

Definitions

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 which surrounds, and protects, the brain and spinal cord

cervical - the upper part of the spine; the neck area

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

diplopia - double vision

Epstein-Barr virus - virus which causes mono

esotropia - when one or both eyes turns inward towards the nose; cross-eyed

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

magnetic resonance imaging (MRI) - diagnostic test which uses a large magnet to create images of internal body parts

nystagmus - involuntary, rapid eye movement

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

syrinx - fluid filled cyst in the spinal cord

vertebra - segment of the spinal column (see [Spinal Anatomy](#))

Chiari Siblings; Spontaneous Resolution

Case Studies is a feature designed to highlight interesting patient cases reported in the research. Given the lack of knowledge about CM/SM, much of the published research comes in the form of case studies - doctors describing one or two patients they have seen and treated - as opposed to rigorous scientific studies. While this type of publication doesn't advance the scientific cause as much, it does give us a window into some of the issues surrounding CM/SM, including lasting side effects and related conditions. And hopefully, some of our readers will say, "Hey, that's just like me!" and know they are not alone in what they are going through.

Familial Chiari Type I Malformation

Authors: S George, AB Page

University/Hospital: Royal Victoria Hospital, Belfast UK

Journal: Eye (advance publication on-line)

Introduction: Family clusters of Chiari have been reported both in the US and Japan. This is the first report of siblings in the UK with Chiari.

Patient 1: 49 year old man went to an eye clinic in 1996 with double vision, headaches, vertigo, and left arm pain. Exam revealed even more eye problems, such as nystagmus. Neurological exam revealed minor problems. MRI however and blood tests were normal. In 1997 the man developed worse eye problems and had trouble walking. Repeat MRI revealed a Chiari to the level of C2.

Treatment: Decompression surgery.

Outcome: Significant improvement in symptoms. Some eye problems returned however and required eye surgery. Man was asymptomatic after eye surgery.

Patient 2: 52 year old woman, the older sister of Patient 1, went to a neurologist in 1995 with shaking and weakness in her left hand. Neurological exam and CT scan were normal. She was given a medication to control the shaking. Symptoms progressed and in 1998 she was found to have decreased sensation and abnormal reflexes. MRI revealed a very large syrinx plus Chiari.

Treatment: Decompression surgery.

Outcome: Initial improvement in symptoms, however shaking returned along with other symptoms. Repeat MRI showed good decompression, but the syrinx had not fully resolved.

Author's Discussion: Family cases of Chiari have been reported since 1990, including a study by Speer which examined 28 pairs of siblings. Milhorat's landmark study showed that 12% of 364 Chiari patients had at least one close relative with Chiari and/or syringomyelia.

Editor's Discussion: In addition to adding to the growing evidence that some percent of Chiari cases have a genetic component, this case highlights the varied ways in which Chiari manifests. In addition to the classic headache, many people experience problems with their eyes and ears. Thus, in order to improve the accurate diagnoses of Chiari, it is important that many different types of doctors be aware of what to look for.

Spontaneous resolution of acquired tonsillar herniation caused by isolated cerebellar tonsil inflammation

Authors: Shokouhi G, Naghili B.

University/Hospital: Tabriz University of Medical Sciences, Tabriz, Iran

Journal: Neurosurgery. 2005 May;56(5):E1158

Introduction: The authors present the case of a young boy whose acquired tonsillar herniation (Chiari) resolved after conservative treatment for the Epstein-Barr virus.

Patient 1: 6 year old boy had headaches in the back of his head for one week, neck pain, and balance problems. Upon examination, he was found to have a fever and sore throat as well. Neurological exam showed abnormal reflexes in his arms and legs. Blood tests showed some irregularities as did a spinal tap. MRI showed that his cerebellar tonsils had herniated to the level of C1.

Treatment: The doctors suspected that the herniation was due to swelling related to the Epstein-Barr virus and chose to treat it conservatively.

Outcome: Three weeks later the boy was asymptomatic. Two months later a follow-up MRI showed the tonsils were higher, but had not returned completely to their normal position.

Author's Discussion: The authors point out that this is a rare complication of the Epstein-Barr virus and go on to emphasize that in cases where the Chiari is thought to be acquired, treating the underlying problem may resolve the Chiari.

Editor's Discussion: Other causes of acquired Chiari include hydrocephalus, cysts, and certain types of shunting. This case highlights the differences between congenital Chiari (meaning you are born with it) and acquired Chiari. In trying to explain why a certain percent of Chiari surgeries fail, some surgeons have focused on whether a different disorder (such as increased intracranial pressure) is at work, meaning a Chiari that was thought to be congenital was really acquired.

--Rick Labuda

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