

Key Points

1. Study identified 17 children who developed acquired Chiari as a long term complication of shunting
2. 5 of the children had significant symptoms, 4 had mild headaches, and 8 were asymptomatic
3. Six of the children underwent successful surgery for their symptoms
4. Researchers found that all the children had small posterior fossas and thickened skull bases
5. This finding is in line with previous work which identified the same thing in shunted children
6. The implications for the pathogenesis of Chiari are not clear but do raise questions about the role that CSF pressure plays in the development of a normal skull size and shape

Definitions

acquired - in relation to a medical condition, refers to something that is caused by something else, as opposed to being born with the problem

asymptomatic - without symptoms

herniation - protrusion of a body structure, such as the cerebellar tonsils - into a space where it doesn't normally belong

hydrocephalus - condition characterized by a build-up of CSF in the brain

pathogenesis - the cause of a disease

posterior fossa - region in the back of the skull where the cerebellum sits

shunt - implantable tube which diverts CSF from the brain

ventricles - spaces in the brain which are filled with CSF

Long Term Shunting Can Lead To Acquired Chiari

January 31st, 2009 -- A recent study from a group of Italian surgeons (Caldarelli et al.) adds to the evidence that Chiari can develop as a long-term complication of CSF shunting. While most cases of Chiari are considered to be congenital - meaning people are born with the anatomy that leads to symptoms - in the case of shunting, the Chiari is actually acquired, meaning that it develops over a period of time. Acquired Chiari can also be due to some type of space occupying mass in the brain - such as a tumor or cyst - which forces the cerebellar tonsils out of the skull.

Published on-line first in the journal, Child's Nervous System, the Italian research looked at 1,700 children who had shunts placed over a 30 year period. Out of that large group, they identified 17 who had developed acquired Chiari malformations of at least 5 mm (Figure 1). Thirteen were boys and 4 were girls. Most of the children had hydrocephalus, which was the reason they were shunted. Interestingly, while most of the children had the shunts placed while they were less than a year old, the Chiari did not develop until years later.

The surgeons were certain that the Chiari was acquired because imaging from the shunt surgery (and follow-ups) clearly showed there was no herniation at that point. Interestingly, not all the children had Chiari related symptoms. In fact, eight were asymptomatic, four had mild headaches, and five were considered to be truly symptomatic. In studying the MRIs, the surgeons found that 5 children had herniations of 5mm, 6 had herniations between 5mm-10mm, and 5 had significant herniations greater than 10mm. However, as with most Chiari cases, the amount of herniation did not correlate with symptoms.

In terms of treatment, six of the children underwent surgery to relieve their Chiari related symptoms, which was successful in each case. Beyond identifying a long-term complication of shunting, this study may have implications for the pathogenesis of Chiari in general.

Specifically, the researchers found that the children with acquired Chiari had small posterior fossas and thickened skull bases. The alert reader will recognize that many studies have found that Chiari patients in general have small posterior fossas. This finding is similar to work by Lazareff which found that children who developed Chiari related symptoms due to shunting had skull dimensions similar to general Chiari patients.

While this paper does not go into detailed discussion of this finding, one logical implication is that CSF pressure may play an important role in the normal development of the posterior fossa size and shape. In fact, this has been hypothesized in relation to Chiari II and spina bifida and is known as the hydrostatic theory. In terms of Chiari II, it is proposed that the opening in the spinal canal results in a loss of CSF pressure, which in turn results in a small posterior fossa and the herniation of the cerebellar tonsils. In the shunting cases, because CSF is diverted out of the brain, it might be that in some cases this results in not enough CSF pressure for normal skull development.

Interestingly, and despite this, most attendees at the recent Conquer Chiari research symposium felt that Chiari I and Chiari II are separate entities and not related. Similarly, one could argue that just because long-term shunting results in a small posterior fossa in some cases, does not mean a similar mechanism is at work in congenital Chiari cases. Rather, there could be other reasons for the development of a small posterior fossa, such as genetic transcription errors which result in what should be bone forming as something else instead.

Still, it is curious that to date no one has really explored whether low CSF pressure during fetal development - such as due to a CSF leak which later closes - could lead to a small posterior fossa, and in turn Chiari.

Figure 1: Select Characteristics of Acquired Chiari Patients

Patient #	Age at Shunt (months)	Age at Chiari Dx (years)	Herniation (mm)
1	1	10	5
2	3	17	>10
3	3	21	5
4	60	10	5
5	2	12	>10
6	1	11	>10
7	2	13	5-10

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

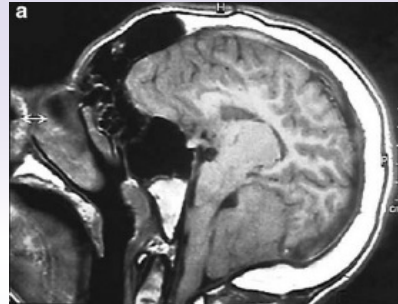
Chiari malformation I - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

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

8	36	6	5-10
9	.3	8	5
10	.6	14	>10
11	1	12	5-10
12	.3	3	>10
13	1	3	5
14	.6	11	>10
15	3.5	5	5
16	54	12	5-10
17	.63	24	5-10

Figure 2: MRI Taken 13 Years After Shunt Placement



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Source

A late complication of CSF shunting: acquired Chiari I malformation. Caldarelli M, Novegno F, Di Rocco C. Childs Nerv Syst. 2008 Dec 5.

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