PedsCases Podcast Scripts

This is a text version of a podcast from Pedscases.com on "**Cerebral Palsy**." These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at www.pedcases.com/podcasts.

## Cerebral Palsy

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Hi everyone, my name is Dayae Jeong and I am a medical student at McMaster University in Ontario, Canada. This PedsCases podcast on cerebral palsy was developed with the help of Dr. Kristen Hallett, a consultant pediatrician and associate professor at McMaster Children's Hospital, and Dr. Sara Long-Gagné, a complex care pediatrician at Montreal Children's Hospital. This podcast is designed to give you an overview of cerebral palsy.

We will be addressing the following questions in our podcast:

1. What is Cerebral Palsy?
2. What are some of the risk factors that can cause CP?
3. How is CP classified, in terms of type and severity?
4. How is CP diagnosed?
5. What is the management for CP?
6. What is the prognosis for patients with CP?

### *Defining CP*

Let's get started with a clinical case. You are a third year medical student working in a general pediatrician's office, and you are asked to see Rosie. Rosie is a 16 month old girl, who was born prematurely at 32 weeks. She is being brought in by her mother because she is not yet sitting independently or pulling herself up to stand. On further questioning, you find out that she was slow to crawl as well. She has no delays in any of the other developmental domains. On exam, you find that she has a good pincer grasp, but hypertonia and hyperreflexia in her lower extremities.

This case is an example of cerebral palsy. So, what is CP? Cerebral palsy is used to describe any non-progressive motor disorder that results from injury or damage to the developing brain, typically defined as before 2 years of age. Depending on the nature and extent of the damage, CP presents with a wide range of symptoms and severities.

### *Etiologies*

So, what exactly causes cerebral palsy? It helps to divide up the etiology into factors that damage the brain before birth, during birth, and after birth.

Prior to birth, intrauterine factors can be divided into innate issues affecting the baby and external factors affecting blood flow to the baby. Innate problems can include congenital anomalies, primary central nervous system malformations, and metabolic or genetic disorders. Extrinsic factors include maternal infections, pre-eclampsia, or placental concerns, which may all limit blood flow to the fetus and interfere with development.

Perinatally, any neurological hypoxic-ischemic injury puts a child at risk for CP. For example, need for prolonged resuscitation at birth or prolonged premature lung disease reduce oxygen delivery to the brain and lead to neuronal death in areas crucial for motor function. Premature and low-birth weight infants are also at higher risk for intraventricular hemorrhage, or bleeding into the ventricles due to fragile blood vessels.

Postnatally, CP can result from metabolic conditions that cause damage to the brain, such as kernicterus from untreated unconjugated hyperbilirubinemia, trauma, or CNS infections like bacterial meningitis or viral encephalitis.

It is important to note that CP refers solely to the motor deficits caused by these varied processes. A child with CP does not necessarily have other neurological deficits like an intellectual disability or seizure disorder. However, it is not uncommon for other areas of the brain to be affected by the insult, leading to concurrent co-morbidities.

***Clinical presentation: Types of CP***

So, as you can see, CP results from sort of disruption to the motor areas of the brain. There are two main neuronal tracts involved with motor function, and we can categorize CP by which of these is affected.

Spastic cerebral palsy results from damage to the pyramidal tract, which originates from the motor cortex. Non-spastic CP results from damage to the extrapyramidal tract, which originates from the basal ganglia and cerebellum.

70-80% of CP cases are of the spastic type, and patients typically present with spasticity and rigidity. Spasticity is a velocity-dependent increase in muscle tone that leads to resistance during movement, while rigidity is an increase in tone at baseline. Spastic CP can be further characterized based on which body parts are affected.

- Spastic hemiplegia occurs when just one side of the body is affected, usually with more involvement of the legs than arms.
- Spastic diplegia occurs when there is involvement of both lower extremities.
- Spastic quadriplegia occurs when there is involvement of all extremities and the trunk.
- Rarely, children have spastic monoplegia, which occurs when just one limb is involved.

Children with spastic CP have muscle imbalance in their limbs and stronger muscle groups dominate, leading to certain characteristic positions. Since the quads are stronger than the hamstrings, plantar flexors stronger than dorsiflexors, and biceps stronger than triceps, CP patients often have leg extension (also known as scissoring), toe flexion and arm flexion. Eventually, this imbalance can lead to joint contractures or scoliosis. Involvement of the nerves supplying the tongue, mouth or pharynx muscles lead to difficulty swallowing and speaking.

Moving on, non-spastic CP results from damage to the extrapyramidal tracts, which originate from the basal ganglia and cerebellum. Depending on which of these is affected, non-spastic CP can be further classified into dyskinetic and ataxic CP.

Dyskinetic CP results from damage to the basal ganglia, and makes up 15% of total cases. These children present with involuntary movements that are choreoathetoid or dystonic in nature. Choreoathetoid movements occur primarily in the extremities, and can involve rapid, irregular contractions of smaller muscles in the face or digits, or dance-like writhing movements of the limbs. Dystonic movements are involuntary movements of the trunk and result in a fixed, twisted posture.

Finally, ataxic CP results from damage to the cerebellum, and makes up only 5% of CP cases. Since the cerebellum is primarily involved in balance and coordination, loss of function here leads to symptoms like a wide-based gait or difficulty with fine motor skills like writing. Ataxic CP does not usually involve involuntary movements.

Often, there is damage to multiple areas of the brain, leading to mixed CP.

**Clinical Presentation: Severity**

Now that we have talked a bit about how to classify CP based on presentation, we can discuss how we classify it based on severity. A commonly used classification system for children 2 to 18 years old is the Gross Motor Function Classification System, or GMFCS. This system stratifies CP patients into 5 levels depending on what they can do and their level of dependence on mobility assistance devices.

Very broadly, a child in level 1 can walk without limitations, one in level 2 can walk with some limitations, one in level 3 can walk with a hand-held mobility device such as a cane or crutches, one in level 4 requires a wheelchair but can move by themselves by using their arms, legs or a joystick, and one in level 5 requires a wheelchair and need someone else to push them. It may help to give more specific examples of all these levels.

- A child in level 1 is pretty similar to a typical child, but might have some difficulty with advanced motor abilities like playing coordinated sports.
- A child in level 2 might be able to walk and run like other kids, but might need to take more frequent breaks or use the railing to climb stairs. They may require

crutches, a cane or walker when first learning to walk, but usually don't need one after the age of 4.
- A child in level 3 will continue to need a hand-held mobility device, but can still sit without support and are usually able to stand independently.
- A child in level 4 usually needs a manual or powered wheelchair to mobilize, but they can move themselves. They also often need support while sitting and cannot stand independently.
- Finally, a child in level 5 usually has limited head and trunk control, so they need someone else to move them in a wheelchair. Sometimes, they can learn to move themselves if they have a powered wheelchair with a joystick.

***Diagnosis***

Diagnosing CP depends on obtaining a good history and physical examination. But when do you suspect CP in a child? Some important red flags to keep in mind include delays in reaching motor milestones, hypotonia for the first 6 months of life followed by hypertonia, hand preference prior to the age of 1, asymmetric crawling and early standing due to rigidity.

You can begin your history by asking about possible etiologies of neurological damage in a systematic fashion. Be sure to ask about maternal prenatal screening, and infections during the pregnancy. Ask whether any resuscitation was required at delivery, whether the baby stayed in the NICU, or if they were given any antibiotics. Make sure you ask about any traumas or falls after birth.

Next, you can investigate the child's motor development. Was he or she able to sit, stand and walk at the proper ages? Is the child able to complete fine motor movements? Does the child have difficulty with balance or favor one side? Are there any other neurological symptoms like difficulties with vision, hearing or seizures?

A physical exam relevant to CP focuses on a thorough neurological and MSK exam. It is particularly important to examine tone and reflexes. Hypertonia and hyperreflexia are associated with upper motor neuron loss in the pyramidal tracts, or spastic CP, whereas hypotonia and hyporeflexia would be more consistent with non-spastic CP. Persistence of primitive reflexes, such as the Moro, stepping or asymmetrical tonic neck reflexes, suggest a lesion in the pyramidal tract. In older kids, be sure to look for any contractures, hip dislocations, and scoliosis that may have developed over time.

Investigations for a diagnosis of cerebral palsy would mainly focus on looking for a cause of the motor deficits. Head imaging, preferably an MRI compared to a CT, can assess for structural anomalies, lesions from hemorrhages or trauma, and infections. Additionally, an MRI may reveal periventricular leukomalacia (PVL) which refers to inflammatory damage to the white matter surrounding the lateral ventricles. It is especially common in premature and low birth weight infants and frequently causes spastic cerebral palsy.

In general, the investigations you choose to do will depend on what you suspect is the underlying etiology.

***Differential diagnosis***

So let's go back to Rosie. Remember, Rosie is a 16 month old child who was born prematurely at 32 weeks. She has delayed motor milestones, and lower limb hypertonia. You suspect she may have spastic diplegic CP, but what else should you be considering in your differential?

There are certain findings on history or physical that can point you towards a diagnosis other than CP. If the symptoms are present for well-defined and limited periods of time, they could indicate a seizure disorder. If they were limited to one episode, they could represent an acute stroke. If the child deteriorates during periods of fasting, stress, or illness, there may be an inborn error of metabolism. If there is progressive worsening of symptoms or developmental regression, consider a tumor, or neurodegenerative conditions because remember, CP is non-progressive.

***Management***

Rosie's story does not sound like any of these other conditions, so you tell your preceptor that you wish to order an MRI of her head. The results come back showing periventricular leukomalacia, confirming your diagnosis. At the next visit, Rosie's parents ask you what the treatment options are. What do you tell them?

Unfortunately, there is no treatment for CP. That is, the neurological insult that results in the motor deficits cannot be reversed, although the injury is static over time. The goal of management for CP is to increase how well the child can function in day-to-day life, with those deficits. It is always best to involve a multidisciplinary team as early as possible, with a pediatrician or family physician to help coordinate care

Many different subspecialties will be involved in care of a child with CP. Neurologists should monitor tone and other developmental delays, while orthopedic surgeons can increase range of motion through orthotics and surgical release of muscle tendons. Otolaryngology can help manage sialorrhea or increased secretions, and ophthalmology can screen for potential visual deficits.

A team will also include many other kinds of healthcare practitioners. Occupational therapists use goal-oriented activities to improve function, and assess for risk of aspiration. Physiotherapists help work on mobility and reduce muscle tone. Dieticians address any feeding concerns and can suggest gavage tube feeds. Speech language therapists manage oromotor challenges, and can offer assistive device programs to optimize communication abilities.

Pharmacologically, there are certain medications that can reduce hypertonia in spastic and mixed CP. Botox injections into contracted muscles inhibit presynaptic release of

acetylcholine at the neuromuscular junction and relax the muscle. However, these need to be repeated every 3-8 months and into several areas of the muscle. Other oral medications, such as benzodiazepines, dantrolene and baclofen can be used to treat spasticity, but are less effective and have side effects. Benzodiazepines work by increasing neuronal inhibition by GABA, dantrolene works by inhibiting the influx of calcium that is necessary for muscle contraction, and baclofen is a GABA analog.

Lastly, an important part of managing CP is preventative care. Because children with CP have muscle imbalance, they are at risk of hip dislocations, scoliosis and contractures. Thus, vigilant routine monitoring should be arranged by a pediatrician or family doctor.

### *Prognosis*

So, with the proper management, what is the prognosis of children with cerebral palsy? As we've discussed, CP is not a progressive disease and the majority of patients survive into adulthood. However, survival is related to degree of impairment and co-morbidities; high-functioning adults have a similar life expectancy as the general population, but children who are severely affected and at level 5 on the GMFCS tend to have a substantially shorter life span.

### *Summary*

We've reached the end of this PedsCases podcast, so it's time for a quick recap. Here are some take-home points to remember:

1. Cerebral palsy describes non-progressive motor disorders that result from pre-, peri-, and post-natal insults to the developing brain.
2. CP is classified as spastic or non-spastic. Non-spastic CP can further be divided into dyskinetic and ataxic CP.
3. The severity of CP is usually based on the Gross Motor Function Classification System, or GMFCS.
4. Red flags that can point towards CP include delay in reaching motor milestones, asymmetric crawling, early hand preference, early standing, and hypotonia followed by hypertonia.
5. Diagnosis of CP comes from a thorough history and physical examination, but investigations can be used to confirm the underlying neurological damage.
6. Treatment is multidisciplinary and focuses on supportive management. Regular medical surveillance is necessary for preventative care.

Thank you for listening to this PedsCases podcast. Stay tuned for more!

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