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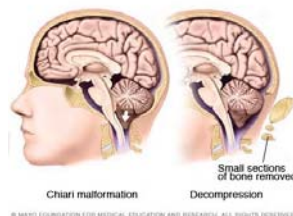
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Chiari malformation (kee-AH-ree mal-for-MAY-shun) is a condition in which brain tissue extends into your spinal canal. It occurs when part of your skull is abnormally small or misshapen, pressing on your brain and forcing it downward.

Chiari malformation is uncommon, but increased use of imaging tests have led to more frequent diagnoses.

Doctors categorize Chiari malformation into three types, depending on the anatomy of the brain tissue that is displaced into the spinal canal, and whether developmental abnormalities of the brain or spine are present.

Chiari malformation type I develops as the skull and brain are growing. As a result, signs and symptoms may not occur until late childhood or adulthood. The pediatric forms, Chiari malformation type II and type III, are present at birth (congenital).



Chiari malformation

In Chiari malformation surgery, doctors remove a small section of bone at the back of your skull to make room for part of your brain (cerebellum) and relieve pressure on your brainstem, cerebellum and spinal cord.

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Treatment of Chiari malformation depends on the form, severity and associated symptoms. Regular monitoring, medications and surgery are treatment options. In some cases, no treatment is needed.

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Symptoms

Many people with Chiari malformation have no signs or symptoms and don't need treatment. Their condition is detected only when tests are performed for unrelated disorders. However, depending on the type and severity, Chiari malformation can cause a number of problems.

The more common types of Chiari malformation are:

- **Type I**
- **Type II**

In Chiari malformation type I, signs and symptoms usually appear during late childhood or adulthood.

Chiari malformation type II is usually noted by ultrasound during pregnancy. It may also be diagnosed after birth or in early infancy.

Although these types are less serious than the more rare pediatric form, type III, signs and symptoms still can be life disrupting.

Chiari malformation type I

Headaches, often severe, are the classic symptom of Chiari malformation. They generally occur after sudden coughing, sneezing or straining. People with Chiari malformation type I can also experience:

- Neck pain
- Unsteady gait (problems with balance)
- Poor hand coordination (fine motor skills)

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- Numbness and tingling of the hands and feet
- Dizziness
- Difficulty swallowing, sometimes accompanied by gagging, choking and vomiting
- Vision problems (blurred or double vision)
- Speech problems, such as hoarseness

Less often, people with Chiari malformation may experience:

- Ringing or buzzing in the ears (tinnitus)
- Weakness
- Slow heart rhythm
- Curvature of the spine (scoliosis) related to spinal cord impairment
- Abnormal breathing, such as central sleep apnea, characterized by periods of breathing cessation during sleep

Chiari malformation type II

In Chiari malformation type II, a greater amount of tissue extends into the spinal canal compared with Chiari malformation type I.

The signs and symptoms can include those related to a form of spina bifida called myelomeningocele that nearly always accompanies Chiari malformation type II. In myelomeningocele, the backbone and the spinal canal haven't closed properly before birth.

Symptoms may include:

- Changes in breathing pattern
- Swallowing problems, such as gagging
- Quick downward eye movements
- Weakness in arms

Chiari malformation type III

In one of the most severe types of the condition, Chiari malformation type III, a portion of the lower back part of the brain (cerebellum) or the brainstem extends through an abnormal opening in the back of the skull.

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This form of Chiari malformation is diagnosed at birth or by an ultrasound during pregnancy.

This type of Chiari malformation has a higher mortality rate and may also cause neurological problems.

When to see a doctor

If you or your child has any of the signs and symptoms that may be associated with Chiari malformation, see your doctor for an evaluation.

Because many symptoms of Chiari malformation can also be associated with other disorders, a thorough medical evaluation is important.

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Causes

Chiari malformation type I occurs when the section of your skull containing a part of your brain (cerebellum) is too small or is deformed, thus putting pressure on and crowding your brain. The lower part, or tonsils, of the cerebellum are displaced into your upper spinal canal.

Chiari malformation type II is nearly always associated with a form of spina bifida called myelomeningocele.

When the cerebellum is pushed into your upper spinal canal, it can interfere with the normal flow of cerebrospinal fluid that protects your brain and spinal cord.

This impaired circulation of cerebrospinal fluid can lead to the blockage of signals transmitted from your brain to your body, or to a buildup of spinal fluid in the brain or spinal cord.

Alternatively, the pressure from the cerebellum upon the spinal cord or lower brainstem can cause neurological signs or symptoms.

Risk factors

There's some evidence that Chiari malformation runs in some families. However, research into a possible hereditary component is still in its early phase.

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Complications

In some people, Chiari malformation can become a progressive disorder and lead to serious complications. In others, there may be no associated symptoms, and no intervention is necessary. The complications associated with this condition include:

- **Hydrocephalus.** An accumulation of excess fluid within your brain (hydrocephalus) may require placement of a flexible tube (shunt) to divert and drain the cerebrospinal fluid to another area of your body.
- **Spina bifida.** Spina bifida, a condition in which your spinal cord or its covering isn't fully developed, may occur in Chiari malformation. Part of the spinal cord is exposed, which can cause serious conditions such as paralysis. People with Chiari malformation type II usually have a form of spina bifida called myelomeningocele.
- **Syringomyelia.** Some people with Chiari malformation also develop a condition called syringomyelia, in which a cavity or cyst (syrinx) forms within the spinal column.
- **Tethered cord syndrome.** In this condition, your spinal cord attaches to your spine and causes your spinal cord to stretch. This can cause serious nerve and muscle damage in your lower body.

By Mayo Clinic Staff

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