

## Key Points

1. Pseudotumor Cerebri is a condition where the pressure of CSF in the brain is chronically elevated for unknown reasons
2. PTC appears to be somehow linked to Chiari as a high percentage of PTC patients have Chiari
3. PTC could cause Chiari, Chiari could cause PTC, Chiari surgery could cause PTC, or some combination of the three
4. PTC has been identified as a possible cause of failed Chiari surgeries
5. Report details two cases from India where patients developed PTC symptoms a week after Chiari surgery
6. In both cases, medicine was able to control the PTC

## Definitions

**acetazolamide** - also known as Diamox; medicine used to lower elevated ICP

**idiopathic** - due to an unknown cause

**idiopathic intracranial hypertension (IIH)** - condition where a person suffers from elevated intracranial pressure with no visible cause

**intracranial pressure (ICP)** - the pressure of the cerebrospinal fluid in the skull

**lumbar puncture (LP)** - procedure where a needle is inserted into the CSF space of the lower back region; can be used to remove CSF or insert drugs

**papilledema** - swelling of the optic nerve due to increased intracranial pressure.

**posterior fossa volume** - measurement of the size of the posterior fossa, which is the space in the back of the skull where the cerebellum is situated

## 2 Cases of Pseudotumor In Failed Chiari Surgery

May 31st, 2009 -- Pseudotumor cerebri (PTC), also known as idiopathic intracranial hypertension (IH) is a condition characterized by an increase in intracranial pressure (the pressure of spinal fluid in the head) with no apparent cause. The most common symptom is a pressure headache, with other symptoms including double vision, visual blurring, nausea, vomiting, dizziness, and ringing in the ears. The most serious symptom associated with PTC is vision loss. The sustained pressure associated with PTC can eventually damage the optic nerve - the bundle of fibers which connect the eye to the brain - and if not treated can lead serious vision problems.

A growing body of research has shown what appears to be a strong link between Chiari and PTC. Specifically a high percentage of PTC patients also have Chiari. In addition, PTC has been identified as a possible cause of failed Chiari surgeries (Fig 1). It is not clear, however, if PTC causes Chiari, if Chiari causes PTC, if decompression surgery can cause PTC, or if any or all of the above can come into play for a given patient.

**Figure 1**  
**Possible Reasons For Failed Surgery (From Presentation By Dr. Ghassan Bejjani)**

- Inadequate Decompression
  - No duraplasty
  - Not enough bone removed
- Recurrent CSF Obstruction
  - Scarring
  - Retethering
  - Regrowth of bone
- Surgical Complications
  - Pseudomeningocele
  - Cerebellar sag
  - Altered neural hydrodynamics
  - Cranio-cervical instability
  - Muscle adheres to dura
- Concurrent Conditions
  - Pseudotumor cerebri
  - Basilar invagination
  - Other (genetic)
- Symptoms Not Due To Chiari
  - Asymptomatic tonsillar ectopia

It could be that the sustained increase in pressure associated with PTC eventually leads the cerebellum to herniate out of the skull and creates a Chiari malformation. If this were the case, then decompression surgery would help symptoms associated with direct compression of the malformation, but would not relieve the symptoms associated with the elevated pressure of PTC.

It may also be the case that the blockage caused by a Chiari malformation, which we know can elevate the intracranial pressure, may lead to a fundamental change in the CSF system and eventually PTC. In this case, even though the region around the Chiari malformation is decompressed surgically, for unknown reasons, the intracranial pressure remains high.

A third possibility is that both Chiari and PTC are manifestations of a more fundamental problem, such as a too small posterior fossa (the skull region where the cerebellum sits). Perhaps some people with a small or

**pseudotumor cerebri (PTC) -**

another name for idiopathic intracranial hypertension; not used as much anymore

**cerebellar tonsils** - portion of the cerebellum located at the bottom, so named because of their shape

**cerebellum** - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

**cerebrospinal fluid (CSF)** - clear liquid in the brain and spinal cord, acts as a shock absorber

**Chiari malformation I** - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

**decompression surgery** - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

**syringomyelia** - condition where a fluid filled cyst forms in the spinal cord

**Source**

*Pseudotumor cerebri: as a cause for early deterioration after Chiari I malformation surgery.* Furtado SV, Visvanathan K, Reddy K, Hegde AS. Childs Nerv Syst. 2009 Mar 19. [Epub ahead of print]

Skau M, Brennum J, Gjerris F, Jensen R. *What is new about idiopathic intracranial hypertension? An updated review of mechanism and treatment.* Cephalalgia. 2006 Apr;26(4):384-99

abnormally shaped skull develop PTC, while others develop Chiari, and still others develop both.

Finally, it may be that Chiari surgery itself plays a role in the development of PTC. There appear to be some cases where symptoms associated with PTC don't appear until after Chiari surgery. How, and even if, decompression surgery could lead to PTC is not at all clear.

Since the cause of PTC is unknown, treatments tend to focus on the symptoms and can involve drugs or surgery. Unfortunately, to date there have not been any rigorous studies comparing the effectiveness of drug treatments versus surgery.

On the medicine side, acetazolamide is commonly one of the first drugs tried. It is believed to act by reducing the production of cerebrospinal fluid and thus leads to a decrease in ICP. If the acetazolamide does not work, it is sometimes supplemented with a second medicine, furosemide. Recently, an anti-convulsant, topiramate, has been reported to be used to treat PTC, but more research is required to determine its effectiveness. In addition to treating the symptoms with medicine, if the patient is obese, weight loss has been shown to be an effective approach to alleviating symptoms.

On the surgical side, a shunt can be inserted to divert CSF and lower the pressure in the head. While shunting is very effective in relieving symptoms, a patient then has to deal with potential problems related to having a shunt inside of them; namely mechanical malfunction and infection. Such problems are not uncommon can lead to the need for additional surgeries to revise or replace the shunt.

For a patient whose vision is at risk, a surgeon may elect to decompress the optic nerve by perforating the sheath over the nerve bundle. This reduces pressure on the nerve and according to published reports is usually successful in stabilizing or improving visual function. However, this type of surgery does not address the problem of elevated pressure directly and relief from other PTC symptoms is not as great.

One approach to treating PTC which may be falling out of favor is to repeatedly drain CSF through lumbar punctures. Draining a large quantity of CSF in this way usually provides temporary relief from symptoms, but again does not really address the underlying problem.

Overall, treatments for PTC are effective for a majority of people. Research has found that 70% or more of patients experience symptom relief or resolution within a couple months of starting treatment. However, there does appear to be a subset of patients, perhaps as many as 25%, for whom PTC becomes a long, difficult battle. In addition, much like Chiari, recurrence of symptoms, even years down the road, has been noted.

Although the exact mechanism which leads to the increased pressure in PTC is not known, from an abstract point of view there are several possibilities: increased production of CSF, abnormal absorption of CSF, increased brain mass, and obstruction of blood outflow from the brain. However, and somewhat surprisingly, research has failed to consistently find any of the above to be the problem in PTC patients. Interestingly, studies have shown an increase in resistance to CSF flow (much like exists with Chiari) in 75% or more of PTC patients.

In terms of PTC related to failed Chiari surgery, in several research publications, Dr. Bejjani has identified a sub-group of patients for whom decompression surgery provides temporary relief of symptoms, which then return after a period of weeks or months. Lumbar punctures on these patients have shown that they have elevated ICP, which is the likely reason that the symptoms came back after a period of time. Again, it is not entirely clear if the PTC was present, but undetected before surgery, as the symptoms of PTC overlap such much with Chiari symptoms.

A recent publication from surgeons in India (Furtado et al) in Child's Nervous System provides more evidence that PTC should be considered in cases where Chiari surgery appears to succeed initially, but then symptoms return after a short period of time. The publication details the cases of a 12 year old girl and a 24 year old man who both developed serious symptoms associated with PTC after Chiari surgery.

In the girl's case, six days after surgery, she began to have trouble walking and had a severe headache with vomiting. There was no evidence of a CSF leak, but a lumbar puncture showed that her ICP was indeed elevated. Medication was used to lower her pressure and her symptoms resolved.

One week after surgery, the man was taken to an emergency room in a coma and struggling to breathe. There were indications that his intracranial pressure was so high that the doctors could not do a lumbar puncture. Again, medication brought the pressure down, but not before it had caused bleeding in his brain.

Interestingly, the researchers used the two patients MRIs to measure their posterior fossa volume and found them to be small, even as compared to other Chiari patients. However, the precise meaning of this finding is not clear.

Although pseudotumor is by no means the only reason for which Chiari surgery can fail (as detailed by Dr. Bejjani), it should definitely be considered when symptoms initially go away for a period of time and then come back. This is yet another example of the importance of researching and identifying patient sub-groups, so that

they may get the proper treatment and improve their outcomes.

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