Cerebrospinal fluid shunting and hearing loss

The clinical presentation of hydrocephalus includes increased size of head, seizures, headaches, blurred vision, memory problems, hearing loss, changes in personality, loss of bladder control and precocious or delayed onset of puberty etc [1].

Sensorineural hearing loss (SNHL) in hydrocephalus patients has been reported in the literature and the proposed mechanism hypothesized is that increased intracranial pressure (ICP) is transmitted to the perilymph by the cochlear aqueduct, resulting in a relative perilymphatic hydrops and this hydrodynamics leads onto sensorineural hearing loss [2] [3] [4].

Hearing loss is a potential cerebrospinal fluid shunt complication that may lead to severe impairment and deafness.

Mechanism

Although the mechanism for this is unclear, it is thought that changes in cerebrospinal fluid pressure can affect cochlear physiology via endolymph expansion in the setting of a patent cochlear aqueduct [5].

Several indirect mechanisms may explain the association between hydrocephalus and hearing loss, including mass effect, compromise of the auditory pathway, complications of prematurity, and genetically mediated hydrocephalus and hearing loss. Nevertheless, researchers have proposed a direct mechanism, which Satzer et al. term the hydrodynamic theory. In this hypothesis, the intimate relationship between CSF and inner ear fluids permits relative endolymphatic hydrops or perilymphatic hydrops in the setting of CSF pressure disturbances. CSF is continuous with perilymph, and CSF pressure changes are known to produce parallel perilymphatic pressure changes. In support of the hydrodynamic theory, some studies have found an independent association between hydrocephalus and hearing loss. Moreover, surgical shunting of CSF has been linked to both resolution and development of auditory dysfunction. The disease burden of hydrocephalus-associated hearing loss may be large, and because hydrocephalus and over-shunting are reversible, this relationship merits broader recognition. Hydrocephalic patients should be monitored for hearing loss, and hearing loss in a patient with shunted hydrocephalus should prompt further evaluation and possibly adjustment of shunt settings [6].

Hearing thresholds may increase following VP shunt placement, possibly due to secondary endolymphatic hydrops [7].

Case reports

A case suggests that patients with a ventriculoperitoneal shunt should be monitored for hearing loss. Auditory impairment in these patients should prompt further evaluation and possibly adjustment of shunt settings [8)].

A 77-year-old man with unilateral dominant sensorineural hearing loss after a shunt placement for normal pressure hydrocephalus (NPH) combined with a patent cochlear aqueduct. Based on our experience and a review of the literature, we suggest an early restoration of the reduced CSF pressure using a programmable valve as a treatment strategy, which might prevent the persistent hearing loss [9)].

The case of an elderly physician who endured a slowly progressive, ambulatory illness, which was erroneously diagnosed as Parkinson's disease. After ten years of progressive illness, the correct diagnosis of normal pressure hydrocephalus (NPH) was finally made, revealing itself, by accident, through incontinence and mild dementia. The patient-physician enjoyed an instantaneous remission induced by a large lumbar puncture (LP) sustained by a ventriculosystemic shunt. The patient-physician dedicated his renewed life to informing the medical profession about this dramatic syndrome, which he believes is more common and more reversible than generally thought. Although the patient had been virtually restored to normal, a series of complications typical of ventriculosystemic shunting (VSS) occurred, including significant hearing loss and subdural haematoma (SDH). The patient feels, however, that his clinical improvement far outweighs the complications and that every patient with NPH should have the opportunity to decide whether or not to have a VSS [10)].

A case of intracranial hypotension syndrome due to overdrainage of cerebrospinal fluid presented with hearing loss after ventriculoperitoneal shunting procedure. A 69-year-old man suffering from subarachnoid hemorrhage presented with an angiogram showing two aneurysms, one of the right internal carotid and one of the middle cerebral artery. Neck clipping was performed. One month later, he developed normal pressure hydrocephalus (NPH), which was treated by ventriculoperitoneal (NPH), which was treated by ventriculoperitoneal shunting procedure using low pressure Pudenz system. Trias of NPH were improved by insertion of shunt system. However, he complained of hearing loss which was worsened by upright position and improved by lying down. Such kinds of phenomenon were demonstrated by audiogram showing that the transitory decrease of hearing and electrocochleography showing the elongation of N1 latency at upright position. These data suggested that his hearing loss was caused by inner ear or auditory nerve lesion. After the shunt system was replaced into the antisiphon device, his hearing disturbance improved. Axial computed tomography of bone window at the level of orbitomeatal line demonstrated widely perilymphatic duct on both sides. This finding suggested that the fluctuation of intracranial pressure was easily transmitted into the cochlear through the widened perilymphatic duct, resulting in hearing disturbance [11)].

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