

Chiari Malformation Type I



What is a Chiari malformation?

A Chiari malformation is a structural defect in which a part of the brain (the cerebellum) at the back of the skull bulges through a normal opening in the skull where it joins the spinal canal. This puts pressure on parts of the brain and spinal cord and can cause mild to severe symptoms. In most cases, the problem is present at birth (congenital).

There are several types of Chiari malformations, but type I is the most common. In type I, the cerebellum bulges through the normal opening at the base of the skull. This type is most often congenital. It is also called primary Chiari malformation type I. But it often isn't found until a person is a teen or young adult.

In rare cases, this type may also develop later in life. This is known as acquired or secondary Chiari malformation type I.

What causes a Chiari malformation type I?

The exact cause of a congenital Chiari malformation type I isn't known. A problem during fetal growth may cause the defect. It may be caused by contact with harmful substances during pregnancy. Or it may be linked with genetic problems that run in families.

An acquired Chiari malformation type I happens to a person after birth. It's caused by excess leaking of spinal fluid from the lower back (lumbar) or chest (thoracic) areas of the spine. This can happen because of traumatic injury, disease, or an infection.

What are the symptoms of a Chiari malformation type I?

You may not have any symptoms. Or symptoms may develop slowly over time. Most people don't have symptoms until they are teens or young adults.

The most common symptoms are headaches or pain in the back of the head or neck. The headaches and pain are made worse by coughing, laughing, or sneezing.

Other symptoms of a Chiari malformation type I are:

- Hoarseness or trouble speaking
- Trouble swallowing
- Excessive drooling, gagging, or vomiting
- Rapid, back and forth eye movements (nystagmus), or double or blurred vision
- Periods of not breathing during sleep (sleep apnea)
- Weakness, dizziness, or abnormal movements
- Trouble with hearing or balance
- Abnormal reflexes
- Abnormal shape of the spine (scoliosis)

You may also have a pocket of spinal fluid in the spinal cord or brain stem. This is called a syrinx. A syrinx can cause trouble walking or pain in the arms or legs. It can also lead a person to develop weakness in the arms and legs.

How is a Chiari malformation type I diagnosed?

If you have no symptoms, the problem may be found when you have imaging tests for other reasons. There are currently no tests available to determine if a baby will be born with Chiari malformation. If you have symptoms, your healthcare provider will ask about your health history and give you a physical exam. They may refer you to a specialist.

Imaging tests are done to find a Chiari malformation type I. You may have one or more of these tests:

- **MRI.** This test is the one most often used to diagnose Chiari malformations. It uses large magnets and a computer to make detailed pictures of the inside of the body without using X-rays.
- **CT scan.** This test uses a series of X-rays and a computer to create detailed pictures of the inside of the body. A CT scan is more detailed than a regular X-ray.

How is a Chiari malformation type I treated?

You may be treated by a neurologist or neurosurgeon. These are experts in brain and spinal cord problems. Treatment will depend on your symptoms, age, and general health. It will also depend on how severe the condition is.

- **With no symptoms.** Your health may be watched closely. This may include frequent physical exams and MRI tests.
- **With symptoms.** Your healthcare provider may prescribe medicines to reduce pain. Or they may choose surgery. This is done to relieve pressure on the brain or restore the flow of spinal fluid.
- **With few or no symptoms, but a syrinx.** Your healthcare provider may suggest close watching of the defect with a special type of MRI called cine phase contrast. This test looks at the flow of spinal fluid. It also looks at areas where the fluid is blocked. You may need surgery, based on the MRI results or if symptoms get worse.
- **With signs of sleep apnea.** You may need a sleep study if you have sleep apnea. Sleep apnea means that you stop and start breathing during sleep. A sleep study can also help your healthcare provider decide if you need other treatment.

What other health problems are caused by a Chiari malformation type I?

These health problems can include:

- Long-term pain
- A pocket of spinal fluid (syrinx) in the spinal cord or brain stem. This forms over time.
- Lifelong damage to muscles or nerves
- An inability to move your arms or legs because the muscles no longer work (paralysis)

Carefully watching for changes in your health can help prevent these problems. This helps to make sure that treatment is done early.

In addition to the physical problems, the emotional and financial stress caused by a Chiari malformation diagnosis can be hard for you and your family members. Ask your healthcare provider for community support resources or professional counselors if you need help managing the stress caused by this diagnosis.

When should I call my healthcare provider?

Call your healthcare provider if you notice any changes or new symptoms. Also if you notice signs of depression or problems with:

- Breathing

- Swallowing
- Eating
- Speaking
- Walking or moving

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