Key Points

1. Increased use of MRI has resulted in identifying Chiari malformation earlier and in cases where there are no symptoms
2. Deciding whether to perform surgery on border-line cases can be difficult and relies on a surgeon's judgment and experience
3. Balance problems are common among Chiari patients
4. Study involved 77 patients with Chiari who underwent a variety of vestibular tests (VFT)
5. Researchers showed that abnormal VFT results was suggestive of problems caused by Chiari
6. VFT test results can be used to help make surgical decisions in border-line cases

Definitions

audiometric - having to deal with hearing tests

asymptomatic - without symptoms

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

cine MRI - type of MRI which shows CSF flow

decompression surgery - common term for any of several variations of a surgical procedure

Looking To The Ear For Treatment Guidance

As MRI's become more common, Chiari malformations are being identified both earlier and in more people than ever before. While this is undoubtedly a good thing, it also poses some new challenges for the medical community. Tonsillar herniations are being found on people with mild to no symptoms, symptoms that don't seem to relate to Chiari, and even incidental to other medical problems. With no clear, objective test to determine when surgery is necessary, doctors need to rely on their experience and judgment in determining the proper course of treatment for so called border-line cases.

Aggravating the situation is the fact that compression of the cerebellum and brainstem, or abnormal CSF flow, can manifest in a wide variety of symptoms. Some people with large malformations, and even syrinxes, are asymptomatic, while others with mild malformations suffer from a seemingly endless list of woes.

Despite the variety of possible symptoms, many Chiari patients experience similar symptoms as well, such as balance problems. The human balance system - also known as the vestibular system - is a delicate system which involves a number of parts. In order to maintain balance, the body integrates input from three sources: the eyes, muscles and joints, and the inner ear. The inner ear has fluid-filled structures (see Side Bar) which send balance information to - of all places - the cerebellum. Obviously, if the cerebellum is compressed, the system may not work properly.

Dr. Arvind Kumar, a neurotologist at the University of Illinois - Chicago, noticed that he was seeing a number of patients with vestibular problems who turned out to have Chiari malformations. He decided to study these patients, in conjunction with the neurosurgeons there, to see if the information provided by his vestibular testing would be useful in deciding whether to recommend surgery. Dr. Kumar and his colleagues published their results in the September, 2002 issue of the journal Otology and Neurotology.

Dr. Kumar retrospectively reviewed the medical records of 77 patients from his clinic with MRI confirmed Chiari I malformations. The researchers looked at the results of Vestibular Function Tests (VFTs), audiometric tests, MRI's, cine-MRI's, demographic information, symptoms, treatment, and clinical outcome.

Demographically the group was diverse with patients ranging in age from 11 to 67 years old, however there were predominantly more women (56) than men (21). The symptoms for the group overall were typical with imbalance in 45% of the patients, occipital headaches in 36%, dizziness in 32%, and neck pain in 27%, to name a few. Neurologically, 43% of the group showed signs of cerebellar deficit, and more than a third suffered from sensory and strength deficits in their upper body. Forty-four percent of the patients demonstrated some type of hearing problem when tested.

The vestibular testing - a series of 6 tests which involved looking for abnormal eye movements in the vertical and horizontal direction, as a result of head/body position, as a result of irrigating the ears, the ability to visually track objects, and two other tests - revealed abnormalities in more than half of the patient group. In fact, 75% of the group showed abnormal results to the test which involved irrigating the ears.

In order to make sense of the data, the researchers divided the patients into three groups: Group A (19 patients) was comprised of people with moderate to severe symptoms and noted neurological deficits. Many in this group also had a syrinx confirmed by MRI. Group B (34 patients) had relatively mild symptoms that would come and go, and no syrinxes. Group C (24 patients) had symptoms that did not seem to be related to their Chiari malformations, but did have neurological deficits.

In Group A, a remarkable 18 of the 19 patients had abnormal VFT results. In addition, the results suggested a lesion - or problem - in the posterior fossa region. In Group B, the VFT results were abnormal in 82% of the patients. Interestingly, two patients in this group who did not have surgery become asymptomatic, and VFT tests repeated when they were symptom free were normal. In contrast, in both Groups A & B, VFT tests remained abnormal for patients who either declined surgery or did not improve clinically.

In Group C, seven patients (29%) had normal VFT results. Interestingly, 5 of the 7 were determined to have other conditions, such as cervical disc disease that were causing their symptoms. Of the remaining patients, 12 became symptom free, and 5 are still experiencing symptoms. Four of the symptom free patients were retested and their VFT results were normal. VFT results in the symptomatic patients remained abnormal.

Based on these results, vestibular testing appears to be a strong predictor of dysfunction due to a Chiari malformation. While the researchers do not suggest the testing can replace a surgeon's judgment, it does provide the surgeon with more information to make a decision in border-line cases. Specifically, in cases with a demonstrated Chiari malformation, but mild symptoms, abnormal VFT results may add to the case for performing surgery. On the other hand, the researchers recommend that in cases where an MRI shows a Chiari malformation, but VFT tests are normal, patients should undergo thorough testing to determine the cause of their symptoms, as it might not be Chiari.

to alleviate a Chiari malformation

lesion - injury

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

neurotologist - an ear-nose-throat medical specialist who obtains an additional subspecialty in conditions of the ear

nystagmus - involuntary movement of the eyes, can reveal neurological problems

occipital - pertaining to the back part of the head, the occiput bone

otology - scientific study and treatment of the ear

otolaryngologist - doctor who specializes in the ear, nose, and throat

posterior fossa - a depression on the inside of the back, base part of the skull, where the cerebellum is situated

retrospective - type of scientific study which looks back in time, often at medical records

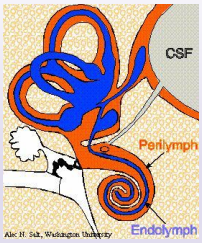
syringomyelia - neurological condition where a fluid filled cyst forms in the spinal cord

vestibular - relating to the balance system; more specifically to the vestibule of the inner ear

The Inner Ear

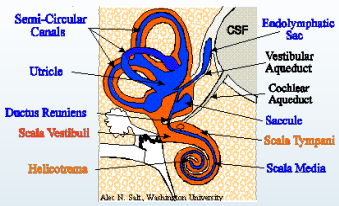
Pictures and Text Courtesy of Alec Salt, Ph.D.; Washington University
http://oto.wustl.edu/cochlea/

Inner Ear Fluids



The anatomy of the inner ear is dominated by large fluid-filled spaces. As shown in this figure, the inner ear consists of a complex series of tubes, running through the temporal bone of the skull. The bony tubes (sometimes called the bony labyrinth) are filled with a fluid called perilymph. Perilymph is shown colored orange. Within this bony labyrinth is a second series of tubes made out of delicate cellular structures (called the membranous labyrinth). The fluid inside these membranous structures is called endolymph, and is shown colored blue. The different spaces of the perilymphatic and endolymphatic compartments are each interconnected by a series of ducts.

Inner Ear Fluid Compartments



The inner ear is comprised of two main sections. The semi-circular canals, utricle and saccule make up the vestibular system and are involved in balance. The scala tympani, scala media and scala vestibuli make up the cochlea which is involved in hearing.