

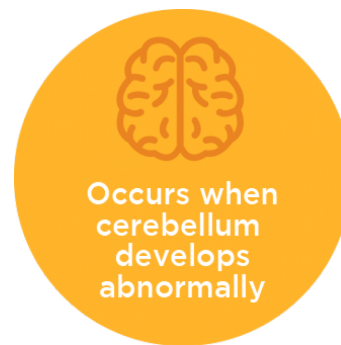
Chiari Malformation

What Is Chiari Malformation?

A Chiari (kee-AH-ree) malformation occurs when the back of the brain develops abnormally and pushes into the spinal canal.

The downward movement of the brain tissue puts pressure on the cerebellum. This pressure limits the flow of the protective liquid (cerebrospinal fluid) that surrounds the brain and spinal cord.

The pressure on the cerebellum can prevent parts of the brain and spine from functioning properly. It sometimes causes problems with balance and coordination.



What Causes Chiari Malformation?

Most children who have Chiari malformation are born with the condition. It often occurs along with conditions such as hydrocephalus, craniosynostosis, [Ehlers-Danlos syndrome](#) and Klippel-Feil syndrome.

The most common type of Chiari malformation is caused by genetic changes (mutations) or a lack of certain vitamins and nutrients in a mother's diet. Secondary Chiari malformation is typically caused by an injury, exposure to toxic substances, or an infection.

Chiari Malformation Types

Chiari malformation types get categorized based on which parts of the brain push down into the spinal canal.

- Type I: The cerebellum pushes through the bottom of the skull into the upper spinal canal. Symptoms might not appear until adolescence or early adulthood. Chiari 1 malformation is the most common type in children.
- Type II (also known as Arnold-Chiari malformation): A larger part of the cerebellum and part of the brain stem push into the spinal canal. Chiari malformation type 2 is almost always associated with myelomeningocele, a severe form of spina bifida.

- Type III: Parts of the cerebellum and brain stem push into the spinal cord, sometimes causing a pouch-like structure to form on the back of the neck. Type III often causes life-threatening complications and is very rare.
- Type IV: Also called cerebellar hypoplasia. This type is very rare and occurs when the cerebellum does not completely develop. It is the most severe form of the condition. Most infants born with Type IV Chiari malformation do not survive.

Chiari Malformation Symptoms and Effects

Some children who have Chiari malformation don't experience any symptoms. Or, they might not experience symptoms until later in adolescence or even adulthood.

Symptoms of Type I Chiari malformation can include:

- Headaches.
- Pain in the arms, legs or neck.
- Numbness or weakness in the hands and arms.
- Unsteadiness when walking.
- Dizziness.
- Vomiting.
- Loss of the ability to sense temperature.
- Curvature of the spine (scoliosis).
- Fluid collection in the spinal cord (syringomyelia).

Some younger children and infants might have difficulty drinking or swallowing.

Symptoms of Type II Chiari malformation are similar to those found in Type I. However, Type II symptoms are more severe and usually happen at a younger age.

Symptoms of Type II Chiari malformation include:

- Abnormally slow or noisy breathing.
- Sleep apnea.
- Accidentally breathing fluids or saliva into the lungs.
- Numbness or weakness in nerves of the throat, face or tongue.

Chiari Malformation Diagnosis and Treatment

Some Chiari malformations don't cause symptoms, and therefore don't require treatment. However, others cause a wide range of symptoms. The Gillette Children's Specialty Healthcare team of experts works with specialists to

diagnose the condition in your child.

Next, we partner closely with you to find the best treatment plan. Because Chiari malformation affects each child differently, we offer a wide range of tests, treatments and services for affected children, teens and adults. We start with a review of your child's medical history. Some of the tests we might use to diagnose Chiari malformation include an [MRI](#) scan of the brain and/or [CT scan](#).

Treatments for Chiari malformation include:

Medication

Pain-relieving or anti-inflammatory medicines can help manage symptoms such as frequent headaches and pain.

Surgery

Our pediatric neurosurgeons work closely with you to decide whether your child could benefit from surgery.

The Chiari malformation surgery (also known as posterior fossa decompression) increases space for the cerebellum and brain stem. This procedure relieves pressure on the brain and lets cerebrospinal fluid flow normally. If your child also has hydrocephalus—especially in combination with spina bifida and Type II Chiari malformation—shunt surgery might be required before Chiari decompression.

Integrated Care

Treatment for the complex symptoms and effects of Chiari malformation require a team approach. At Gillette, your family will work with experts in a wide range of specialties and services that might include:

- [Orthotics, Prosthetics and Seating](#).
- [Chaplaincy](#).
- [Child life](#).
- [Gastroenterology](#).
- [Medical genetics and genetic counseling](#).
- [Neurodiagnostics](#).
- [Neurology](#).
- [Neuropsychology](#).
- [Neurosurgery](#).
- [Psychology](#).
- [Radiology and imaging](#).
- [Rehabilitation medicine](#).
- [Rehabilitation therapies](#).

- [Physical therapy.](#)
- [Social work.](#)
- [Therapeutic recreation.](#)
- [Urology.](#)

Our multidisciplinary team of experts is here to support your family, answer questions, and help your child feel their best every step of the way.

Explore Chiari Malformation Resources

- Find education, research and advocacy resources at the [Chiari & Syringomyelia Foundation, Inc.](#)

[Make An Appointment](#) [651-290-8707](#) [Refer a Patient](#) [651-325-2200](#)

This information is for educational purposes only. It is not intended to replace the advice of your health care providers. If you have any questions, talk with your doctor or others on your health care team.

If you are a Gillette patient with urgent questions or concerns, please contact Telehealth Nursing at [651-229-3890](#).