THE RELATIONSHIP BETWEEN PERILIMPH AND CEREBROSPINAL FLUIDS IN THE MENIERES DISEASE. NEW FINDINGS IN RMI AFTER GADOLINIUM INTRA-TYMPANIC ADMINISTRATION

Giampiero Neri
Zou J et al. In vivo visualization of endolymphatic hydrops in guinea pigs: magnetic resonance imaging evaluation at 4.7 tesla 2003


Naganawa S, et al Separate visualization of endolymphatic space, perilymphatic space and bone by a single pulse sequence; 3D-inversion recovery imaging utilizing real reconstruction after intratympanic 2008


Naganawa S, et al MR imaging of Ménière’s disease after combined intratympanic and intravenous injection of gadolinium using HYDROPS2 2014
What we know today about the characteristics of the perilymph and endolymph is that the two liquids have different composition, as clarified by Smith and a different electric potential, as clarified by von Bekesi (endococlear potential = 85 mV).

Regarding the perilymph we also know that the characteristics of scala tympani and the scala vestibuli are different so as to suggest two different origins; in fact, in addition to having different ionic concentrations between them, radioactive tracers injected into the vestibular scale not be traced in the scala tympani and the presence of K-glucose - protein is greater in the vestibular scale than in the scala tympani.
The perilymph is produced both from plasma, that filters slowly through the blood-labyrinth barrier and from the CSF, that communicates with the cochlear perilymph through the cochlear aqueduct, but is not the major source of perilymph (Zou 2003).
The perilymph is removed both through the production of endolymph and its excretion by the cochlear aqueduct toward the cerebellar angle (cisterna magna). The endolymph, produced by the transepithelial exchange, is transported towards the ES where will be reabsorbed.
Considering the difference between the endolymphatic and perilimphatic spaces, while the smaller amount of endolymph communicates with the outside of the system through the bigger way (VA), a bigger amount of perilymph communicates through the narrow cochlear aqueduct considered at the same time the gateway and the way out of the perilymph.
Cochlear aqueduct
The critical point

- Discovered by Du Verney (1683) and described in detail by Cotugno (1761).
- A narrow canal between subaracnoid space and internal ear.
- It communicates with the scala tympani (Ghiz et al., 2001).
- In human it is 1 cm long and with 0.14 mm minor diameter.
- Its endocranial opening is funnel-shaped. (Gopen et al., 1997).
- It contains a vein draining from the cochlea into the internal jugular vein.
- The csf enters the duct with speed of 30 nl/min pushed to the pulse of breath (Salt 2015)
The cochlear aqueduct is really important for the perilymphatic flow?

- The closure of the cochlear aqueduct is a physiologic age-related event (Marchbanks RJ, 1990).
- The cochlear duct is almost always a virtual duct (Jackler 1993).
- In human adult, the cochlear aqueduct is (Naganawa 2008):
  - Completely closed in the 7% of the general population,
  - Filled by connective tissue in the 59%;
  - Its central lumen is patent only in the 34%.
- Others possible communications between the CSF and the perilymph are the modiolus pores and the perineural spaces in the singular nerve canal (Naganawa 2008, Yamazaki 2012).
- In Meniere’s patients, after its intratympanic injection, with RM 3 Tesla the gadolinium was observed in the IAC in the 100% of patients and not in the cochlear aqueduct (Naganawa 2008).
Useful to demonstrate the anatomical features of inner ear related to perilymph
<table>
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<tr>
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<th>Seo Y.J. 2012</th>
<th>Our experience 24 pz</th>
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<tbody>
<tr>
<td>No response</td>
<td>20%</td>
<td>17% (4/23)</td>
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<tr>
<td>No diffusion in semicircular canals</td>
<td>53%</td>
<td>47% (8/19)</td>
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<tr>
<td>Cochlear response</td>
<td>81%</td>
<td>63% (12/19)</td>
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<tr>
<td>Utricolo-saccular response</td>
<td>69%</td>
<td>84% (16/19)</td>
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<tr>
<td><strong>Cochlear duct absent</strong></td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td><strong>Diffusion of Gd in the Internal auditory canal</strong></td>
<td>98% (naganawa)</td>
<td>100%</td>
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The diffusion of gadolinium from the IAC in Meniere’s patients could be considered as a labyrinthine fistula sensible to the pressure changes of the CSF?
The CSF is contained in the ventricles (25%) and in the spinal and cortical subarachnoid spaces (75%).

Normal CSF pressure is equal to intracranial pressure (ICP), varies from 60 to 250 mm H_2O in healthy adults, and fluctuates during the day because of many factors.

In adults, 500–600 mL of CSF is secreted daily at a rate of 0.3–0.6 mL/min.

The total CSF volume of 160 mL is replaced about four times a day.

A substantial part of water in the brain is drained into the extracranial cervical lymphatics from the Virchow Robin space, and from the CSF space via the perineural subarachnoid space of cranial nerves.
Can CSF generate audio-vestibular diseases?

- **In Eva Syndrome**
  - The increasing pressure of the Vestibular aqueduct (even caused by soft trauma) can lead to a hearing loss.

- **In the Minor Syndrome**
  - Changes of the intracranial pressure can lead to vertigo and oscillopsia (positional)
  - Changes of pressure caused by natural plugging can lead to Meniere-like syndrome (Modugno 2013)

- The link between perilymph and IAC could determine Gusher’s disease during stapedotomy or cochleostomy procedures (Phelps 1991)

- Otoacoustic emissions change after a ventriculoperitoneal shunt due to a “vacuum” of the internal ear (Chomicki 2007)

- A low CSF pressure determines a low perilymphatic pressure with consequent edolymphatic hydrops (Gordon 1984, Walsted , 2002).
The bulging of Reissner’s membrane might be due to differences in hydrostatic pressure between endolymph and perilymph and these pressure changes might be responsible for the hearing impairment that occurs in Menière disease.
Affects young women with obesity, contraceptive use, pregnancies, obstructive sleep apnoea, arterial hypertension (Biousse 2012, Andrews 2014, Wall 2014)

Headaches and papilloedema can be associated with blurred vision, photopsia, or transient visual obscuration, insidious progressive visual loss, which is usually reversible with treatment.

70% of patients with severe or moderately severe Meniere’s disease have headache … It is concluded that MD is associated with headache that can be handicapping. (Eklund 1999)

The neuro-otological symptoms rarely are as a presenting symptom, but these were common when asked to the patients with IIH (Ozer 2013)

‘Congestive inner ear’: otologic signs in patients with increased intracranial pressure (tinnitus, hearing loss and vertigo (Fischer and Wolfson 1943)

The audiograms of IIH patients reported SNHL at low frequencies from 52% to 90% of patients (Saxena 1969, Sismanis 1987)

ECOG analysis of the patients with IIH showed abnormal SP/AP ratio in 74.1% (Ozer 2013)

Acetazolamide, a carbonic anhydrase inhibitor that reduces CSF production and ICP, is the main drug used to treat idiopathic intracranial hypertension (Wall 2014)
Why monitor perilymphatic pressure in Menière's disease?

Marchbanks R¹.

- Tympanic Membrane Displacement (TMD) technique
- Patients with Intracranial hypertension without the usual headache and visual symptoms
  - 62% complained of tinnitus
  - 28% suffered from paroxysmal rotary vertigo
  - In such cases, the patient may be referred to the otolaryngological clinic and the condition may be mistaken for Menière's disease or a labyrinthine disorder.

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*Posted by: deleted_user 6 years ago Mood: Ok*

Anyone here have Menieres disease? I have had Menieres disease for 10 years and was recently diagnosed with Pseudotumor Cerebri. I have read a few items suggesting there is a link between the two. Just wondering if anyone else here has both or has heard or anyone having both diseases.

*deleted_user 6 years ago*

That is an interesting theory. However, I have found my PTC symptoms to be very different than my Menieres attacks. I go almost completely deaf after a Meniere's attack and it takes several days for my hearing to return, whereas my PTC symptoms come on very suddenly, and are usually gone within a couple of days. 

*ategan 12 days ago*

I have been diagnosed with both diseases in the last year. I was told that I had PTC by an and visual disturbances that were not eye related. Diamox dropped my BP too low, so I took constant ringing and fluid in my ears. My ENT did a balance test and hearing test, which I misdiagnosed because people with meniers do not complain of debilitating headaches.
The pathophysiology of Ménière’s Disease is, in some cases, associated with the interconnecting pathways between the inner ear and the intracranial fluid space. It seems probable that the hydrodynamics of the intracranial/labyrinthine interface need to be considered and that the newly discovered intracranial pressure waves will feature in our understanding.
Intracranial hypotension can result from spinal CSF leaks through small spinal dural tears. (Schievink 2008, Brinker 2014, Mokri 2014)

Leakage causes CSF hypovolaemia, that causes pachymeningeal thickening, pituitary hyperaemia, venous engorgement, endolymph hypotension, and downward displacement of the brain, with sagging of the cerebellar tonsils and brainstem (Mea 2012)

A Menière’s disease patient affected by normal pressure hydrocephalus presented after shunting a reversible and reproducible CSF pressure-dependent hearing impairment in Menière’s disease hearing might be very sensitive to small reductions of intracranial pressure (Kurzbuch, 2009)

In at least 50% of cases the hypotension is associated with cochlear-vestibular signs include tinnitus, ear fullness, echoing or distortion of sounds, hypoacusia, dizziness, or even rotational vertigo (Fontaine 2012)
In the Menière’s disease the attention was always focused on the endolymph while the perilymph was considered as a fluid adapting to endolymphatic modifications.

The GD leaks in the IAC demonstrates

- A different cochlear physiology (or pathophysiology) in which the IAC is the place of the perilymph dispersion
- A continuous flow that leads to the diffusion of the GD from the round window to the IAC exists almost in the patients with Menière disease.

The changes of pressure of the CSF, modifying the perilymphatic pressure, could play a role in determining or maintaining the Meniere’s disease.

It’s not a new theories but a new approach to pathophysiology that starts from the IAC, largest and most direct, and not from the CA.

Would be useful to investigate about the intracranial pressure when the Menière disease is suspected.

More studies are needed.