genetic factors involved in cerebral palsy.

Researchers generally believe that a genetic disposition to characteristics such as:

- prematurity
- small size for gestational age; and
- heart problems ...

... may start a chain of events (a causal pathway). The combination of one or several events along a pathway can result in a child having cerebral palsy.

Any number of problems can occur in the neonatal period when key organs are still developing. For example, a baby in utero may have a genetic predisposition to prematurity and then contract a virus or infection. The baby's immune system may not respond appropriately which may in turn lead to a premature birth. This baby may need intensive care after birth, but then progress well. It might not be until they reach 6-12 months of age, when developmental milestones are not reached, that a diagnosis of cerebral palsy is made. In this case, it becomes difficult to say that any one factor caused the cerebral palsy. Some children who have a genetic

predisposition to a known risk factor might not experience any other events that lead them on one of the pathways that result in cerebral palsy.

In 10% of children with cerebral palsy, brain malformations are present (where the injury to the developing brain happens very early in development). It is thought that this group, as well as those who have had a neonatal stroke (due to inherited thrombophilia), are the most likely to have genetic factors involved.

Families often ask, "I have had one child with cerebral palsy - how likely is it that my next child will also have cerebral palsy?" If you think along the lines of a causal pathway, this is very difficult to answer with certainty. It is likely that your chance is the same as for the first baby. What that chance is, we are unable to tell. For the population as a whole, the chance of having a baby who has cerebral palsy is 1:500. For some, this chance might be higher.

To discuss your concerns, it is advisable to seek genetic counseling before conceiving your next child.

## Can Cerebral Palsy Be Prevented or Cured?

At present, there is no way to totally prevent or cure cerebral palsy.

Two interventions are now being used to reduce the risk or severity of cerebral palsy:

- **Magnesium Sulphate** – is given to pregnant mothers when they are at risk of very preterm birth. This can help protect babies from the brain injury that leads to cerebral palsy.
- **Cooling Cap** – Newborn babies who have suffered a brain injury due to lack of oxygen before birth (hypoxic ischemic encephalopathy) may be treated with a special cooling cap which aims to reduce the impact of the brain injury.

## Brain Plasticity

Recent exciting advances in research have shown that the brain is 'plastic' – that is, the brain changes with every different activity it performs. When certain parts of the brain are damaged, sometimes other areas of the brain can, and will, take over to compensate for the injury.

This offers great hope that gene and stem cell therapies may one day be used, even before a baby is born, to repair some or all of the damage to the developing brain that leads to cerebral palsy.

## 3. Diagnosing Cerebral Palsy

### Introduction

Cerebral palsy is a complex disability and diagnosis is not always an easy process. Doctors may suspect cerebral palsy if a baby has slow motor development (does not reach movement milestones), has tight or floppy muscle tone, or displays unusual postures.

### When is Cerebral Palsy Diagnosed?

The time at which a child is diagnosed with cerebral palsy varies. If a baby is very premature, early scans (such as an MRI - magnetic resonance imaging) might show that they have an injury to the brain, but at that stage it is often too early to predict the impact.

The General Movements Assessment can be conducted from birth until 3 months of age. It has been shown to be a strong predictor of cerebral palsy, particularly when certain changes to the brain are seen on an MRI. If the result is that a baby is "at risk of cerebral palsy", then intervention can start as early as one month. The General Movements Assessment cannot, however, predict the severity of cerebral palsy.

Most children with cerebral palsy are not born prematurely. Most are born at full term and it is not until they do not meet the usual infant milestones that any form of disability is considered.

### Milestones

All babies develop at different rates, but they generally reach a set of milestones around 3, 6, 9 and 12 months of age.

These might include:

- holding own head up when lying on their stomach or in a supported sitting position
- sitting and rolling over by 6 months
- walking by 12-18 months
- speaking in simple sentences by 24 months.
- In NSW, questions relating to these milestones are described in the Personal Health Record book (Blue Book) provided to families when their child is born. Parents are usually the ones to notice that their child is not reaching these milestones and may alert their early childhood nurse, general practitioner or paediatrician

## How Do Doctors Diagnose Cerebral Palsy?

Cerebral palsy is a condition that:

- is permanent but not unchanging
- involves a disorder of movement and/or posture and of motor function
- is due to a non-progressive lesion or abnormality which has originated in the immature brain.

In order to make a formal diagnosis of cerebral palsy, doctors will take a complete medical history and carefully examine the child. They will pay special attention to the child's movements – both their voluntary movements as well as their muscle tone. Some children may have very relaxed, floppy muscles, while others have stiff, tight muscles. Doctors will also look for any unusual postures or if the child favours one side over the other.

Scans such as MRI (magnetic resonance imaging) or CT (computed tomography) may be ordered by the doctor.

One of the frustrations for parents is that sometimes a diagnosis can take a long time, with repeated tests and visits to specialists. This may be because the child has a mild form of cerebral palsy, but it could also be because the doctor needs to make sure it is not another type of movement disorder that may be progressive (get worse over time).

## 4. Types of Cerebral Palsy

Cerebral palsy can be described or classified in different ways - by:

### 1. Its impact on movement

#### A. Spastic Cerebral Palsy

This is the most common type of cerebral palsy. Spasticity means stiffness or tightness of muscles. The muscles are stiff because the message to the muscles is sent incorrectly through the damaged part of the brain. This is the most common type, affecting approximately 77-93% of people with the condition.

When we perform a movement, some groups of muscles become tighter and some groups of muscles relax. In people with spastic cerebral palsy, both groups of muscles may become tighter. This makes movement difficult or even impossible.

#### B. Dyskinetic Cerebral Palsy

This type affects about 2-15% of people with cerebral palsy. There are two forms:

- Athetosis is characterised by uncontrolled, slow, 'stormy', writhing movements;
- Dystonia is characterised by sustained or intermittent muscle contractions causing twisting or repetitive movement.

#### C. Ataxic Cerebral Palsy

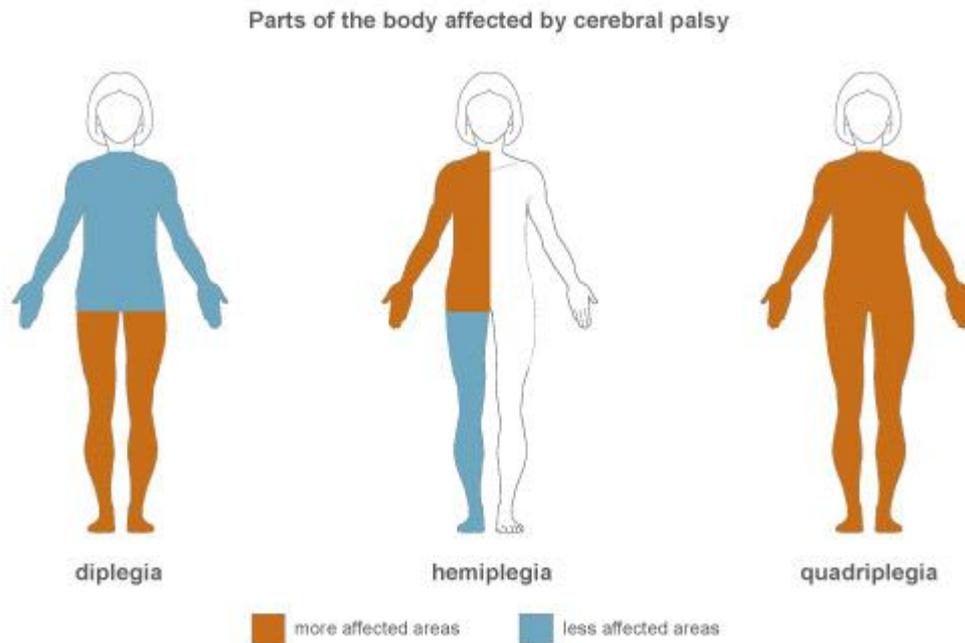
This is the least common type of cerebral palsy (2-8%) and is characterised by shaky movements. It affects a person's balance and coordination.

#### D. Mixed Forms

Many people have signs of more than one of the three types. The most common mixed form includes spasticity and dystonic movements, but other combinations are also possible.

## 2. The part of the body that it affects

The parts of the body affected by cerebral palsy differ from one person to another. There are specific words used to describe the parts affected:



- **Hemiplegia** - the leg and arm on one side of the body are affected.
- **Diplegia** - both legs are affected significantly more than the arms. People with diplegia may have some clumsiness with their hand movements.
- **Quadriplegia** - both arms and legs are affected. The muscles of the trunk, face and mouth can also be affected.

The impact of cerebral palsy in terms of motor ability or function is commonly described by:

## 3. Its impact on gross motor skills

The gross motor skills (e.g. sitting and walking) of children and young people with cerebral palsy can be categorised into 5 different levels using a tool called the Gross Motor Function Classification System (GMFCS).

GMFCS looks at movements such as sitting and walking. It is helpful because it provides families and clinicians with:

- a) a clear description of a child's current motor function, and
- b) an idea of what equipment or mobility aids a child may need in the future, e.g. crutches, walking frames or wheelchairs.

## **GMCFS Levels**

The GMFCS levels are as follows:

**Level I** - Walks without limitations.

**Level II** - Walks with limitations.

**Level III** - Walks using a hand-held mobility device.

**Level IV** - Self-mobility with limitations; may use powered mobility.

**Level V** - Transported in a manual wheelchair.

Generally, a child or young person over the age of 5 years will not move between GMFCS levels so, if for example, a child is at Level IV at the age of 6 then it is likely that they will need to use a mobility device throughout their life.

### **Level I: Uses no assistive devices (such as crutches)**

1. Can walk indoors and outdoors and climb stairs without using hands for support.
2. Can perform usual activities such as running and jumping.
3. Has decreased speed, balance and coordination.

### **Level II: Walks independently**

4. Is limited in outdoor activities.
5. Has the ability to walk indoors and outdoors and climb stairs with a railing.
6. Has difficulty with uneven surfaces, inclines or in crowds.
7. Has minimal ability to run or jump.

### **Level III: Uses assistive mobility devices**

8. Walks with assistive mobility devices indoors and outdoors on level surfaces.
9. May be able to climb stairs using a railing.
10. May propel a manual wheelchair (with assistance needed for long distances or uneven surfaces).

### **Level IV: Severely limited**

11. Self-mobility severely limited even with assistive devices.
12. Uses wheelchairs most of the time and may propel their own power wheelchair.
13. May participate in standing transfers.

## Level V: No Self-Mobility

14. Has physical impairments that restrict voluntary control of movement and the ability to maintain head and neck position against gravity.
15. Is impaired in all areas of motor function.
16. Cannot sit or stand independently, even with adaptive equipment.
17. Cannot independently mobilise, though may be able to use powered mobility.

## 4. Its impact on fine motor skills

The ability of children with cerebral palsy to handle objects in everyday activities can be categorized into 5 levels using the Manual Ability Classification System (MACS).

Knowing a child's MACS level can help parents, teachers and others to understand in which situations a child is independent and the extent to which they need support or adaptations.

MACS level is determined based on knowledge about the child's actual performance in daily life. It is not done by conducting a specific assessment, but by asking someone who knows the child and how that child performs typically. MACS is based on the use of both hands in activities, not an assessment of each hand separately.

	<b>Level</b>	<b>Distinction Between Levels</b>
<b>1.</b>	<b>Handles objects easily and successfully</b> - At most, limitations in the ease of performing manual tasks requiring speed and accuracy. However, any limitations in manual abilities do not restrict independence in daily activities.	<b>Distinction between Levels I and II</b> - Children in Level I may have limitations in handling very small, heavy or fragile objects which demand detailed fine motor control, or efficient coordination between hands. Limitations may also involve performance in new and unfamiliar situations. Children in Level II perform almost the same activities as children in Level I but the quality of performance is decreased, or the performance is slower. Functional differences between hands can limit effectiveness of performance. Children in Level II commonly try to simplify handling of objects, for example by using a surface for support instead of handling objects with both hands.

2.	<p><b>Handles most objects but with somewhat reduced quality and/or speed of achievement</b> - Certain activities may be avoided or be achieved with some difficulty; alternative ways of performance might be used but manual abilities do not usually restrict independence in daily activities.</p>	<p><b>Distinction between Levels II and III</b> - Children in Level II handle most objects, although slowly or with reduced quality of performance. Children in Level III commonly need help to prepare the activity and/or require adjustments to be made to the environment since their ability to reach or handle objects is limited. They cannot perform certain activities and their degree of independence is related to the supportiveness of the environmental context.</p>
3.	<p><b>Handles objects with difficulty; needs help to prepare and/or modify activities</b> - The performance is slow and achieved with limited success regarding quality and quantity. Activities are performed independently if they have been set up or adapted.</p>	<p><b>Distinction between Levels III and IV</b> - Children in Level III can perform selected activities if the situation is prearranged and if they get supervision and plenty of time. Children in Level IV need continuous help during the activity and can at best participate meaningfully in only parts of an activity.</p>
4.	<p><b>Handles a limited selection of easily managed objects in adapted situations</b> - Performs parts of activities with effort and with limited success. Requires continuous support and assistance and/or adapted equipment, for even partial achievement of the activity.</p>	<p><b>Distinction between Levels IV and V</b> - Children in Level IV perform part of an activity, however, they need help continuously. Children in Level V might at best participate with a simple movement in special situations, e.g. by pushing a simple button.</p>
5.	<p><b>Does not handle objects and has severely limited ability to perform even simple actions.</b> - Requires total assistance.</p>	

## 5.Common Issues

## 6.Interventions

## 7.What Does the Future Hold?