Chiari Malformation: Treatment

Rick Labuda, Executive Director
director@conquerchiari.org
724-940-0116

Disclaimer: This presentation is intended for informational purposes only and may or may not apply to you. The editor and publisher are not doctors and are not engaged in providing medical advice. Always consult a qualified professional for medical care. This publication does not endorse any doctors, procedures, or products.

© 2012 C&S Patient Education Foundation, Conquer Chiari®
Treatment Options

Patients evaluated for Chiari-like symptoms or tonsillar herniation do not always have surgery right away. Anecdotally, many surgeons report that they recommend surgery for only 30%-50% of patients.

<table>
<thead>
<tr>
<th>1</th>
<th>Wait &amp; See</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Generally doctor driven</td>
<td></td>
</tr>
<tr>
<td>- Mild or no symptoms</td>
<td></td>
</tr>
<tr>
<td>- No definitive neurological signs</td>
<td></td>
</tr>
<tr>
<td>- Diagnosed incidentally</td>
<td></td>
</tr>
<tr>
<td>- Not clear if patient has Chiari</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>2</th>
<th>Treat symptoms individually</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Can be doctor or patient driven</td>
<td></td>
</tr>
<tr>
<td>- Symptoms not severe enough for surgery</td>
<td></td>
</tr>
<tr>
<td>- Patient does not want surgery</td>
<td></td>
</tr>
<tr>
<td>- Other medical conditions to address first</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>3</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Doctor and patient agree</td>
<td></td>
</tr>
<tr>
<td>- Symptoms clearly due to Chiari</td>
<td></td>
</tr>
<tr>
<td>- Severe or getting worse</td>
<td></td>
</tr>
<tr>
<td>- Most doctors recommend surgery when there is a significant syrinx</td>
<td></td>
</tr>
</tbody>
</table>
Wait and See

Reasons a doctor opts for Wait & See

*Symptoms, if present, may not be due to Chiari*
- For example, frontal or more general headaches as opposed to headaches in the back of the head. Headaches are very common and can be caused by many different things

*Incidental Diagnosis*
- A person is found to have herniation of the cerebellar tonsils, but does not have symptoms associated with Chiari
- For example, a child has an MRI after a head trauma during a sporting event. The MRI shows herniation, but there are no headaches, or neurological signs

*Doctor’s Judgment*
- An experienced neurosurgeon does not think the symptoms will respond to surgery

Procedure:
- Monitor the condition with regular MRIs
- Regular check-ups
- Be on the look out for Chiari type symptoms

Because there is no definitive test for symptomatic Chiari, and because people can have herniation of the cerebellar tonsils without symptoms, many people are advised to wait and see what happens
Treat Symptoms

- There is no overall or direct medical treatment for the condition itself, rather each symptom is treated individually beginning with the most troubling.

- There is no standard on how often MRI should be performed, someone with few or no symptoms may be advised to call if they experience anything unusual.

- Patients tend to find their own way of managing symptoms, whether adopting lifestyle changes, restricting activities, changing careers, trying alternative medicine or conversing with other Chiari patients on what works for them.

Some patients choose not to have surgery because they are afraid of the procedure or think they will end up worse off.
Surgery

Chiari surgery (posterior fossa decompression), is performed by a neurosurgeon to relieve pressure caused by herniated cerebellar tonsils.

50% or less of people diagnosed with Chiari have surgery.

After a successful decompression, it is expected that the cerebellar tonsils will move up and take on a healthier, rounded shape.

If a syrinx is present, a successful surgery should result in a reduction in size or at least prevent it from growing any larger.

American Association of Neurological Surgeons reported in 1997 that approx. 2,000 surgeries were performed each year by only 40% of neurosurgeons.

In 2007 11,000 surgeries were being performed each year by approx. 75% of neurosurgeons, a significant increase.

There is controversy over when to perform surgery for Chiari, especially if headaches are the only symptom.
Finding A Neurosurgeon

Conquer Chiari recommends that anyone diagnosed with Chiari be evaluated by a neurosurgeon. We believe that neurosurgeons are in the best position to provide treatment advice to patients.

You may choose to use all or none of the following tips to aid in your search. Whatever the case, we recommend that each patient do the work necessary to find the neurosurgeon that is right for them.

1. **Set emotion aside:** This can be extremely difficult to do, however it is an important decision, and a methodical approach to the matter can help.

2. **Establish your criteria:** What do YOU, as a patient, feel is important in a doctor. Some items to consider:
   - Location
   - University based or private practice
   - A doctor that specializes in Chiari surgeries or a general surgeon
   - Is bedside manner important
   - Do you have a straightforward case or a complicated one
   - Insurance and cost

*Continued on next page*
Finding a Neurosurgeon cont.

3. Create a list of candidates:
   Possible sources:
   - Professional society websites like: www.neurosurgerytoday.org
   - Neurosurgical faculty at local universities
   - Referrals from doctors, people you know
   - Internet searches
   - Chat rooms and message boards (with caution)

4. Create a short list based on your criteria.

5. Research further into doctors that made the short list.

6. Compare your candidates to your criteria and set up appointments.

7. Trust yourself: When you meet with the doctor do you get a good feeling? Can you communicate with him/her easily? Can you trust this person to make the best decisions regarding your care? If not, consider seeing someone else.

A skilled patient has a strategy, is focused, and in control.
Decision to Operate

- There is no objective measure or test to say whether someone should have surgery.
- There is disagreement among doctors about when surgery should be performed and how to perform it.
- Many factors surround the decision to operate from the patient’s, as well as the doctor’s perspective.

**Patient Perspective**
- No change from wait & see or treating symptoms
- Quality of life
- Desire to relieve symptoms
- Surgeon’s experience and judgment

**Doctor Perspective**
- Severity of symptoms
- Symptom progression
- Neurological signs
- Syrinx present
- MRI findings

Some people choose to get a second opinion (and third or fourth); people who do this should be prepared to hear different opinions on what they should do.
Surgical Procedure

The goal of posterior fossa decompression is to create more room around the malformation and restore the flow of spinal fluid. *There are many variations:*

**Surgical Steps**
- **Craniectomy** - A piece of the skull is removed
- **Laminectomy** - Part of one or more vertebrae are removed
- **Duraplasty** - The covering of the brain is opened and a patch is sewn in to make it bigger
- **Tissue removal** - Cerebellar tonsils are sometimes cauterized *(controversial)*
- **Plate insertion** - Mesh is inserted where skull was removed

**Surgical Variations/Controversies**
- How much bone is removed
- Whether the dura is opened at all
- What type of material is used for a dural graft (patch)
- Whether any brain tissue is removed
- Whether a mesh is put in to take the place of the removed piece of skull

* Images courtesy of Dr. Ghassan Bejjani
Surgery - The Experience

Every case is unique, but a typical experience might be:

- **Surgery:** ~4 hrs.
- **ICU:** One night
- **Hospital:** 3-4 days

**Normal Activity:** Variable

**Rest at home:** 3 wks to 3 mos
Other Surgeries

Sometimes Chiari patients undergo additional surgical procedures:

- **Shunt** - A tube-like medical device which is surgically implanted to divert CSF from one place to another. Patients with hydrocephalus (excess CSF in the brain), or intracranial hypertension (elevated pressure in the head) may benefit from this procedure.

- **Fusion/Stabilization** - As part of a decompression, or at a later date, a Chiari patient may require surgery to stabilize their neck. Stabilization usually involves fusing several vertebrae together and results in a reduced range of neck motion for the patient.

- **Tethered Cord** - The specifics of tethered cord surgery depend on what the cause of the tethering is. For a tight filum terminale (at the base of the spinal cord), the filum is sectioned, or cut, to release the tension.

- **Scoliosis** - Research has shown that decompression surgery can effectively stop the progression of scoliosis related to Chiari; however in cases where it doesn’t, the placement of rods and screws may help to stop the curve from getting worse.

- **Transoral decompression** - The surgery for basilar invagitation is called transoral decompression because the surgeon goes in through the mouth.

- **Acquired Chiari** - For acquired Chiari, the underlying cause of the herniation is addressed, possibly in conjunction with a decompression. For example if a tumor or cyst is pushing the cerebellar tonsils down, then the mass will be removed.
Complications

Complication rates for Chiari surgery vary among surgeons, but are often less than 5%

<table>
<thead>
<tr>
<th>Complication</th>
<th>What it is</th>
<th>Additional Information</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infection</td>
<td>As with any surgery, infection is a possibility with Chiari surgery.</td>
<td>Infections can range from superficial to serious.</td>
</tr>
<tr>
<td>Graft Problems</td>
<td>Patient’s body may react poorly to the graft resulting in inflammation and scarring.</td>
<td>These problems do not always come to light right away, but may surface down the road and if bad enough may require another surgery to replace the graft.</td>
</tr>
<tr>
<td>CSF Leak</td>
<td>Cerebrospinal fluid leaks at the dural graft site.</td>
<td>New materials and techniques have reduced the rate of CSF leaks.</td>
</tr>
<tr>
<td>Pseudomeningocele</td>
<td>Occurs when the subarachnoid space (where the CSF circulates) bulges into the surrounding tissue.</td>
<td>The size of Pseudomeningoceles can range from small ones which do not require any intervention, to large ones which may require surgery.</td>
</tr>
<tr>
<td>Cerebellar Slumping</td>
<td>Also known as ptosis. One of the more serious complications, which involves the cerebellum slumping down even further into the spinal area after surgery.</td>
<td>Can be difficult to treat, surgeons from UCLA published a technique which involves rebuilding bony support for the cerebellum while still maintaining an adequate decompression.</td>
</tr>
</tbody>
</table>
Recovery

Factors that can affect recovery

- Surgical Complications
- Success of Decompression
- Other Diseases
- Duration of Symptoms
- Lifestyle
- Mental Attitude

1 WEEK POST-OP
- 3-4 days hospital recovery
- Rest at home
- 7-10 days remove staples and or stiches
- Restricted activity

3 MONTHS POST-OP
- Resume light activity
- Anticipation to return to normal activities
- Patient’s that are feeling better may push too hard
- Patients not feeling up to par, doubt sucess of surgery

1 YEAR POST-OP
- Some patients are back to normal, full activity, no restrictions
- Others may still feel weak, due to complications or complex cases

Patient should discuss in detail with their surgeon when they can resume work, school, driving, and other activities
Factors that may affect outcomes include: age, syringomyelia, bony abnormalities, scoliosis, duration of symptoms, and concurrent conditions.

Common residual symptoms include pain, muscle weakness, and loss of sensation; patients will adapt their lifestyle accordingly.

Symptoms may fluctuate over time; patients report that trauma can cause symptoms to come back.

Patients in the poor range (10-20%) may end up getting revisions or other surgeries.

Over the very long term (10 years+), symptom recurrence has been reported, but has not been studied extensively.

Published outcomes research has many limitations, such as poorly defined measures and short durations. Patient should discuss their individual case with their surgeon to establish expectations before surgery.
Why Do Surgeries Fail?

*Surgery can fail for a number of reasons*

<table>
<thead>
<tr>
<th>Surgery Related</th>
<th>Patient Related</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inadequate decompression</td>
<td>Concurrent conditions still cause symptoms</td>
</tr>
<tr>
<td>- not enough bone removed</td>
<td>- pseudotumor cerebri; elevated pressure in the brain</td>
</tr>
<tr>
<td>- dura was not opened</td>
<td>- basilar invagination; compression of the brainstem</td>
</tr>
<tr>
<td>Recurrent obstruction</td>
<td>- from the spine</td>
</tr>
<tr>
<td>- scarring disrupts CSF flow</td>
<td>- genetic disorders</td>
</tr>
<tr>
<td>- bone regrowth in children</td>
<td></td>
</tr>
<tr>
<td>- spinal cord retethers</td>
<td></td>
</tr>
<tr>
<td>Surgical complications</td>
<td>Symptoms not due to Chiari</td>
</tr>
<tr>
<td>Altered CSF dynamics</td>
<td>- symptoms were due to something else</td>
</tr>
<tr>
<td>Cervical instability</td>
<td></td>
</tr>
</tbody>
</table>

It is important to try to understand the reason for failure before undergoing additional treatment, but sometimes a revision surgery will be necessary.
Pediatric Treatment

*It is not yet clear if when it comes to Chiari, children are just small adults, or if pediatric Chiari is fundamentally different.*

<table>
<thead>
<tr>
<th><strong>Surgery</strong></th>
<th>Bone only decompression is more common in children. It results in a drastically reduced complication rate and quicker recovery, but the reoperation rate is higher.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>In a study of 130 children, 2.3% experienced complications</td>
</tr>
<tr>
<td><strong>Pain Control</strong></td>
<td>A 2004 study from the University of Alabama-Birmingham found that giving children regularly scheduled doses of pain medicine was more effective than waiting for the children to ask for them.</td>
</tr>
<tr>
<td><strong>Recovery</strong></td>
<td>Overall, children tend to recover from surgery more quickly than adults</td>
</tr>
<tr>
<td><strong>Outcomes</strong></td>
<td>While the data is not conclusive, there are indications that children in general have better outcomes than adults</td>
</tr>
<tr>
<td><strong>Living With</strong></td>
<td>Controversy over whether children should have any restrictions after surgery, especially when it comes to sports.</td>
</tr>
</tbody>
</table>
Articles:
- Study Explores The Natural History Of Chiari...
- Tips On Finding A Neurosurgeon...
- Survey Shows How Doctors Worldwide Treat Chiari ..
- Simple Idea To Improve Chiari Surgery..
- Is Surgery Necessary If There Are No Symptoms?
- Study Compares Surgical Techniques...
- Surgical Technique Alleviates Cerebellar Slumping...
- Study Looks At Long Term Impact Chiari Has On Quality Of Life...
- What Can Be Learned From 130 Chiari Kids...
- Large Study Examines Surgical Outcomes In Children ...
- Trying To Identify Why Surgeries Fail...

Books:
- Conquer Chiari: A Patient’s Guide
- Contents Under Pressure

Videos:
- 2010 Conference: New Developments..

Questions or comments about this presentation can be sent to:
director@conquerchiari.org

© 2012 C&S Patient Education Foundation, CONQUER CHIARI ® This presentation is for informational purposes, consult a qualified professional for medical advice.