

Key Points

1. Surgeons at UAB published their experience with 500 surgical pediatric Chiari patients
2. Outcomes were good with 83% experiencing good symptom relief
3. Complication rate was a low 2.4% and there were no deaths
4. Common symptoms included headache, neck pain, back pain, scoliosis, and upper extremity problems
5. Syringomyelia and hydrocephalus were common, but there were also many other additional diagnoses
6. Children were only out of school for an average of 12 days

Definitions

apnea - condition where a person temporarily stops breathing many times while sleeping

ataxia - trouble walking

cervical - upper part of the spine

dura - covering of the brain and spine

duraplasty - surgical technique where a patch is sewn into the dura to make it bigger

cine MRI - type of MRI which can show the flow of CSF

C1 - the first cervical vertebra

C2 - the second cervical vertebra

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

laminectomy - surgical technique where part of one or more vertebrae are removed

scoliosis - abnormal curvature of the spine

syringobulbia - condition when a syrinx is located in the brainstem as opposed to the spine

Surgery Group Discusses Experience with 500 Chiari Children

June 30th, 2011 -- A group from the University of Alabama - Birmingham has published their experiences with one of the largest patient groups to date. Specifically, the research involves 500 surgical pediatric Chiari patients, spanning more than a decade. Interestingly, the authors state that in general they only operate on about 20% of the people they see who have an MRI indication of Chiari.

The children ranged in age from as young as just 2 months old to a more adult like 20 years, with the average age being 11. In contrast to many adult patient series where women are normally in the majority, the patient was comprised of 271 boys and 221 girls. Headache and neck and back pain were by far the most common presenting symptom (Table 1), although scoliosis and upper extremity issues were also fairly common. A couple of the more rare - but interesting - symptoms were chronic hiccups and rage attacks. Chronic hiccups have been associated with Chiari in published case studies, and Conquer Chiari has had questions from parents and teachers about rage attacks.

Table 1: Most Common Presenting Symptoms Among 500 Pediatric Chiari Patients (Most Patients Exhibited More Than One Symptom)

Symptom	Number With
Headache/neck/back pain	200
Scoliosis	90
Upper extremity pain/weakness	41
Apnea	25
Trouble swallowing	20
Ataxia	19
Irritability	19
Nasal speech	15

There were a number of common associated diagnoses (Table 2), including hydrocephalus of course and neurofibromatosis 1. Interestingly, the group had 21 children with growth hormone deficiency (the authors have previously published on growth hormone deficiency and Chiari). Highlighting the often confusing nature of Chiari, there were 25 associated diagnoses which affected only one patient in the group. The question of how many of these additional diagnoses were related to Chiari by more than just chance is difficult to say and was not addressed by the researchers. Slightly more than half of the group had syringes and there were six cases of syringobulbia.

Imaging studies showed that 22% of the group had herniations between the foramen magnum and the level of C1, 37% were at C1, 39% were at C2, and 1% was at C2 (Table 3). Almost all of the children (97%) had pointed tonsils. The doctors routinely used cine MRI to assess CSF flow for a couple of years but stopped because they felt there was a high rate of false negatives.

Each member of the group underwent a posterior fossa decompression with laminectomy, and all but one had a duraplasty. The surgeons used different materials for the dural patch. About 10% of the group also had their cerebellar tonsils reduced using coagulation. The surgeries took 95 minutes on average and the children were generally in the hospital for 3 days and returned to school after 12 days. The complication rate was only 2.4% and thankfully there were no deaths associated with the surgery.

In terms of outcomes, the children were followed for an average of 5 years, with 83% experiencing good relief of symptoms. Fifteen children required further decompression, and two of that group also required a shunt. Twelve patients underwent cervical fusion and four had to have a transoral decompression to relieve brainstem pressure.

There have now been several very large patient series published which give a pretty good picture of both the average and extremes of the surgical experience. What is needed now is a structured effort to improve the outcomes and experiences of Chiari patients who undergo surgery.

Table 2: Associated Diagnoses in 500 Pediatric Chiari Patients

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transoral - surgical procedure performed through the mouth

vertebra - individual bony segment of the spine

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

Diagnosis	Number With
Hydrocephalus	48
Neurofibromatosis I	25
Growth Hormone Deficiency	21
Basilar Invagination	15
Klippel-Feil	15
Pseudo tumor cerebri	10

Table 3: Extent of Tonsillar Herniation in 500 Pediatric Chiari Patients

Extent	Number	%
FM - C1	110	22
@ C1	187	37.4
@ C2	197	39.4
@ C3	6	1.2

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Source

[Institutional experience with 500 cases of surgically treated pediatric Chiari malformation Type I](#). Tubbs RS, Beckman J, Naftel RP, Chern JJ, Wellons JC 3rd, Rozzelle CJ, Blount JP, Oakes WJ. J Neurosurg Pediatr. 2011 Mar;7(3):248-56.

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