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Understanding Chiari Type II Malformation

October 2024

Chiari type II malformation is often seen in people with myelomeningocele (also called open Spina Bifida). About 8 out of 10 people with spina bifida have this brain difference, which can cause fluid to build up in the brain (called hydrocephalus). This usually needs medical treatment. There are other types of Chiari malformation, but they are different and don't apply to Chiari type II.

What is a Chiari Type II Malformation?

Chiari type II malformation is a condition that some babies are born with. It affects the lower part of the brain, called the cerebellum, and the brain stem. In this condition, parts of the brain are pushed down into the top of the spinal canal through an opening at the bottom of the skull.

This can block the normal flow of fluid (called cerebrospinal fluid) that moves around the brain and spinal cord. When this fluid gets blocked, it can build up inside the brain. This is called hydrocephalus. Hydrocephalus can cause pressure in the brain and make the spaces in the brain (called ventricles) get larger.

Most people with myelomeningocele (a type of open spina bifida) have some form of Chiari type II malformation. In some people, this condition causes problems (symptoms), but in others, it does not. About 1 in 3 people with Chiari type II malformation show symptoms. Sometimes symptoms show up later, like during the teenage years or early adulthood. The more severe the brain changes are, the more likely it is that a person will have symptoms.

Common symptoms of Chiari type II malformation:

- Changes in breathing
- Trouble swallowing
- Eyes that move downward
- Weakness in the arms

How Doctors Check for Chiari Type II Problems

Doctors use different tests to check how the brain stem and spine are working. These tests can be a little different depending on the hospital.

An MRI (magnetic resonance imaging) is the best way to look at the brain and upper spine. It takes very detailed pictures and is safe for most people. The MRI can show if part of the brain is out of place, if there are cysts, or if something looks wrong inside the spinal cord. It can also show how the fluid (called cerebrospinal fluid or CSF) is moving around the brain and spine.

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Other tests can check how well the brain and spine are working. These might include:

- A sleep study (polysomnography) to check breathing during sleep
- Tests that check how signals travel from the arms and legs to the brain
- A hearing test to see how the brain responds to sound
- A visit with an ENT (ear, nose, and throat doctor) to check swallowing and how the vocal cords are working

These tests help doctors understand if Chiari type II is causing any problems and how to treat it.

Treating Chiari Type II Malformation

For children who may have symptoms from Chiari type II malformation, the first and most important step is to make sure there is normal pressure in the brain before doing anything else.

In newborns with myelomeningocele, this means treating hydrocephalus (extra fluid in the brain) first. This is usually done by placing a shunt or using another method to drain the fluid.

If an older child starts having symptoms, doctors will first check the shunt to make sure it's working correctly. This step is very important. If there's any concern that the shunt isn't working, surgery may be needed to fix or replace it. Brain scans alone may not always show if the shunt is working properly, so doctors may need to look at it directly.

Once doctors are sure that the brain pressure is normal and the shunt is working, they check to see how serious the symptoms are. Most children with Chiari type II on brain scans do not need surgery, especially if their symptoms are mild or not getting worse. But if the symptoms are very serious or getting worse over time, surgery may be needed.

Surgery for Chiari Type II Malformation

The goal of Chiari type II decompression surgery is to remove some bone to make more space and lower the pressure on the brain. This also helps the fluid around the brain and spine (called cerebrospinal fluid or CSF) flow more normally.

This surgery is usually safe when done by a doctor who is specially trained to operate on children's brains. These doctors are called pediatric neurosurgeons. Because the surgery is complex, it's important that the doctor has a lot of experience.

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Sometimes, instead of doing surgery right away, doctors may choose to watch and wait, especially if the symptoms are not getting worse. Many people with myelomeningocele do not get worse quickly. Doctors may have different opinions about when surgery is needed. Usually, if the symptoms seem to be clearly caused by Chiari type II, surgery is more likely to be recommended.

It's very important to check and fix the shunt first before deciding on surgery. Regular check-ups are also needed to watch for any changes.

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This information does not constitute medical advice for any individual. As specific cases may vary from the general information presented here, SBA advises readers to consult a qualified medical or other professional on an individual basis.