

What is Chiari malformation?

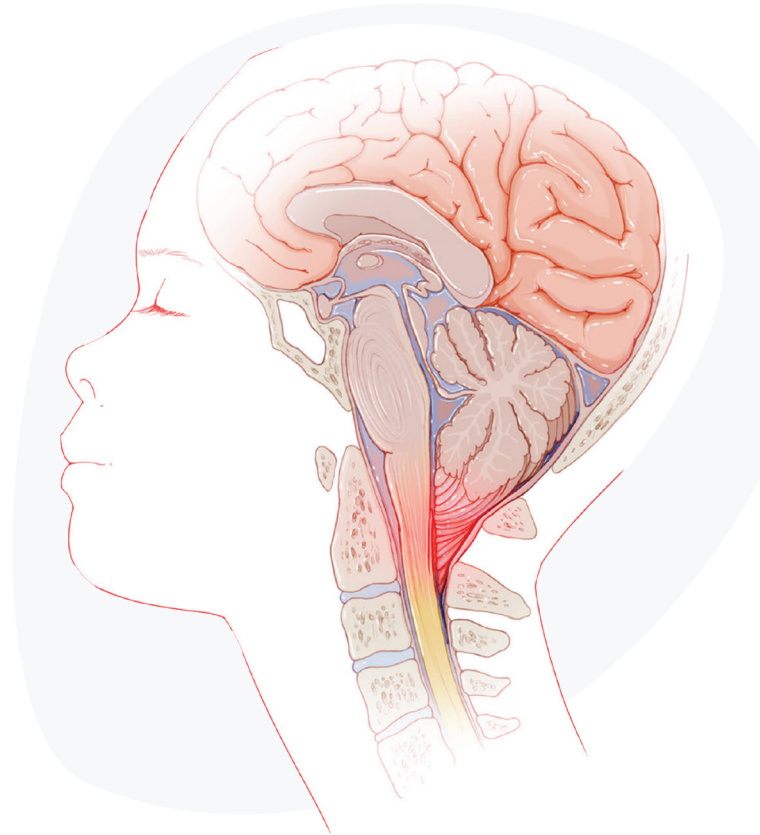
A Chiari malformation is a condition in which the back of the brain (hindbrain) pokes through the base of the skull and into the spinal canal (where the spinal cord is located). This crowding in the hindbrain may lead to a disruption of cerebrospinal fluid (CSF) flow and/or compression of the brain and spinal cord.

There are other conditions that are associated with Chiari malformations:

- Syrxinx/Syringomyelia
- Tethered spinal cord

A syrinx is a buildup of CSF within the spinal cord. It is developed due to the altered CSF flow caused from the chiari malformation. The spinal fluid cannot circulate and absorb properly, so it builds up abnormally in the spinal cord itself, potentially leading to neurological symptoms.

A tethered cord occurs when the spinal cord is attached to tissue that is not elastic (stretchy). The neurological symptoms present as a result of stretching of the spinal cord as the person grows.



What are the symptoms?

The most common presenting symptom of Chiari malformations is headache.

- Headaches are worse in the morning upon awakening
- Headaches are most often located in the back of the head (occipital region) and may radiate behind the eyes (retro-orbital pain) and/or down into the neck
- Neck pain—headaches are worsened by activities that increase the pressure within the brain, such as coughing, sneezing, bending over or straining.
- Difficulty swallowing/choking—difficulty walking
- Central sleep apnea (stop breathing during sleep)
- Balance and coordination issues

Symptoms of a syrinx

- Numbness/tingling in arms/legs—back pain

Symptoms of tethered cord

- Difficulty walking
- Numbness/tingling/weakness in legs—bowel and/or urinary incontinence
- Pain radiating down the legs

The symptoms listed are the most commonly associated ones. Chiari malformations are very complex and individuals may experience a multitude of symptoms.

Diagnosis

If the primary clinician is concerned for the possibility of a Chiari malformation, they may order an MRI of the brain and/or spine. A Chiari malformation may be identified on a CT scan of the brain. However, it is not as evident as it is on an MRI and the diagnosis may be missed. If a Chiari malformation is detected, your child will be referred to a pediatric neurosurgeon.

Depending on the symptoms, further tests may be needed such as a swallow study or a test for sleep apnea.

