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What is This?
Hydrocephalus-Associated Hearing Loss and Resolution after Ventriculostomy

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Keywords
sensorineural hearing loss, hearing loss, ventriculostomy, hydrocephalus, cochlear aqueduct, intracranial pressure, perilymph, endolymph

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Hydrocephalus is not usually considered in the differential diagnosis of sensorineural hearing loss (SNHL). There have been reports in the literature of sudden SNHL after reduction of intracranial pressure (ICP) via ventriculoperitoneal shunt (VPS).1,2 However, a literature search reveals only 1 study that associates elevated ICP with SNHL 3 and 1 case of hydrocephalus-associated SNHL.4 We present a patient who developed hydrocephalus and SNHL and experienced total resolution of SNHL with treatment of hydrocephalus.

Case Report

Formal review and waiver was conducted by the University of Kentucky Office of Research Integrity Institutional Review Board.

A 14-year-old previously healthy girl presented initially with sudden right hearing loss, tinnitus, aural fullness, and headaches. An audiogram revealed moderate right low-frequency SNHL rising to normal in the higher frequencies. The patient’s left side was within normal range. Two months later, the patient developed left-sided hearing loss and tinnitus and was started immediately on steroid therapy. An audiogram revealed mild left and moderate right up-sloping SNHL (Figure 1). The patient did not experience vertigo or disequilibrium.

On examination, the tympanic membranes were normal bilaterally. The extraocular movements were normal bilaterally with no nystagmus. Cranial nerve and extremity neurological examinations were normal. Romberg, gait, and tandem walk were normal. Magnetic resonance imaging (MRI) obtained after initial onset of right SNHL revealed dilated lateral and third ventricles with a normal-appearing fourth ventricle. There was no evidence of cerebellopontine angle tumor.

The patient was referred to pediatric neurology and eventually underwent ventriculostomy without shunt placement by neurosurgery. At 2-month follow-up, she noted resolution of hearing loss and headaches, and an audiogram revealed hearing within normal limits bilaterally (Figure 2).

Discussion

Transient SNHL is a known sequela after loss of cerebrospinal fluid (CSF) due to procedures such as lumbar puncture, spinal anesthesia, and myelography.1 In addition, there are documented cases of SNHL associated with reduction of ICP with VPS placement.1,2 Lee et al1 describe a case of unilateral SNHL and tinnitus after shunt placement in a patient with a patent cochlear aqueduct on the affected side. The authors postulate that reduction in ICP is transmitted through the cochlear aqueduct to the perilymph, resulting in a relative endolymphatic hydrops similar to Meniere’s disease. Russell et al2 similarly described a patient with bilateral patent cochlear aqueducts who developed delayed SNHL following shunt placement and improved with shunt revision.

Sensorineural hearing loss as a result of increased ICP is a rarely documented entity. Tandon et al3 studied 138 patients with elevated ICP from a variety of etiologies and found that 81.5% had some degree of hearing impairment. Of these patients, 71.5% had improvement in their hearing after operations that resolved the elevated ICP. The only recent documentation of SNHL associated with hydrocephalus is by Sammons et al.4 They present a patient with hydrocephalus and SNHL who had resolution of SNHL after VPS placement.

Tandon et al3 describe mechanisms by which increased ICP results in SNHL. They hypothesize that increased ICP is transmitted to perilymph by the cochlear aqueduct, resulting in a relative endolymphatic hydrops. The altered hydrodynamics results in a SNHL. They also contend that elevated perilymphatic pressure could increase the mechanical load on the stapes footplate and result in a conductive hearing loss.

This case gives merit to the theory of elevated ICP resulting in SNHL due to transmission through the cochlear aqueduct.

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Figure 1. Audiogram prior to ventriculostomy exhibiting mild left and moderate right up-sloping sensorineural hearing loss.

Figure 2. Audiogram 2 months after ventriculostomy revealing resolution of sensorineural hearing loss.
The patient had SNHL and no other explanation for the loss. Her hearing improved with resolution of the elevated ICP.

Based on this patient’s clinical course and others similar to her, changes in ICP should be on the list of differential diagnoses of SNHL. Suspicion should be particularly high in young patients, who are more likely to have patent cochlear aqueducts, and in patients who present with concomitant headaches. Eye examination for papilledema and imaging, which is often obtained anyway for unexplained SNHL, will aide in establishing the diagnosis.

Conversely, patients with hydrocephalus should be evaluated with an audiogram. This is especially important in infants and children, in whom self-reporting is not reliable and hearing is vital for development.

Author Contributions

Joshua F. Dixon, acquisition of data, review and compilation of literature, drafting, revision, final approval of article; Raleigh O. Jones, conception and design, revision and final approval of article.

Disclosures

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