Chiari Malformation

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What is Chiari Malformation?

Chiari Malformation (CM) is a condition where the lower part of the brain (the cerebellum) protrudes down into the spinal canal. CM is divided into 4 classifications that range from mild to severe. It is not uncommon for children who have CMs to also suffer from hydrocephalus (excess fluid in the brain), spina bifida (opening of the spine), syringomyelia (excess fluid in the spinal cord), or some inherited conditions. (i.e. dwarfism)

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What causes Chiari Malformation?

The cause of CM is unknown. However, there are several theories about why it occurs. One theory suggests that, in some individuals, the base of the skull is too small and the cerebellar region of the brain is forced downward. While another theory states that overgrowth of the cerebellar region causes compression in that area of the brain.

What are the symptoms of Chiari Malformation?
Symptoms of CM often begin during infancy, although they may be delayed until adolescence or adulthood. Symptoms usually include vomiting, muscle weakness in the head and face, difficulty swallowing and varying degrees of mental impairment. Other symptoms can involve:

- Headache
- Neck pain
- Progressive scoliosis (curvature of the spine)
- Difficulty with balance and coordination
- Problems with fine motor skills
- Vision problems including blurred or double vision and hypersensitivity to bright light
- Tinnitus (buzzing in the ear)
- Hearing loss
- Voice alteration or paralysis
- Frequent gagging and/or choking
- Sleep apnea (briefly ceasing to breathe while sleeping)
- Paralysis or weakness of the arms and legs
- Spasticity (abnormally high muscle tone or tightness).

How is Chiari Malformation diagnosed?
Magnetic Resonance Imaging (MRI) is a safe, non-evasive, and painless way to scan the brain and is currently the most reliable way to diagnose Chiari Malformation. Since the introduction of the MRI, the diagnosed incidence of Chiari Malformation has risen.

How common is Chiari Malformation?
Approximately 1% of infants are born with malformations of the cervico-medullary junction, the connection between the spine and the brain. Not all of these malformations are Chiari Malformations, although CM is the most common.

How is Chiari Malformation treated?
Children who suffer from CM may require surgery to repair myelomeningocele, a protrusion from the spinal cord. Hydrocephalus can be treated with an implanted shunt to relieve pressure from fluid on the brain.

What is the Prognosis after surgery?
Most children who undergo surgical treatment experience a reduction in their symptoms. Infants with more severe cases may have life-threatening complications.

Are there any other names for Chiari Malformation?
Chiari Malformation is also known as herniation of the cerebral tonsils, cerebellar ectopia, hindbrain herniation, and Arnold-Chiari malformations.

Is Chiari Malformation hereditary?
In a few cases, more than one member of a family has been documented to have CM. It is possible that CM could be inherited but there is no conclusive evidence to support this. Any close relative of a child suffering from CM who is experiencing symptoms should seek genetic counseling.

Fact Sheet by:

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