Key Points

1. Study reviewed 130 children with Chiari treated over a 23 year span.
2. Most common symptoms were head/neck/back pain and scoliosis.
3. 58% of the children also had a syrinx.
4. Surgery provided some measure of relief in 83% of the patients.
5. Head/neck/back pain was not improved in 12% of the patients; scoliosis was not improved in 17% of the patients.
6. 2.3% of the patients experienced surgical complications.
7. 9 children required additional surgery.
8. Amount of tonsillar herniation did NOT correlate with symptoms or surgical outcome.

Definitions

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

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

dura - thick outer layer covering the brain and spinal cord

duraplasty - surgical procedure where a patch is sewn into the dura

dysesthesia - unpleasant abnormal sensation

dysphagia - trouble swallowing

foramen magnum - opening at the base of the skull, through which the spinal cord passes

hydrocephalus - condition where there is an abnormal collection of CSF in the skull area

idiopathic growth hormone deficiency - abnormally low level

What Can Be Learned From 130 Chiari Kids?

If you're the parent of a child with Chiari, have you ever wondered how your situation compares to others? Here's a chance to find out. In one of the largest - if not the largest - reviews of its kind, Dr. Tubbs, Dr. McGirt, and Dr. Oakes, from the University of Alabama at Birmingham, reviewed their experience with 130 pediatric patients who underwent surgery for Chiari malformations over a 23 year time span. They reported the details of their exceptionally large patient series in the August issue of the Journal of Neurosurgery.

The children described in the study revealed a broad range of demographics, symptoms, and associated conditions; yet in aggregate, also begin to define norms for most common symptoms, surgical success rates, and the relationship between Chiari and syringomyelia. The children ranged in age from 2 months to 20 years with an average age of 11 years, with 69 males and 61 females. All patients underwent at least one surgery, with an average hospital stay of 2.7 days.

As would be expected, the most common symptoms included head/neck/back pain, headaches, abnormal nerve sensations, and scoliosis (see Table 1). However there was also a wide variety of less common symptoms, including irritability, facial numbness, trouble walking, hoarseness, rage attacks, and even chronic hiccups. The researchers point out that with the increased availability and use of MRI, Chiari malformations are being diagnosed at younger ages, sometimes even before a child can adequately verbalize their symptoms. In a truly symptomatic case, the only symptom a very young child may show is irritability. On the flip side, generalized symptoms like irritability can have many causes and an MRI may show a malformation that may not be causing any symptoms. The authors suggest doctors should not assume every malformation requires surgery and should strive to exclude all possible non-Chiari causes of a young child's symptoms.

Of the 130 children, in 26 patients the cerebellar tonsils were descended to a point between the foramen magnum and C1, in 61 patients the tonsils were at C1, in 42 patients they were at the C2 level, and in one patient the tonsils were descended to C3. Interestingly, the study found that the magnitude of the herniation did not correlate with either symptoms or surgical outcome.

58% of the children had syringomyelia; this is in line with previous research which has placed the rate of SM in CM patients between 50%-75%. In addition to SM there were a range of associated conditions, with the most common being ridging of the frontal skull bones (metopic ridging), hydrocephalus, and neurofibromatosis (see Table 1). Although these were the most common associated conditions, others ranged from epilepsy to cerebral palsy to Chron disease. The authors identified seven children who exhibited low levels of growth hormones and speculate this may be related to the development of a small posteria fossa (back of the skull) which some researchers believe is essentially the cause of Chiari malformations.

Every patient in the study underwent a decompression surgery that included a craniectomy, laminectomy, and duraplasty. Some patients also had their tonsils partially burned away. In the first 26 patients, a shunt was also placed out of the fourth ventricle. The overall success rate was good, with 83% of the patients experiencing some level of relief [Ed. note: this is another example where the definition of success is too vague and probably not sufficient from a patient's point of view]. It appears as if the operative technique of the surgeon improved over time because the patients with the shunt did not fare as well as the ones without. Of the primary symptoms, headache and neck pain did not resolve in 12% of the cases and scoliosis did not improve in 17% of the cases. The authors note that scoliosis with an initial curve of greater than 40 degrees was less likely to improve than more mild curves.

Of the entire group, surgical complications occurred in 2.3% of the patients and included hydrocephalus and one case of severe, life-threatening brainstem compression which was treated immediately with surgery. Overall, nine patients had to undergo additional surgeries; eight for syrinxes that didn't reduce and one for persistent headaches. The patient who had persistent headaches had not received a duraplasty in the initial operation and the headaches went away after a duraplasty was performed in a second operation. For the group with persistent syrinxes, only one required the insertion of a shunt and the rest responded well to re-operation. The doctors noted that in every case of re-operation for persistent syrinx, CSF was not flowing out of the fourth ventricle the way it should.

So what can be learned from 130 Chiari kids? That Chiari and syringomyelia are complex conditions that can present with a wide variety of symptoms and associated conditions, yet at the same time exhibit a high level of commonality among patients. In addition - and perhaps more importantly - in the hands of a skilled, experienced surgeon, decompression surgery which includes a duraplasty, can help a majority of people with fairly low surgical risk.

Table 1
Most Common Symptoms

of growth hormones due to an unknown cause

Klippel-Feil syndrome - congenital condition where 2 or more cervical vertebra are fused together; often associated with other neurological conditions

laminectomy - surgical removal of part (the bony arch) of one or more vertebrae

metopic - relating to the forehead or front part of the skull

neurofibromatosis - set of genetic disorders in which tumors grow on different types of nerves, bone and skin; type 1 is characterized by spots on the skin

suboccipital craniectomy - surgical removal of part of the skull, or cranium, in the back of the head, near the base

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

ventricle - a cavity in an organ, the fourth ventricle is a space in the brain where CSF collects

Source

Tubbs RS, McGirt MJ, Oakes WJ; Surgical experience in 130 pediatric patients with Chiari malformations. Journal of Neurosurgery 99(2); August 2003, pg 292.

Symptom	# of Patients
Head/neck/back pain	55
Migraine like headache	28
C2 dysesthesia	26
Scoliosis	23
Pain/weakness/ numbness in arms/hands	22
Trouble swallowing	20

Note: *Most patients suffered from more than one symptom*

Table 2
Most Common Associated Conditions

Condition	# of Patients
Metopic ridging	20
Hydrocephalus	14
Neurofibromatosis type I	7
Idiopathic growth hormone deficiency	7
Klippel-Feil syndrome	7