ENDOLYMHPHATIC HYDROPS

(ELH, MENIERE’S DISEASE)

The inner ear hearing and balance mechanisms resemble convoluted tubes of membranes immersed in fluid within the otic capsule, the bony casing of the inner ear. The tubules are filled with endolymph, fluid of a consistency necessary for the functions of the inner ear. Outside the tubules, the structures are bathed in perilymph, which closely resembles the cerebrospinal fluid around the brain and which differs substantially from the endolymph.

To maintain its consistency, the endolymph is continually formed within the inner ear, then drained away via a small out-pocketing: the saccus endolymphaticus, which absorbs the fluid back into the bloodstream.

Meniere's Disease: The swollen inner ear

For reasons uncertain, the saccus drainage may fail. When this occurs, the membranous inner ear structure swells, and may rupture. The swelling produces episodes of typical symptoms (Otalgia).
Characteristics

The condition is typified by episodic severe distress (Acute Otitis Media). Initially, Low frequency nerve deafness may be mild and fluctuant, becoming more severe with time, possibly to total loss. A feeling of fullness, pressure or blockage is felt in the affected ear. Humming or buzzing tinnitus may be severe, worse with the episodes. Severe rotatory vertigo may cause nausea, vomiting and prolonged prostration. Diarrhoea is not uncommon. Later, the condition may develop drop attacks – sudden collapse, without warning, and without loss of consciousness.

The cause of the hydrops problem is uncertain; it is unknown whether the problem is a unique condition or, alternatively, common to several causes (e.g. like a headache). This impacts on the ability to formulate and target treatment adequately.

Treatment

Medical management may aim to treat the cause or relieve the symptoms. The former employs diuretics (Moduretic, urea) or vascular agents (Serc). Others settle dizziness or nausea (Stemetil, Zofran).

Dietary reduction of salt intake appears to be beneficial, but as many foodstuffs have considerable covert salt content, a limited-salt diet is best accessed via a specialist dietitian.

Several office procedures may be helpful. Grommet insertions have been used in the past, although with uncertain efficacy.

More commonly, middle ear perfusions are used to control symptoms. Filling the middle ear with steroid solutions is reported as being beneficial in earlier cases, although further verification is needed.

More often, gentamicin perfusions are used. Gentamicin is a common antibiotic that has a toxic effect on the balance function of the ear. If the middle ear is flooded with a gentamicin solution, some crosses into the inner ear, reducing the vertigo produced by the ELH. Several perfusions may be needed, performed with little discomfort under local anaesthetic as an office procedure, usually employing a vent tube. The aim of the procedure is to produce mild unsteadiness (dysequilibrium) that comes on in the days after the perfusion, and after which rotatory vertigo generally diminishes. Further perfusions are given if the vertigo persists. The technique is effective in over 80% of cases, although some require a “top-up” many months later.

The downside of perfusions is the degree of dysequilibrium encountered. This balance upset is due to reduction of balance function in the ear treated, and may be severe in some cases, especially in females, where the lighter bodily tissues presumably admit more of the drug into the inner ear. This more severe reaction may take 6-8 weeks to clear; the patient needs to know this risk prior to treatment. Fortunately, severe reactions generally herald a complete cure of the rotatory vertigo.

A further complicating factor arises if the patient is unable to distinguish between the treatment-induced dysequilibrium and another attack of true ELH: the surgeon may be uncertain whether to administer a further dose that may exacerbate matters.

Gentamicin may also damage the hearing. The effect of perfusions in this respect is uncertain, as active ELH also causes progressive deafness.
Surgery

Operative intervention may be employed in several roles.

i. Saccus Decompression/Drainage.

Removal of the mastoid bone and its air cells from over the saccus endolymphaticus relieves or cures ELH in up to 70% of cases. The surgery is controversial, as the exact beneficial action is uncertain. The efficacy of sac drainage undertaken during this surgery is also questionable. When successful, symptoms are alleviated and hearing stabilised, although cases may relapse later. This is often the treatment of choice in early cases, in order to retain hearing.

ii. Section of the Eighth Nerve.

Neurosurgical-type division of the nerve attempts to halt the dizziness without loss of hearing. The surgery is more invasive, requires advanced skills, and may be ineffective in salvaging hearing, as the ELH continues to damage this aspect in the ear; the nerve section does not halt this aspect. Dysequilibrium may be severe.

iii. Labyrinthectomy.

Removal of the balance elements of the inner ear is effective in eliminating rotatory vertigo, and is done when the hearing is severely damaged, or when the symptoms dictate this course, usually prostration or drop attacks. The surgery removes any remaining hearing and may induce lasting dysequilibrium.

Rehabilitation

**Hearing:** ELH or its management commonly result in severe or profound deafness, with loss of speech discrimination ability, such that conventional hearing aids may be ineffective. Several hearing options are available.

**CROS / BiCROS** aids help to eliminate the “head shadow” dead zone of single sided deafness. These use a receiver on the deaf ear that transmits by Bluetooth to an aid in the better ear (which, in the BiCROS variant also aids the better ear).

**Active bone conduction devices** (Med EL Bonebridge, Cochlear BAHA) act similarly to the CROS aid using skull vibration, and can be implanted at labyrinthectomy surgery. They can be used only when the better ear has excellent hearing, and do not restore full audiological ability (direction finding, stereo, or the improved hearing with two ears – binaural summation). They help greatly at a table, groups, committees etc.

The concern with the bone conduction implants is the possibility of later onset ELH in the contralateral ear, which would negate their effectiveness.

**Cochlear implants** may also be effective. When the hearing is severely damaged, particularly in rapid onset cases or when contralateral disease is present, cochlear implantation of the worse ear is considered, to provide a stable and effective means of communication. The sound quality from a cochlear device is not as clear as the bone conduction implants, but is independent of the other ear and immune to further ELH effects. They help greatly to avoid the socio-economic disruption of bilateral Meniere’s and, particularly in younger patients, are an increasingly employed management.
Balance

Gentamicin perfusion, VIII section and labyrinthectomy carry a significant risk of severe dysequilibrium, as outlined above. This occurs when the brain has not compensated for the drop in function of the affected ear. In younger patients, recovery may be relatively rapid, but in the older case this is more protracted and long-term difficulties may persist. Expert balance rehabilitation physiotherapy is strongly advocated, and persistence with this management is desirable for best outcomes.

When dysequilibrium is severe, however, the intellectual centres are used to help compensate. Distraction of these centres can significantly impede compensation; the patient should avoid a number of such situations:

- **Financial** concern: avoid debt, excessive spending.
- **Friction**: minimise family confrontations.
- **Fatigue**: avoid physical over-exertion (e.g. over-zealous rehabilitation).
- **Movement**: Reduce excessive multidirectional movements.
- **Decision-making**: Constant demands for decisions impact heavily on executives/managers. Other worries, stresses: to be avoided

Benign Positional Vertigo (BPV)

Lastly, ELH cases may be prone to BPV, due to debris resulting from the disease. BPV occurs when particulate debris irritates balance receptors in the ear, causing rotatory vertigo in specific bodily positions or movements, including arising, looking up, rolling over in bed, or sudden horizontal movements. The problem is often remedied by specialized exercises (Epley manoeuvres) that reposition the offending particles away from the sensitive site.

ELH is a complex, damaging and often distressing condition. Fortunately treatment will terminate the vertigo, or this will burn out spontaneously in the long term, if untreated.

More information

- [Benign Positional Vertigo](#)