Spina bifida and Chiari malformation

What is Chiari malformation?
In Chiari (kee-ARE-ee) malformation, the brainstem and cerebellum are pushed down because there is less space in the brain. Because the normal flow of fluid is blocked, it builds up and increases pressure on the brain.

There are four types of Chiari malformation. While type I is most common, type II is often associated with spina bifida.

What happens when my child has Chiari malformation?
In a normal skull, the cerebellum (which controls balance) sits just above the spine. When the space for the cerebellum is too small, part of the cerebellum squeezes down the foramen magnum, a hole beneath the skull. Part of the brainstem, which contains many nerves for the head, eyes, and neck, is pushed down as well.

A Chiari malformation can cause:
- The brainstem, spinal cord, and cerebellum to stop working properly
- Cerebrospinal fluid (CSF) to stop flowing, which means less protection for the brain and spine
- Hydrocephalus (a buildup of CSF in the brain)

How do I know if my child has Chiari malformation?
If your child shows some or all of the following symptoms, have them checked for Chiari malformation:
- Headaches
- Difficulty breathing
- Not breathing (apnea)
- High-pitched noisy breathing (stridor)
- Problems swallowing or feeding (in babies)
- Arms that are weak and numb
How is Chiari malformation diagnosed?

An MRI is the best way to see a Chiari malformation. This test uses a powerful magnet and radio signals to see inside the body. Your child’s neurosurgeon will determine when it is necessary to do this test.

Once your child has an MRI, the neurosurgeon will look at the images to see if your child’s brainstem looks abnormal and CSF is flowing.

If there is a Chiari malformation, fluid pressure can build up causing breathing problems and weakness. Treatment is needed to decrease fluid pressure.

How is Chiari malformation treated?

There are a few ways to ease the pressure:

- **Decompression surgery:** This is the most common treatment for Chiari malformation. Your child’s neurosurgeon will make a cut at the back of your child’s head and remove a small piece of bone. This makes more room for the cerebellum and may relieve symptoms. The neurosurgeon may also make a cut in the dura mater, the brain and spinal cord covering, to make more room.

- **Inserting a shunt:** If your child has hydrocephalus, the neurosurgeon will insert a tube in their head to drain the excess fluid from the brain.

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Chiari malformation symptoms are different at different ages. This table will give you the most common signs of Chiari malformation.

<table>
<thead>
<tr>
<th>Age</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infants</td>
<td>Poor feeding, not breathing (apnea), or arm weakness</td>
</tr>
<tr>
<td>10 years and younger</td>
<td>High-pitched noisy breathing (stridor)</td>
</tr>
<tr>
<td>Older than 10</td>
<td>Arm weakness, trouble breathing, and sometimes high pitched noisy breathing (stridor).</td>
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</tbody>
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If you notice any of these problems, tell your child’s healthcare provider.

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Notes