

## **Chiari II Malformation**

(Arnold Chiari II)

### **What is Chiari II (2) malformation?**

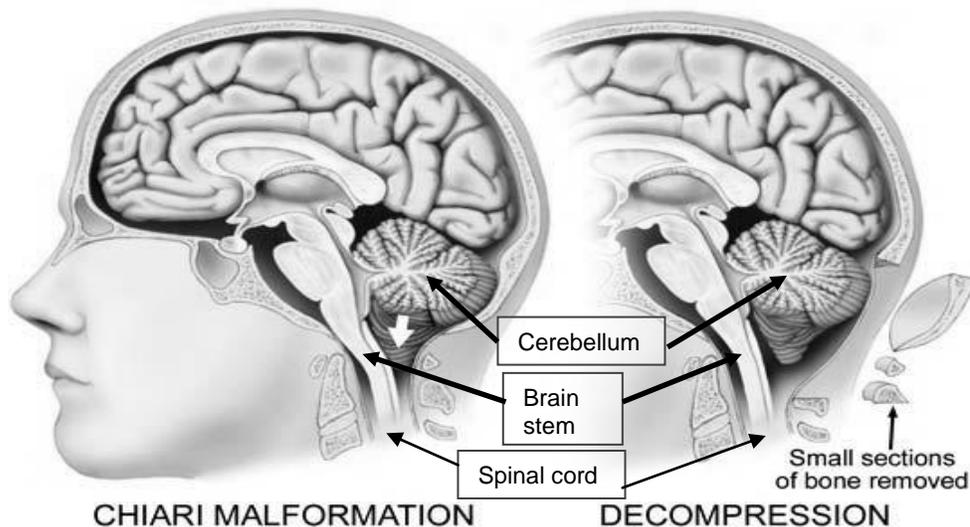
This problem occurs in the cerebellum, the brain stem and the spinal cord.

The cerebellum and the brainstem are not shaped properly. There is not enough space for them at opening at the base of the skull.

- There are two pegs of tissue that hang off the bottom of the cerebellum at the base of the brain. These pegs are called cerebellar tonsils. \_\_\_\_\_
- In Chiari II malformation, the cerebellar tonsils hang lower than normal. They hang past the base of the skull, into the spinal canal.
- Part of the brainstem also hangs lower than normal and gets pushed down into this space, making it very crowded. Spinal fluid is not able to flow normally through it.

This crowding may cause:

- Increased fluid in the brain. This is called hydrocephalus
- Fluid in the spinal cord. This is called syringomyelia.



### **How often is it found?**

Chiari II malformation occurs in most patients with spina bifida and is screened for regularly.

### **How does it affect my child?**

Not all children will have symptoms. It causes symptoms in one out of every three (1/3) children with spina bifida. Symptoms are more likely to occur when brain tissue is seen below the level of the fourth cervical vertebrae (C4). When brain tissue stops above the level of C3, the Chiari II malformation is less likely to cause obvious symptoms.

## Symptoms (continued)

Symptoms can start at any age and look different depending on the age of the child.

Babies usually have symptoms related to difficulty swallowing due to the brainstem not working properly.

You may see:

- Choking or sputtering during feedings.
- Weak cry
- Coughing, gagging.
- Noisy breathing.
- Arching the head backwards.

Older children may have symptoms due to brain stem dysfunction like those found in babies. They also may have symptoms due to fluid in the spinal cord (syringomyelia).

These include:

- Weakness, pain, numbness or tingling of the hands or arms.
- Pain in the back of the head or neck.

## What tests will be done?

An MRI (magnetic resonance imaging) of the brain and spinal cord will be done. This is the best way to see if Chiari II is present and to see how severe it is.

## How is Chiari II treated?

Chiari II malformation is treated with surgery. The surgery is called Chiari decompression. Surgery is most often recommended if your child has:

- Symptoms that are severe or getting worse.
- A syrinx of the spinal cord.

If symptoms are mild or absent, surgery may not be needed at all.

The goal of Chiari decompression surgery is to make more room at the base of the skull. This will allow spinal fluid flow easier in this area. It will also relieve pressure on the spinal cord. Children with hydrocephalus may need to also have a shunt placed in the brain.

It is not always possible to fix all of the symptoms your child is having.

**ALERT:** Call your child's doctor, nurse or clinic if you have any questions or concerns or if your child has special health care needs that were not covered by this information.

**This teaching sheet is meant to help you care for your child. It does not take the place of medical care. Talk with your healthcare provider for diagnosis, treatment, and follow-up.**