Conquering Chiari: Treatment

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Treatment Options

Patients evaluated for Chiari do not always have surgery. Anecdotally, many surgeons report that they only recommend surgery 30%-50% of the time.

Wait & See

- Generally doctor driven
- Mild or no symptoms
- No definitive neurological signs
- Diagnosed incidentally
- Not clear if patient has Chiari

Treat symptoms individually

- Can be doctor or patient driven
- Symptoms not severe enough for surgery
- Patient does not want surgery
- Other medical conditions to address first

Surgery

- Doctor and patient agree
- Symptoms clearly due to Chiari
- Severe or getting worse
- Most doctors recommend surgery when there is a significant syrinx



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.

Procedure

- Be on the look out for Chiari type symptoms
- Follow-up MRIs, at regular intervals, may or may not be recommended



Treat Symptoms Individually

If symptom(s) are not severe, the patient may be advised (or choose) to try to manage them without surgery.

Patients tend to find their own way:

- Medication for headaches and pain
- Alternative treatments
- Lifestyle changes
- Avoiding certain activities

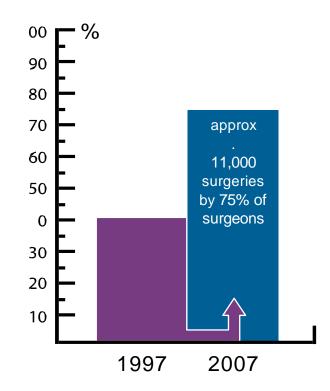


Some patients choose not to have surgery because they are afraid of the procedure or think they will end up worse off.



Surgery

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



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

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







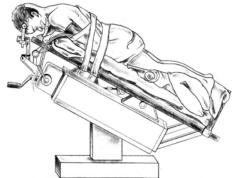


Some people choose to get a second opinion (and third or fourth); people who do this should be prepared to hear different opinions on what they should do.



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:**



* Images courtesy of Dr. Ghassan Bejjani



- 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

Opening the Dura

There is a significant amount of research on whether the dura should be fully opened during surgery. Basically, each approach has advantages and disadvantages:



- Opening
 - Lower rate of reoperation, better syrinx improvement
- Not Opening
 - Much lower complication rate, shorter surgical time and hospital stay

- Opening
 - Higher complication rate, longer hospital stay
- Not Opening
 - Higher reoperation rate, worse outcomes with syrinx reduction

Not opening the dura is used more often in treating kids



Surgery – The Experience

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







ICU: One night



Hospital: 3-4 days







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.
- * <u>Scoliosis</u> 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.
- * <u>Transoral Decompression</u> The surgery for basilar invagination is called transoral decompression because the surgeon goes in through the mouth.
- * <u>Acquired Chiari</u> 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	What it is	Additional Information
Infection	As with any surgery, infection is a possibility with Chiari surgery.	Infections can range from superficial to serious.
Graft Problems	Patient's body may react poorly to the graft resulting in inflammation and scarring.	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.
CSF Leak	Cerebrospinal fluid leaks at the dural graft site.	New materials and techniques have reduced the rate of CSF leaks.
Pseudomeningocele	Occurs when the subarachnoid space (where the CSF circulates) bulges into the surrounding tissue.	The size of Pseudomeningoceles can range from small ones which do not require any intervention, to large ones which may require surgery.
Cerebellar Slumping	Also known as ptosis. One of the more serious complications, which involves the cerebellum slumping down even further into the spinal area after surgery.	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.

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



Recovery

Factors that can affect recovery

- Surgical complications
- Success of decompression
- Other diseases
- **❖** Duration of symptoms
- **❖**Lifestyle
- Mental attitude

Patients should discuss in detail with their surgeon when they can resume work, school, driving, and other activities

1 WEEK POST-OP

3-4 days hospital recovery

Rest at home

7-10 days remove staples/stiches
Restricted Activity



3 MONTHS POST-OP

Resume light activity
Patient's not feeling up to par, doubt
success of surgery

Anticipation to return to normal activities Patient's that are feeling better may push too hard



Some patients are back to normal, full activity, no restrictions

Other's may still feel week due to complications or complex issues



Surgical Outcomes According to Doctors

Poor Good Excellent 10-20% 30-40% 30-40%

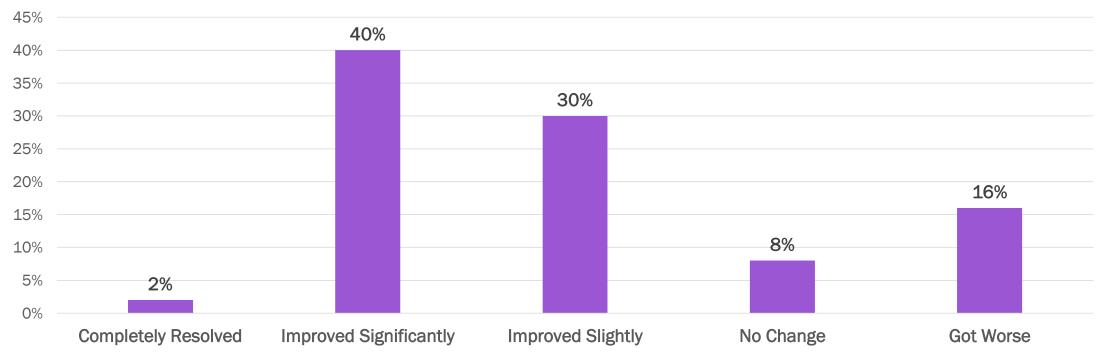
- ❖ Valsalva (Chiari) headaches are the most likely to improve with surgery
- ❖ Patients in the poor range (10-20%) may end up getting revisions or other surgeries.
- Over the very long term (10 years+), symptom recurrence has been reported, but has not been studied extensively.
- Sometimes new symptoms appear after surgery

Many journal publications report surgical "success" rates of 80% or higher; patients should discuss in detail with their doctors what to expect and what will define success



What Do Patients Say?





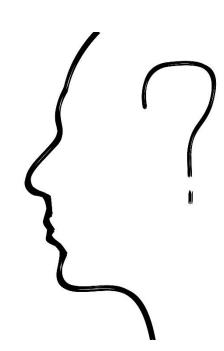
Nearly 80% of people say that knowing in advance how they would feel, they would go through with it again



Why do surgeries fail?

Surgery Related

- Inadequate Decompression
 - Not enough bone removed
 - Dura was not opened
- Recurrent Obstruction
 - Scarring disrupts CSF flow
 - ❖ Bone regrowth in children
- Surgical Complications
- Altered CSF dynamics
- Cervical Instability



Patient Related

- Concurrent conditions still cause symptoms
 - Pseudotumor cerebri; elevated pressure in the brain
 - Basilar invagination; compression of the brainstem from the spine
 - Genetic disorders
- Symptoms not due to Chiari
 - Symptoms were due to something else

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



Pediatric Surgery

Surgery

Bone only decompression is more common in children.

Pain Control

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.

Recovery

Overall, children tend to recover more quickly than adults.

Outcomes

Research indicates that children in general have better outcomes than adults.

Living with

Controversy over whether children should have any restrictions after surgery, especially when it comes to sports.

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



Predicting Outcomes

From Imaging:

CCRC is actively working to identify quantitative measures that can be used to identify good surgical candidates

Negative Factors:

Scoliosis, syringomyelia, duration of symptoms before surgery, additional bony abnormalities such as cervical instability, and surgical complications have all been shown to be associated with poorer outcomes

Long Term Symptom Recurrence:

- Although the data is limited a couple of studies have shown that for 10% or more of adult patients, symptoms can come back years after surgery, perhaps due to the build-up of scar tissue
- Studies have also shown that for young kids, bone can grow back leading to recompression and symptoms coming back



Summary

- ❖ Up to half of patients who are evaluated by a neurosurgeon will be advised to have surgery
- For many types of cases, there is NOT agreement on whether surgery is the best approach
- The biggest surgical controversy is whether to open the dura
- ❖ Doctors report that about 80% of adults will experience some improvement in symptoms
- Many patients report some residual symptoms, especially chronic pain
- Kids tend to recover more quickly and have better outcomes than adults
- For some patients, multiple surgeries will be required

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