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CASE REPORT

Extreme sensitivity of hearing to decreases of ICP in Menière's disease

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Abstract We report the case of a Menière's disease patient affected by normal pressure hydrocephalus (NPH) who presented a cerebrospinal fluid (CSF) pressure-dependent hearing impairment after shunting. This side-effect was not only reversible and reproducible but occurred at a high opening pressure when the valve setting was lowered by only 0.7 mmHg (10 mmH_2O). This observation suggests that hearing in Menière's disease might be very sensitive to small reductions of intracranial pressure (ICP) and that these patients should be informed of this potential risk, which can compromise the efficacy of the shunt.

Keywords Hearing impairment · Ventriculo-peritoneal shunt · Normal pressure hydrocephalus · CSF pressure · Menière's disease

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Introduction

Transient, permanent, unilateral or bilateral hearing impairment has been described after ventriculo-peritoneal (VP) shunting [5, 8, 10, 12, 14]. The probable mechanism is the decrease of the cerebrospinal fluid (CSF) pressure consecutive to the CSF loss [8, 10, 12, 14].

We report the case of a patient suffering from Menière's disease who presented, subsequently to VP shunt insertion for normal pressure hydrocephalus (NPH), a non-postural bilateral hearing impairment. The manifestation and resolution of the latter was dependent on the opening pressure value of the implanted programmable valve. The striking particularity of this case was that the hearing deficit appeared and disappeared in the high opening pressures range after a minute change of the valve setting.

An attempt at explaining the possible underlying pathophysiological mechanisms of the particular sensitivity to post-shunting altered hearing in Menière's disease patients is made.

Case report

A 55-year-old woman suffering since 1993 from Menière's disease presented in 2003 with the classic clinical triad of NPH comprising gait disturbance, memory impairment and urinary incontinence. Based on the clinical picture, radiological imaging (CT and MRI) and a determination of the resistance to CSF outflow performed in 2005 by a lumbar infusion test ($R_{CSF} \geq 20$ mmHg/ml/min), the diagnosis of NPH was made and she underwent VP shunt insertion in 2006, with the placement of a Codman Hakim programmable valve with antisiphon device set initially at 70 mmH_2O, according to the baseline ICP of 6 mmHg (82 mmH_20)

measured at the time of the infusion study. The patient complained of headache in the erect posture one day after the operation and the valve was adjusted to 90 mmH_2O, which made the headaches disappear. However, 2 days after the shunt implantation she noticed a disabling bilateral hearing decrease, proving to be non-dependent of the posture. A CT scan excluded over-drainage. The opening pressure of the valve was, therefore, further increased in a stepwise fashion up to 160 mmH_2O where the patient did not perceive any amelioration of the hearing impairment yet. However, when the valve was set at 170 mmH_2O she noticed a frank improvement in hearing that subjectively returned to the level of her pre-operative perception. It deteriorated again when the valve was lowered to 160 mmH_2O, but improved when it was reset at 170 mmH_2O. This course was documented by repeated audiograms (Fig. 1). The patient's gait was nevertheless significantly better improved at 160 mmH_2O than at 170 mmH_2O, but the lady was so disabled by the hearing loss that she preferred remaining with the 170 mmH_2O setting to the detriment of NPH treatment.

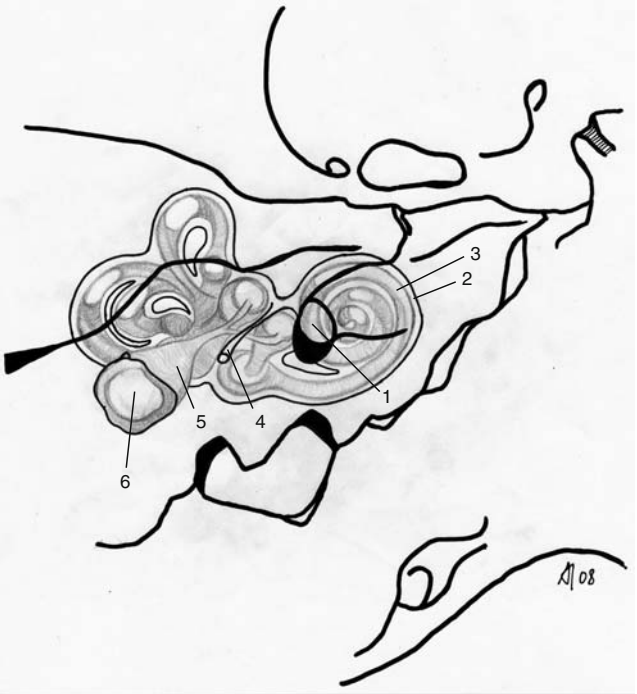

Fig. 2 Schematic diagram illustrating the postero-medial view of a left petrous bone. *1* Inner auditory canal, *2* perilymphatic space, *3* endolymphatic space, *4* CA, *5* endolymphatic duct, *6* endolymphatic sac

Discussion

If hearing impairment subsequent to shunt placement for hydrocephalus has already been reported in the literature, the notable particularity of the present case is the triggering and cessation of the phenomenon by a minute adjustment of the CSF pressure and its appearance at a high pressure setting of the valve compared with previous reports of hearing impairment after shunting [8, 10, 12, 14]. Indeed, the reported valve pressures triggering a hearing impairment were always lower than the pressure setting provoking the deficit in the present case, and in the previously published cases the hearing impairment improved after revising the shunt with a high opening pressure valve [8, 12].

The decrease of CSF pressure caused by the CSF loss has been looked upon as the cause of hearing impairment that was supposed to be an effect of pressure transmission between the subarachnoid space (SAS) on one side and the inner ear on the other side [3, 8, 10, 11, 12, 14, 15]. The inner ear itself, however, houses two fluid spaces, the perilymphatic and the endolymphatic compartments. The former communicates directly via the cochlear aqueduct (CA) with the SAS of the posterior cranial fossa and the latter communicates indirectly with the SAS via the endolymphatic duct and the endolym-

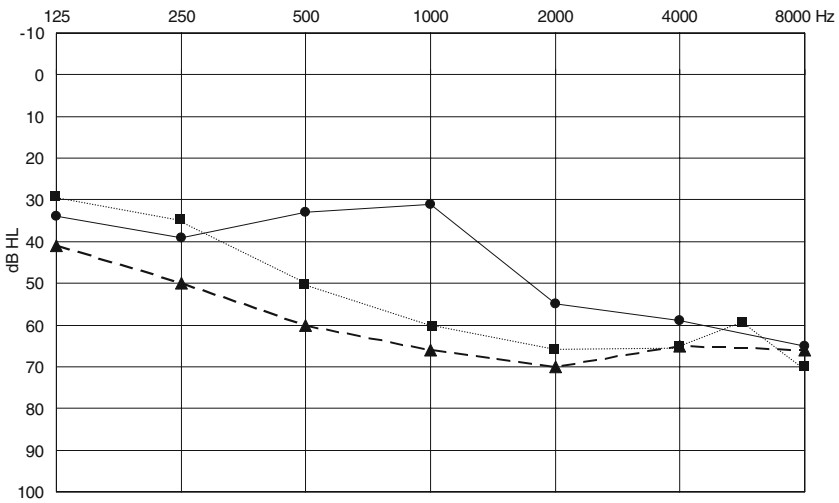

Fig. 1 The right ear pure tone hearing thresholds 10 years before VP shunting (●), after shunt placement at the opening pressure of 170 mmH_2O (12.5 mmHg) (■) and at the opening pressure of 160 mmH_2O (11.8 mmHg) (▲). The left ear pure tone hearing thresholds were almost identical

phatic sac situated in the subdural space (Fig. 2). The lumen of the CA may be patent, partially or completely occluded, while its width is inconstant [2, 7, 13]. This might explain why in previous reports patients had unilateral, bilateral or no hearing impairment [8, 10, 11, 12, 14]. However, relating hearing impairment after procedures provoking CSF loss uniquely to the patency of the CA does not consider that after the occlusion of the CA the pressure transmission between the CSF and perilymphatic spaces persists, albeit smaller and delayed, which does not seem to result in inner ear dysfunction [2, 3]. Hearing impairment may, however, more likely result from the apparition of a disequilibrium between the pressures present in the inner ear compartments. In fact, a change in CSF pressure may more strongly influence the perilympahitic pressure, owing to the direct communication between the CSF space and the perilympahitic space, than the endolympatic pressure, which is only indirectly influenced by the CSF pressure through the subdurally located endolymphatic sac. An imbalance of magnitude and/or phase may, therefore, happen between the pressures of the perilymphatic and endolymphatic spaces and, as equal perilymphatic and endolymphatic pressures are necessary to ensure the normal transduction of sounds by the endolymph-bathed organ of Corti, result in hearing impairment.

The overall particularity of the present case, compared with the previously published cases, is the extreme sensitivity of the patient's inner ears to a pressure decrease. Although the patient already had an audition deficit, a subjectively noticeable and objectively confirmable worsening happened at the high pressure threshold of 160 mmH_2O (11.8 mmHg). Moreover, the reproducible trigger was a minute ICP change of only 0.7 mmHg (10 mmH_2O), from 170 mmH_2O to 160 mmH_2O. These differences are most likely explained by the pre-existence of the associated inner ear disorder in this particular patient.

The classic triad of Menière's disease includes recurrent vertigo, fluctuating sensorineural hearing loss and tinnitus [6]. Despite a rich literature, the definitive cause and the pathogenis of Menière's disease have not been identified yet. However, although the exact patho-physiology of Menière's disease in humans is still unsolved, the presence of an endolymphatic hydrops is generally accepted as the most important pathologic alteration, and the decoupling of the pressures ratio resulting in an endolymphatic hydrops seems to play an important role in guinea pigs [1, 9]. As shearing movements between the basilar and the tectorial membranes are necessary for the conversion of mechanical into electrical energy and hence audition, the enlargement of the distance between the basilar and the tectorial membranes by the hydrops may explain the hearing impairment in Menière's disease [4, 11, 14, 15]. The marked CSF pressure sensitivity and dependence of the hearing of our patient may be related to the pre-existence of a hydrops. In fact, when an endolymphatic hydrops is present, the importance of preserving the perilymphatic pressure to stick the basilar membrane onto the tectorial membrane might increase. Thus, the apparition of a minor imbalance between the perilymphatic and endolympatic pressures will have a more pronounced detrimental effect than in normal ears on the transduction of sounds.

Conclusion

The present case suggests that hearing in Menière's disease patients may be extremely sensitive to CSF pressure decreases, notably after VP shunting. This observation should render neurosurgeons aware of this disabling side-affect when considering shunting in patients with a pre-existing Menière's disease and, consequently, these patients should be informed of this potential risk. Indeed, as in our case, this side effect may quite compromise the efficacy of the shunt on NPH symptoms and renders NPH treatment difficult. As the hearing impairment could be reversed by slightly raising the CSF pressure, the implantation of a programmable valve is advocated for patients needing a VP shunt but also suffering from a pre-existing Menière's disease.

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