Resolution of hydrocephalus-associated sensorineural hearing loss after insertion of ventriculoperitoneal shunt

Case report

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The authors present a pediatric patient with severe hearing loss due to communicating hydrocephalus. This is the first clearly documented case of de novo sensorineural deafness caused by hydrocephalus, with subsequent improvement in hearing after shunt insertion.

The patient initially presented with otitis media and was found to have hearing loss. After reporting ongoing headaches, he received a diagnosis of communicating hydrocephalus, which was treated with the insertion of a ventriculoperitoneal shunt. Formal hearing tests showed dramatic improvement postsurgery; his hearing was normal at 2 months. At 3 years postsurgery the patient’s hearing remains within normal limits.

Hearing loss is a rare complication of hydrocephalus. Based on this case, the authors suggest that the diagnosis of hydrocephalus be considered as a cause of unexplained hearing loss, and conversely, that patients with hydrocephalus might benefit from hearing assessment. (DOI: 10.3171/2009.4.PEDS09103)

Key Words • hydrocephalus • hearing loss • treatment planning • shunt

Abbreviation used in this paper: ICP = intracranial pressure.

Hearing loss is not generally considered a symptom of hydrocephalus. Whereas auditory symptoms have rarely been described in cases of raised ICP, hearing loss has most commonly been associated with CSF shunting and an acute reduction in ICP.1,7 This case is one of documented hearing loss apparently caused by hydrocephalus, and resolution of the hearing loss after shunt placement.

Case Report

History. In this 13-year-old boy, acute otitis media was diagnosed after he presented with severe unilateral ear pain. He described no headache or any other symptoms. A hearing assessment conducted using tuning forks suggested right-sided hearing loss; the hearing loss persisted after the otitis media had resolved. Formal audiological evaluation demonstrated severe right-sided and moderately severe left-sided sensorineural hearing loss (Fig. 1). Tests were repeated at 2 weeks, with good concordance. Results had been normal on a formal audiometry test that had been performed when the patient was 6 years old. There was no medical history relevant to his symptoms.

Over the following 2 months, the patient experienced daily headaches. The headaches were typically bifrontal and frequently associated with dizziness. They often occurred in midmorning, and by midafternoon the patient reported being quite tired. The headaches were occasionally so disabling that the patient could not attend school. He had no tinnitus. There was no associated vomiting, but the patient was intermittently nauseous.

Examination. On examination the patient’s head circumference was 56 cm (> 97th percentile for age), results of limb examination were normal, and cranial nerve examination revealed the hearing impairment and mild papilledema on the right. He had no dysmorphic features.

Neuroimaging. An MR imaging study was suggestive of communicating hydrocephalus, with all 4 ventricles enlarged (Fig. 2). There was no venous thrombosis identified and the vestibular apparatus appeared to be anatomically normal. The inner ear appeared normal (Fig. 3). There was no fluid seen in the middle ear.
Hydrocephalus and deafness

**Operation and Postoperative Course.** The patient underwent insertion of a ventriculoperitoneal shunt for treatment of the hydrocephalus. Postoperatively, his headaches were better, and he reported improved hearing. On Day 2, audiological assessment showed mild hearing loss in the right ear and normal hearing in the left (Fig. 4). Further testing at 2 months demonstrated normal hearing bilaterally. At 3 years postshunting, the patient’s hearing remains normal.

**Discussion**

Mild-to-moderate conductive but not sensorineural hearing loss is a recognized symptom of acute otitis media; it can persist for weeks to months, with an average of 3 weeks, as long as fluid remains in the middle ear. This patient suffered from documented severe sensorineural hearing loss.

Hearing loss is rarely reported as a symptom of either hydrocephalus or benign intracranial hypertension. However, there are a series of unique cases described in the literature that link auditory dysfunction and alterations in ICP. These include hearing loss after ventriculoperitoneal shunt insertion, reduction in hearing after lumbar puncture, and also after spinal anesthesia. In a prospective study, transient but significant hearing loss after neurosurgery was found, suggesting that perilymph pressure is directly affected by the CSF pressure. Conversely, resolution of Ménière-like symptoms, including hearing loss, after shunt placement has been reported in 2 patients with aqueductal stenosis. Tandon et al. investigated the auditory function in a series of patients with raised ICP. They found that 81.5% of the 138 patients investigated had some degree of hearing impairment; 74% of these cases were mild. Some degree of improvement in hearing was observed in 71.5%, with 12.7% showing a significant improvement. Lopponen et al. studied the audiological function in 47 children with shunt-treated hydrocephalus;
38% had sensorineural hearing loss. It is not clear whether the hearing loss was related to the original hydrocephalus or to shunting.

The coexistence of hydrocephalus and altered hearing can be explained anatomically: the inner ear and the CSF spaces are intimately related. A number of mechanisms of pressure homeostasis between the cochlear fluids and the CSF has been described, but the complete picture remains imperfectly understood. The pressure in the perilymph is controlled by the endolymphatic sac, which lies within the subdural space. Although there is usually no direct communication between the endolymph and the perilymph, the surrounding CSF pressure is transmitted to the perilymph. The pressure of the endolymph is controlled by the endolymphatic sac, which lies within the subdural space. Although there is usually no direct communication between the CSF and endolymph, the surrounding CSF pressure is transmitted to the perilymph. The pressure differentials may cause displacement of the Reissner membrane and may affect hearing.

The rarity of overt hearing problems in hydrocephalus raises the possibility of an additional derangement of the normal anatomy or normal anatomical barriers where hearing loss occurs. Walsted,12 in assessing hearing in 126 patients after loss of CSF, noted that not all of the patients had hearing threshold changes. A possible explanation given was that individual factors influence the clinical manifestation of the pathological change; for example, differences in patency and the dimensions of the cochlear aqueduct. This may be congenital, as with an anatomical narrowing, or due to some insult (for example, head injury or infection).

Conclusions

We suggest that acute hearing loss is a rare and probably underestimated complication of hydrocephalus. This case suggests the consideration of hydrocephalus as a cause of otherwise unexplained hearing loss, and conversely, that patients with hydrocephalus might benefit from some form of hearing assessment. This may be of particular importance in the pediatric patient. Doctors providing treatment should consider the possibility of hearing impairment in pediatric cases of hydrocephalus, where self-reporting of poor hearing is inherently low.

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Disclaimer

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References


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