The Journal of Laryngology and Otology
November 1996, Vol. **110**, pp. 1008–1011

Main Articles

The effect of surgical removal of the extraosseous portion of the endolymphatic sac in patients suffering from Menière's disease

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Abstract

Between April 1990 and June 1996, the extraosseous portion of endolymphatic sac has been removed from the affected ear in 77 patients suffering from Menière's disease. Removal of the extraosseous part of the endolymphatic sac without any drainage procedure did not increase the frequency or severity of the attacks of vertigo in any patient. The results of 43 patients with unilateral disease who had a follow-up period of two years are presented. Only eight of the patients had more than two recurrent attacks of vertigo lasting over two minutes within the two years after the surgery, and in three of these patients the severity of the attacks was greatly reduced. In 56 per cent of the operated ears, the hearing deteriorated at least 10 dBHL across five audiometric frequencies (250 Hz, 500 Hz, 1 kHz, 2 kHz and 3 kHz). To the author, endolymphatic sac removal appeared to provide better relief from vertigo than a simple drainage procedure with less tendency for recurrence several months or years after the initial surgery. At present, the statistical analysis of the results shows no significant difference between removal of the extraosseous portion of the endolymphatic sac and the 'so called' endolymphatic sac drainage procedures.

Key words: Menière's Disease; Vertigo; Endolymphatic sac, surgery

Introduction

Endolymphatic sac (ELS) surgery remains a controversial surgical therapy for Menière's disease. The fickle nature of Menière's disease and the lack of any clear rationale to explain the apparent successes claimed for the surgery fuel the controversy. In man, the ELS usually has an intraosseous portion which lies within the petrous bone and an extraosseous portion which lies on the dura medial to the lateral sinus. The extraosseous portion is difficult to define surgically and, from the surgeon's viewpoint, appears only as a thickened area of dura. The ELS is usually 'opened' by splitting the layers of the ELS, but no endolymph is seen and, usually, no significant electrophysiological changes occur (Gibson, 1993). How does such surgery influence the course of Menière's disease?

The anatomy of the endolymphatic sac

Over the past ten years, electron microscopic studies have done much to explain the anatomy and histology of the ELS. The human ELS shows an enormous variability in size. In contrast to animals such as the guinea pig, only a few human ELS lie entirely on the dura outside the temporal bone. Usually only about half of the human ELS is extraosseous (Friberg *et al.*, 1988).

The ELS is not a simple 'sack-like' structure. There is usually some free lumen but much of the ELS is composed of tubules which lie tranversely to its length (Bagger-Sjöbäck *et al.*, 1990). Histologically, the ELS is composed of ciliated cells. There are also light and dark cells, supporting cells, etc. (Lundquist *et al.*, 1964).

The physiology of the endolymphatic sac

Recent advances in the understanding of the physiology of the ELS have occurred (Wackym *et al.*, 1987). There is clear evidence of longitudinal flow of endolymph towards the ELS but it appears unlikely that the ELS is a passive receptacle of endolymph. Endolymph is propelled towards the ELS by an osmotic gradient which is maintained by the stria vascularis (Sterkers *et al.*, 1984). The final passage of fluid down the endolymphatic duct may

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Accepted for publication: 19 August 1996.

be further regulated by the secretion of hydrophilic proteins within the lumen of the ELS.

There appear to be four main functions of the ELS.

(1) *Longitudinal absorption of endolymph*

How essential the longitudinal flow of endolymph is in man is unknown. In animals, such as the guinea pig, removal of the ELS results in endolymphatic hydrops within a few weeks but in higher mammals such as dog and monkeys, endolymphatic hydrops may take years to develop.

In man, it appears that the water component of endolymph can be regulated by radial absorption of endolymph (Lundquist, 1976). Longitudinal flow of endolymph is needed to carry debris, virus particles, etc. to the ELS. Longitudinal absorption may also be needed if excessive production of endolymph has occurred, which is probably the situation in Menière's disease.

(2) *Removal of debris and virus particles*

The ELS is the only part of the inner ear which has the mechanisms for phagocytosis of debris, lymphocytes, etc.

(3) *Secretion of immunoproteins*

Several different immunoproteins have been identified in the ELS and this has raised speculation that an autoimmune process could underlie Menière's disease (Harris and Tomiyama, 1987).

(4) *Secretion of glycoproteins*

There is now evidence in animals and man that the ELS secretes glycoprotein (Rask-Andersen *et al.*, 1991). Glycoprotein is extremely hydrophilic.

Pathophysiology of the endolymphatic sac

When Hallpike and Cairns (1938) described endolymphatic hydrops, they also noted fibrosis of the ELS. Subsequently, many authors have described abnormalities of the ELS in Menière's (Schuknecht, 1976; Fraysse *et al.*, 1980; inter alia). It appears that as the disease progresses, the sac becomes inactive. Schuknecht (1976) suggests that the duct leading into the proximal part of the ELS becomes blocked, but this is defined by others (Rask-Andersen *et al.*, 1991).

The human ELS can usually be found on the dura on a line drawn through the longitudinal axis of the posterior semicircular canal as seen within the mastoid cavity (Donaldson's line). In Menière's disease, the sac usually assumes a more inferior position and may appear smaller, less vascular and more difficult to open surgically than the normal human ELS (Arenberg *et al.*, 1977).

Proposed mechanisms for the vertiginous attacks of Menière's

There have been several theories to account for the acute attacks of vertigo in Menière's disease. In the ears of animals deprived of their ELS, hydrops develops but the animals do not suffer acute attacks of vertigo. The acute attacks of vertigo seem to be at their worst when the ELS is still active in the early stages of the disease.

(1) *The rupture theory* (Schuknecht, 1968)

It seems unlikely that ruptures of Reissner's membrane can be the cause of the vertigo. Ruptures are unusual in the early stages of the disease when the vertigo is at its worst. Furthermore, ruptures would be expected to cause some loss of haircells within a few millimetres of the rupture site, but no islands of low frequency audiometric loss are detectable.

(2) *The distension theory* (Shea, 1993)

The concept is that a rise in the volume of endolymph flattens and inactivates the vestibular haircells, resulting in hypofunction. The ELS becomes active and removes this excessive endolymph returning the system back to normal.

(3) *The drainage theory* (Gibson and Arenberg, 1991)

The drainage theory suggests that the attacks of vertigo are due to periods of rapid drainage of endolymph towards the endolymphatic sac. It is postulated that there is an abnormal build up of fluid in the inner ear which eventually causes a hyperactive response in the endolymphatic sac. A smaller endolymphatic duct than usual may be a prerequisite as a large or open endolymphatic duct may allow passive drainage of endolymph and prevent a build up occurring. The endolymphatic sac secretes glycoproteins which induces a sudden, rapid longitudinal flow. This hyperactivity of the ELS ultimately results in its self-destruction as the mucosa is damaged by the glycoprotein and eventually the sac becomes fibrosed and functionless.

Patient selection

All the patients chosen had definite Menière's

TABLE I
VERTIGO RESULTS ACCORDING TO AAOO-HNS CRITERIA (1995)

Category	A complete	B substantial	C limited	D insignificant	E worsening
	25	11	5	2	1

https://doi.org/10.1017/S0022215100135637 Published online by Cambridge University Press

TABLE II
THE EFFECT ON THE HEARING OF ELS REMOVAL TWO YEARS AFTER SURGERY

Hearing improvement over 10dB	Hearing unaltered +10dB to −10dB	Hearing worse −11dB to −20dB	Hearing worse −21dB to −30dB	Hearing worse −31 to −40dB	Hearing worse over 40dBHL
2	17	15	7	2	0

disease according to the AAOO-HNS 1995 criteria (Monsell *et al.*, 1995). On a ten point history scale (Gibson, 1991) all scored over seven points. The transtympanic electrocochleogram confirmed the diagnosis in all patients by demonstrating enhanced summating potentials.

All patients had a hearing loss of at least 30 dBHL when averaged at frequencies 250 Hz, 500 Hz, 1 kHz, 2 kHz and 3 kHz (Menière's pure tone average (MPTA)). All patients had attacks of vertigo with the sensation of spinning lasting over 30 minutes. All patients had to have at least six attacks in the preceding six months.

The ELS has been removed in 77 patients. The results are presented for the 43 patients with unilateral disease and a follow-up of at least two years.

Surgical method

The surgical technique has evolved during the course of the study as the author has been encouraged by the results. In 1990, only one patient who was facing a probable labyrinthectomy had the ELS excised, but he did remarkably well. In 1991, two further patients underwent this surgery. In 1992, seven patients, and in 1993 18 patients had surgery to excise the ELS. Initially all the ears were monitored using intraoperative electrocochleography and the sac was excised at the neck similar to the method of Welling and his co-workers (1996). No significant changes on the electrocochleogram occurred. During 1993, the technique was altered so that the duct of the ELS was avulsed. This technique does not appear to alter the electrocochleographic monitoring, probably because the endolymphatic duct is so narrow at the isthmus that no force is transmitted into the vestibule. It is the authors belief that the avulsion of the duct of the sac provides better results than merely cutting out the extraosseous portion of the sac.

Results

The effect of ELS removal on the attacks of vertigo

The crucial reason for undertaking this study was to differentiate between the different hypotheses for the cause of vertigo in Menière's disease. If the rupture or the distension theory were correct it was likely that the patients would be worse after surgery. If the drainage theory of Gibson and Arenberg (1991) was correct, the patients who lost all ELS function would probably lose some hearing but should remain free from vertigo. Patients who had an incomplete removal would have attacks of a lesser severity. The results are shown according to the 1995 AAOO-HNS recommendations in Table I.

Monsell and Weit (1988) reviewed several studies of the effect of endolymphatic sac surgery on the vertigo caused by Menière's disease and showed that most authors report control in approximately 70 per cent of the patients. It would appear therefore that this study provides similar results to those in which an attempt is made to drain the ELS.

The findings do support the drainage theory. None of the patients appeared to have more severe vertigo after the surgery and only six had recurrent attacks within the first year. Complete or substantial freedom from attacks occurred in 83 per cent of the patients.

The effect of ELS removal on the hearing in the operated ear

According to the theory proposed by Gibson and Arenberg (1991), the hearing after disabling the ELS would deteriorate unless the radial flow mechanism is sufficient to prevent the accumulation of endolymhatic hydrops. This may depend on whether or not the mechanism causing the increases secretion of endolymph continues. If the endolymphatic hydrops becomes permanent without the auxillary mechanism for drainage to the ELS, then the hearing should deteriorate to the levels seen in 'burnt out' Menière's disease (Stahle *et al.*, 1991).

Hearing change was measured using the Menière's pure tone average (MPTA) in which the hearing levels in dBHL at 250 Hz, 500 Hz, 1 kHz, 2 kHz and 3 kHz were averaged. There were two individuals in which the hearing improved (24 dB MPTA and 18 dB MPTA) post-operatively and there was less than 10 dB deterioration in 17 ears. The amount of hearing loss depended on how much hearing loss existed prior to the surgery. Several patients already had a hearing loss in excess of 50 dBHL prior to the surgery and, even if this deteriorated to 60 dBHL post-operatively, they still managed to remain in the category of less than ±10 dBHL. Ears that only had a loss of 30 dBHL which deteriorated to the same levels could have a loss of over 30 dB MPTA (Table II).

The effect of ELS removal on tinnitus

Tinnitus is a distressing complaint which appears to relate to the level of anxiety and uncertainty experienced by the patient. Not surprisingly, when the attacks of vertigo ceased and when the patients

TABLE III
THE EFFECT ON TINNITUS OF ELS REMOVAL, TWO YEARS AFTER SURGERY

Gone	Better	Same	Worse
3	13	23	3

https://doi.org/10.1017/S0022215100135637 Published online by Cambridge University Press

TABLE IV
THE EFFECT ON THE SENSATION OF AURAL FULLNESS OF ELS REMOVAL, TWO YEARS AFTER SURGERY

Gone	Better	Same	Worse
7	19	14	3

had faith that their disease was fully explained, the tinnitus levels faded in many patients. In general, those patients who did not receive relief from vertigo did not gain any relief from their tinnitus. The results are shown in Table III.

The effect of ELS removal on the sensation of fullness

It was feared that the sensation of fullness within the operated ear would increase post-operatively because of unrelenting endolymphatic hydrops. In reality, this has not occurred and many of the patients have reported relief from the sensation of fullness. The author cannot offer any explanation. The results are shown in Table IV.

Conclusions

The author does not suggest that removal of the extraosseous ELS is an ideal therapy for Menière's disease. The hearing results are far from satisfactory. The study is important because it helps to clarify the mechanism by which ELS surgery operates.

Removal of the extraosseous portion of the human ELS may only partially damage the function of the ELS as, in the majority of ears, most of the active portion lies within the temporal bone. The more important aspect maybe that the blood supply of the ELS is mostly derived from dural vessels and appears quite fragile. Disruption of these vessels probably does cause some loss of function of the intraosseous portion of the ELS.

The findings of this paper suggest that removal of the ELS has the same (or perhaps better) effect in terminating the attacks of vertigo. These findings suggest that the ELS may have an active role in causing the attacks of vertigo. The author suggests that any form of endolymphatic sac surgery usually causes a temporary or permanent reduction in endolymphatic sac activity. The apparent success of endolymphatic sac surgery may be due to reduction in the secretion of glycoprotein which reduces the movement of endolymph towards the ELS averting the attacks of vertigo.

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