









Definitions

dura - thick, outer covering of the brain and spinal cord

duraplasty - surgical technique where the dura is expanded by sewing a dural patch into it

foramen magnum - opening at the base of the skull through which the brain and spine connect

intracranial pressure (ICP) - the pressure of CSF in the brain

neural hydrodynamics - refers to the flow and motion of CSF and blood in the brain and spinal cord

pseudomeningocele - a possible complication of decompression surgery, refers to when the subarachnoid space bulges into the surrounding tissue

pseudotumor cerebri - also known as idiopathic intracranial hypertension; condition where intracranial pressure (ICP) is chronically elevated

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

Symposium Update: Failed Surgery

July 31, 2007 -- At the 2007 Conquer Chiari Research Symposium, Dr. Ghassan Bejjani, of the University of Pittsburgh and a Scientific Advisor to Conquer Chiari, presented a concise but thorough overview of the different reasons that decompression surgery can fail in adults. His analysis was based on his own experience plus the published medical literature. While there are not really any large scale reports on failed surgery in adults, Dr. Bejjani was able to piece together different causes from references in smaller case series.

While his presentation was focused mostly on adults, it is clear that most, if not all, of the reasons he cited apply equally to children as well. The following is a summary of his analysis.

Possible Reasons For Failed Decompression Surgery

1. Inadequate Decompression

- a. No Duraplasty In an effort to reduce trauma during surgery, there is a growing trend to leave the dura intact, especially for children. If a patient did not receive a duraplasty during the initial surgery and symptoms did not improve, many surgeons will reoperate and open the dura.
- b. Not Enough Bone Removed If not enough bone is removed during decompression then the brain tissue can still be compressed and CSF flow blocked. Essentially, the decompression was not big enough and a reoperation is likely necessary.

2. Recurrent CSF Obstruction

- a. Scarring Sometimes scarring and adhesions even if removed during the initial surgery can redevelop and obstruct CSF flow leading to problems.
- b. Retethering of the Spinal Cord An extension of the scarring problem above, if the spinal cord becomes abnormally anchored and tethered, symptoms are likely to result
- c. Regrowth of Bone at the Foramen Magnum There have actually been several case reports, documented with MRIs, of children in whom the bone that was removed around the cerebellar tonsils actually regrows and recompresses the area, necessitating additional surgery.

3. Surgical Complications

- a. Pseudomeningocele One of the more common complications associated with decompression surgery, pseudomeningoceles can range from asymptomatic to requiring surgical repair.
- b. Cerebellar Sag (Ptosis) One of the most serious complications, cerebellar sag is where the cerebellum slumps down after surgery because its bony support has been removed. Although it can be difficult to treat, Lazareff recently published a surgical technique which involves rebuilding the support for the brain while maintaining an adequate decompression.
- c. Altered Neural Hydrodynamics Neural Hydrodynamics refers to how CSF flows in the brain and spine. Some patients can develop hydrocephalus or chronically elevated intracranial pressure (IIH) after decompression surgery (or it was undiagnosed before the surgery, see below). Naturally, these conditions cause symptoms, often similar to Chiari, and are usually treated surgically by implanting a shunt to divert CSF.
- d. Cranio-Cervical Instability Chiari patients tend to have an unusual anatomy at the junction of the head and neck. This, combined with removing bone during decompression surgery, can lead to instability in some people and cause problems. If the neck is not stabilized during the initial surgery, additional surgery may be necessary to do so.
- e. Muscle Adheres to the Dura Since part of the skull is removed during surgery, if nothing is put back in its place, certain muscles of the neck have nothing solid to attach to and may attach directly to the dura. If this occurs, then when the neck muscles are used, they can pull on the dura (thus lowering compliance) and cause problems, such as headaches. This is why surgical manufacturers are working with neurosurgeons to develop Chiari plates that are put in where the skull pieces were removed. A plate provides the muscles with something to attach to and can greatly reduce this problem.

4. Concurrent Conditions

- a. Idiopathic Intracranial Hypertension The link between chronically elevated intracranial pressure (also known as pseudotumor cerebri) and Chiari is not completely understood. While some patients appear to develop PTC after surgery, it is also likely that some patients have PTC prior to surgery, which means that decompression surgery will only provide temporary relief of their symptoms.
- b. Basilar Invagination A significant percentage of Chiari patients also have some degree of basilar invagination or impression (where the second vertebrae is moved up and pushes on the brainstem).
 In general, this is not directly treated by a posterior fossa decompression, so a patient whose symptoms are due mostly to basilar invagination may not get relief from a standard Chiari decompression.
- Other Chiari has been linked with numerous genetic conditions which obviously can bring their own complications and problems to a Chiari surgery.

5. Symptoms Not Due To Chiari

a. Asymptomatic Tonsillar Ectopia - Because of its many manifestations, Chiari is often misdiagnosed as other diseases and problems. However, there is a flip-side to this phenomenon as well. Namely, because there is not a good, objective definition of Chiari and not everyone with herniated cerebellar tonsils has symptomatic Chiari, a person's symptoms may not always be due to the tonsillar herniation. When this is the case, then of course decompression surgery will do nothing to help the patients.

Dr. Bejjani's presentation has received the most hits in the webcast archive, a possible indication of the unfortunately high failure rate for Chiari surgery.

- Rick Labuda

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