Key Points

1. Chiari I in young children has not been characterized to the extent that Chiari II and spina bifida have been
2. Study examined the records of 31 children - under the age of 6 - with Chiari 1
3. Most common symptoms were oropharyngeal problems, scoliosis, and head/neck pain
4. 69% of the children under 3 years of age had oropharyngeal problems
5. Surgery improved symptoms in 92% of the patients
6. 3 out of 25 patients required re-operations

Definitions

aspiration - act of inhaling

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 - 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 - common term for any of several variations of a surgical procedure to alleviate a Chiari malformation

dura - thick outer layer covering the brain and spinal cord

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

dysphagia - trouble swallowing

gastroesophageal reflux (GER) - reflux of stomach contents into

Trouble Swallowing May Signal Chiari In Children

In young children, Chiari II is very often associated with spina bifida. Spina bifida is one of the most well known and highly researched birth defects. The Medline database lists over 6000 entries relating to spina bifida. In contrast, Chiari I in very young children has not been extensively studied or described. While spina bifida is a very serious condition, it is also important to be able to identify Chiari I in young children, who may not be able to communicate very well, so that proper treatment can be applied before any neurological problems become severe or permanent.

In a study published in the December, 2002 issue of the journal Pediatrics, Dr. Greenlee, Dr. Menezes, and their colleagues, describe their experiences with 31 pediatric Chiari patients under the age of 6. The group reviewed the medical, radiological, and treatment records of the 31 children, who had been treated at the University of Iowa hospital system between 1987 and 2001.

The patients ranged in age from just a few months to almost 6, with an average age of 3. The researchers divided the children into 2 groups: 16 children under the age of 3 and 15 children between the ages of 3 and 6. The most common symptoms (see Fig. 1) were oropharyngeal problems (see Fig 3), headaches, neck pain, and scoliosis. Other symptoms included problems walking, sleep problems, sensory disturbances, and developmental delays. Neurologically, the most common finding was abnormal reflexes. Fifty-two percent of the children also had syringomyelia, and every child with scoliosis also had a syrinx. Interestingly, the level of herniation was not related to either symptoms or the presence of a syrinx. Twenty-six of the children were available for long-term follow-up, with an average follow-up time of almost 4 years.

Surprisingly, 11 out of the 16 children under 3 (69%) presented with oropharyngeal problems. Many of these children were on antacids or other drugs and some had undergone multiple gastrointestinal procedures. While some researchers have noted oropharyngeal problems in pediatric Chiari patients, other reports did not find many problems of this type, and no other series have reported this level of oropharyngeal problems. The authors point out that there are many causes of oropharyngeal problems in young children, but do recommend physicians consider Chiari in cases of severe reflux, failure to thrive, and breathing problems.

For the children between 3 and 6, scoliosis and head/neck pain were the primary symptoms. This is less of a surprise as the link between syringomyelia and scoliosis is well established and a cough/strain headache is the hallmark Chiari symptom in adults. In fact, an NIH study concluded that a cough headache was the best clinical predictor of CSF blockage.

Twenty-five of the patients underwent corrective surgery, which entailed a craniectomy, laminectomy, duraplasty, and shrinkage of the tonsils. In addition, 4 of the syringomyelia patients had shunts placed to help with CSF flow. There were only minor complications from the surgery and overall results were very good (see Fig. 2). Symptomatically, 92% of the children improved, with 46% experiencing complete symptom resolution. With regard to neurological findings, 73% of the patients experienced improvement while 27% remained unchanged. For the children with syrinxes, 1 resolved completely, 10 improved, and there was no change in 1 (MRI's were not available for 4). It should be noted that as has been reported elsewhere, the decompression surgery was very effective in treating the scoliosis related to syringomyelia. Unfortunately, 3 of the children had to undergo additional surgery due to recurring symptoms; however, this initial failure rate - 12% - is in line with other reports.

Oropharyngeal problems can appear in a variety of ways, including chronic cough, reflux, trouble swallowing [Ed. Note: Right before my surgery I was having a difficult time eating certain types of food because of problems swallowing], hoarseness, and poor weight gain just to name a few. It is important for both doctors and parents to note that in very young children, Chiari may not manifest with the classic headaches, but rather with some of the symptoms described above. And it goes without saying that the earlier a Chiari malformation is identified, the better.

Figure 1
Primary Symptom

Symptom	# of Patients <3Yrs Old	# of Patients >3Yrs Old
Oropharyng problem	11	0
Scoliosis	2	5
Headache or neck pain	2	5
Sensory deficit	0	2
Motor deficit	0	1

the esophagus

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

MRI - Magnetic Resonance Imaging; diagnostic device which uses a strong magnetic field to create images of the body's internal parts

oropharyngeal - having to do with the throat or throat area

spina bifida - myelodysplasia; birth defect where part of the spinal cord develops outside of the body

stridor - noisy breathing which indicates a problem

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

Source

Dr. Greenlee, Dr. Menezes, et al. Chiari I Malformation in the Very Young Child. Pediatrics. 2002 Dec;110(6):1212-9.

Other	1	2

Figure 2
Surgical Outcome

Category	Outcome	%
Symptoms	Resolved	46%
Improved	46%	
Unchanged	8%	
Neuro Findings	Resolved	31%
Improved	42%	
Unchanged	27%	

Figure 3
Example Oropharyngeal Problems

- Aspiration
- Reflux
- Choking
- Dysphagia
- Stridor
- Chronic cough
- Hoarseness
- Poor weight gain