

Key Points

1. Chiari can affect the ears in a number of ways, including tinnitus, feeling of fullness, vestibular problems, and sensorineural hearing impairment
2. Study looked at children who were seen for SNHI and had undergone CT and/or MRI as part of their diagnosis
3. Identified six children with SNHI and Chiari
4. Both the type and severity of the hearing loss varied among the children
5. It is not known if Chiari actually causes SNHI
6. However, theories include Chiari pinching the 8th cranial nerve; directly compressing the auditory processing centers in the brainstem; and altered CSF dynamics
7. Chiari surgery for SNHI is controversial although some successful cases have been reported

Definitions

asymmetric - in terms of hearing, a problem that affects the two ears differently

auditory - having to do with hearing

bilateral - in terms of hearing, affecting both ears

brainstem - lowest part of the brain which connects to the spinal cord and controls basic functions such as breathing and heart rate

cranial nerve - any of 12 nerve pairs that originate in the brain as opposed to the spine

decibel (dB) - unit of measure for how loud sounds are

SNHI - sensorineural hearing impairment; loss of hearing due to problems with the 8th cranial nerve, inner ear, or processing centers of the brain

Chiari Related Hearing Loss In Children

July 31, 2008 -- Much like Chiari can cause different types of headaches, research has also shown that Chiari can affect the ear in different ways as well. It can cause tinnitus (ringing in the ear), the feeling of fullness, and problems with the vestibular system. In fact, one study found that formal vestibular testing was an extremely sensitive test for Chiari, meaning most people with Chiari have problems that show up with vestibular testing. However, vestibular testing is not a good diagnostic tool for Chiari, because it can not determine who has Chiari and who has a different problem which leads to balance issues.

On top of this, Chiari can lead to outright hearing impairment and loss. Milhorat's landmark study found that 36% of Chiari patients exhibited some level of hearing impairment, and other patient series have reported between 10% - 44% with hearing problems. Like many Chiari related symptoms, the exact link between Chiari and hearing loss is not known, and in fact, it has yet to be shown that Chiari is the actual cause of hearing loss in these cases. However, the hearing problems associated with Chiari are considered to be sensorineural (SNHI), meaning that they arise from problems with the eighth cranial nerve, the inner ear, or the brain auditory processing centers, as opposed to the tiny follicles in the ear which are critical for hearing.

As most people know from experience, hearing is usually tested by using a series of tones which a person listens for through headphones. By having the person taking the test indicate when they hear a tone, the range of frequencies and tone volumes which they are able to hear can be mapped out and compared to established norms. An alternative method, which is used for young children who may not be able to verbalize when they hear something, is to measure the actual brainstem response to auditory clicks.

The volume of sound is described in units known as decibels (dBs), with louder sounds at higher decibel levels. The threshold of human hearing is said to start at 0 dB, normal conversation is around 50 dB, and a jet taking off is 130 dB (Figure 1). The European Working Group on Genetics of Hearing Impairment has established the following classifications for the severity of hearing impairments:

- 21 to 40 dB = Mild
- 41 to 70 dB = Moderate
- 71 to 95 dB = Severe
- 95 db and up = Profound

Figure 1:Decibel Value of Common Sounds

10 dB	Breathing
20 dB	Mosquito
40 dB	Whispering
50-60dB	Normal Conversation
80 dB	Garbage Disposal
100 dB	Train
120 dB	Thunder
130 dB	Jet Take-off

Source: National Institute on Deafness and Other Communicative Diseases

In other words, a person with mild hearing impairment can not hear sounds below the 20dB level and someone with profound hearing impairment can not hear sounds below the 95 dB level, which is almost as loud as a passing train. In addition to severity, hearing impairment is also classified by type, meaning whether it affects one or both ears (unilateral or bilateral), and if both, whether they are affected the same or different (symmetric or asymmetric).

Perhaps not surprisingly given the nature of Chiari in general, a report in the June edition of the Annals of Otolaryngology, Rhinology, and Laryngology (Simons et al.) by researchers at Children's Hospital of Pittsburgh, has found that both the type and severity of SNHI related to Chiari vary from case to case. For their research, the doctors reviewed the records of 481 children who had been seen at the hospital's hearing clinic over a five year period for SNHI and for whom either CTs or MRIs were available.

symmetric - in terms of hearing, affecting both ears similarly

tinnitus - ringing in the ear

unilateral - in terms of hearing, affecting only one ear

vestibular - referring to the inner ear/balance system

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

Source

Simons JP, Ruscetta MN, Chi DH, [Senorineural Hearing Impairment in Children With Chiari I Malformation](#), J Otol, Rhin, Laryn (2008), 117(6):443-447.

Using the old but standard 5 mm of herniation as the definition of Chiari, six children out of the 481 were found to have both SNHI and Chiari (Note, this is 1.25% of the SNHI cases, but does not indicate what percent of Chiari cases have SNHI). In the group of six, there were 4 boys and 2 girls; in each case, the hearing impairment was first diagnosed before the age of 9. Interestingly, the researchers found that the types of hearing impairment varied among the six (Figure 2) and included cases of unilateral, asymmetric bilateral, and symmetric bilateral.

Figure2: Hearing Impairment in Six Children With Chiari

#	Type of Impairment	Severity of Impairment
1	Asymmetric, bilateral	Moderate
2	Unilateral	Moderate
3	Unilateral	Mild to Profound
4	Symmetric, bilateral	Moderate
5	Unilateral	Mild to Severe
6	Unilateral	Mild

The details of the auditory testing in these cases revealed no clues about a possible causal link between Chiari and SNHI, however there are several possibilities. One thought is that Chiari either stretches the eighth cranial nerve or pinches it against a bone. This nerve is critical to hearing. Another possibility is that the cerebellum puts pressure on the cells in the brainstem which are involved in auditory processing. Finally, some have speculated that the alteration of CSF dynamics due to Chiari can lead to SNHI. Given the variety of manifestations of SNHI associated with Chiari in just these six cases, perhaps the most likely explanation is that Chiari can affect hearing in a number of different ways.

Although there have been a number of case reports where decompression surgery has improved SNHI in patients, the families of these children all chose not to pursue Chiari surgery, possibly because the SNHI was essentially the only symptom the children had. In fact, there seem to be a number of reports where SNHI is the only symptoms associated with Chiari, it would be interesting to see if this the norm, meaning that when SNHI is present related to Chiari, it is usually the only symptom.

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