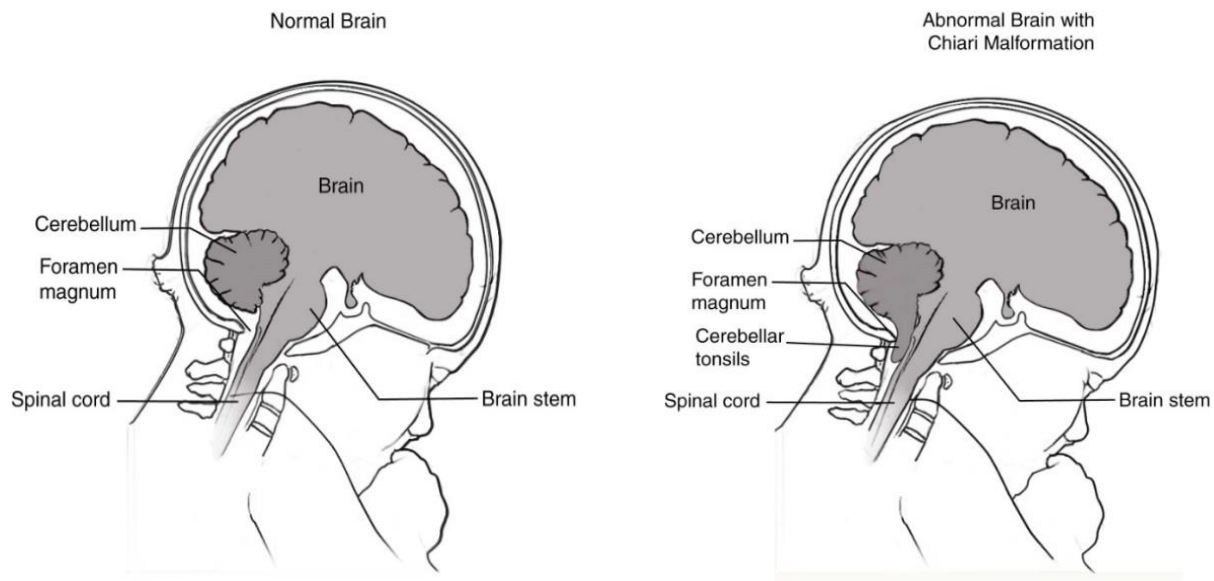


What is Chiari malformation?

Chiari (“kee-ar-ee”) malformation is a condition in which brain tissue drops into a large opening at the base of the skull. The opening is called the foramen magnum. When this happens:

- It creates extra pressure in the brain and spinal cord.
- It blocks the flow of the clear liquid that surrounds and cushions the brain and spinal cord. The liquid is called cerebrospinal fluid or CSF.



What are the types of Chiari malformations?

There are 4 types of Chiari malformations. Your child’s doctor will talk with you about your child’s type. Some general information about each type is listed below.

Type 1 is the most common form of Chiari

- With this type of Chiari:
 - The bottom of the skull and the upper part of the spine do not grow properly. There is not enough space for the brain to get larger.
 - The cerebellar tonsils push into the foramen magnum and the spinal canal. The tonsils are part of the cerebellum, which is the lower part of the brain at the base of the skull.
- A person may not have symptoms at first. Symptoms happen as extra pressure builds in the brain. This is often in late childhood or adulthood since the brain does not reach its full size until then.

In case of an urgent concern or emergency, call 911 or go to the nearest emergency department right away.

Chiari malformation, continued

Type 2 is also called classic Chiari malformation or Arnold-Chiari malformation

- This type of Chiari causes more brain tissue to push into the spinal canal.
- It is more common in children.
- It often happens along with a myelomeningocele, which is a type of spina bifida.
 - Spina bifida happens when the backbone and spinal canal do not close before birth.
 - It can cause part of the spinal cord and tissues that surround it to form outside of the body.

Type 3 is the most severe form of Chiari

- With this type of Chiari:
 - The cerebellum and the brainstem push through the foramen magnum and into the spinal canal. The brainstem is the lower part of the brain that connects to the spinal cord.
 - The covering of the brain or spinal cord can push through an abnormal opening in the back of the skull.
- It is often found on an ultrasound during pregnancy or at birth.

Type 4 is the rarest form of Chiari

- With this type of Chiari:
 - The back of the brain itself is underdeveloped or incomplete (cerebellar hypoplasia).
 - The cerebellum is in its normal position, but parts of it are missing.
 - Parts of the spinal cord and skull may be visible.
- It can be found by ultrasound during pregnancy or at birth

What causes Chiari malformation?

There are 2 possible causes for Chiari malformations.

- Primary or congenital Chiari malformation:
 - Can be caused by changes in the structure of the brain and spinal cord that take place during pregnancy.
 - Might happen as a result of genetic problems (unexpected changes in the genes as the fetus develops) or from the lack of proper vitamins or nutrients during pregnancy.
- Acquired or secondary Chiari malformation:
 - Is caused when large amounts of CSF are drained from the spinal area.
 - Is often due to severe injury, exposure to harmful substances or infection.

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Chiari malformation, continued

What are the possible symptoms?

Symptoms of Chiari malformation depend on the type of defect and how severe it is. They can also differ by age and by child. Some common symptoms by age are listed below.

Child's age	Symptoms
Babies and young children	<ul style="list-style-type: none">• Weak upper body• Problems with feeding or swallowing• Rapid, back and forth eye movements (nystagmus)• Excess drooling• Weak cry• Vomiting or gagging• Stridor or high pitched sound when breathing• Stiff neck• Breathing problems• Problems walking and getting around
Older children and adults	<ul style="list-style-type: none">• Severe headache (pain is most often at the back of the head)• Headache that worsens with coughing, sneezing, straining and bending over• Hoarseness or trouble speaking• Neck pain• Weak upper body or problems with fine movements• Numbness or tingling in the arms or legs• Dizziness• Trouble swallowing• Vision or hearing problems• Vomiting• Breathing problems• Slurred speech• Changes in balance• Problems walking and getting around

What medical tests could my child have?

Some tests that can help the doctor find out more include:

- Computed tomography (CT scan)
 - Uses X-rays and a computer to show pictures of the head and spine
 - Displays cross-sectional images, like slices, of the head and spine
- Magnetic resonance imaging (MRI) with CSF flow study
 - Uses a magnetic field and radio waves to create an even more detailed view of the head and spine

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Chiari malformation, continued

What is the treatment?

Your child's doctor will talk with you about specific care for your child. Some guidelines to follow may include:

- Medicine to control pain and swelling
- Surgery to remove a small section of bone in the back of the skull. This helps to relieve pressure and create more space for the brain to allow for flow of CSF. Sometimes, more than one surgery is needed.

Will my child need any follow-up treatment?

Your child's doctor will talk with you about what is needed for your child. Many times, follow-up includes:

- Routine follow-up exams with your child's neurosurgeon.
- MRI scans to check surgery outcomes.
- Follow-up with other specialists, if needed for your child.

This teaching sheet contains general information only. Talk with your child's doctor or a member of your child's healthcare team about specific care of your child.

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