TESTING FOR PERILYMPHATIC FISTULA BASED ON HENNEBERT'S SIGN AND TULLIO PHENOMENON

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A test procedure for perilymphatic fistula (PLF) using both subsonic and sonic stimuli relying on the patients' reported reactions is described. Its reliability to predict the presence of perilymph fistula is assessed by comparing the pre-operative test results, the surgical findings, and the post-operative reported symptoms. Based on a sample of 40 operated ears and a control group of 129 ears, the test's sensitivity to predict PLF was 76% using a sonic stimulus and 83% using a subsonic stimulus. The procedure gave false positive results in ears with Meniere's disease.

Hennebert's sign is the observation of nystagmus elicited by positive and negative pressure in the ear canal. It was described by Hennebert in 1911 after observation of patients with congenital syphilis. Hennebert attributed the reaction to a bony fistula in the labyrinth and considered it to be a "fistula sign" (Hennebert 1911).

In 1929, Tullio reported a similar phenomenon in an experimental study. After creating a hole in a pigeon's semi-circular canal, he observed "neck nystagmus" in reaction to loud sound stimulation produced by a flute (Tullio 1929).

Both Hennebert's sign and the Tullio phenomenon were initially considered to be pathognomonic of congenital fistulae. Later observations, however, showed that their presence were strong indicators of fistulae in the cochlear windows.

There is a similarity between the Hennebert's sign and the Tullio phenomenon, the difference being the stimulus used to elicit the reaction. Both have been known as "fistula tests". The former requires a subsonic stimulus (pressure waves) while the latter requires a sonic stimulus (sound waves). Both stimuli must have large amplitudes in order to activate the fistula and to elicit a reaction.

During the last 20 years, different procedures have been described to record the Hennebert's sign and the Tullio phenomenon as a means of testing for perilymph fistula (PLF). Initially, recording of nystagmus using electrodes was attempted (Hemenway 1968, Singleton et al 1978, Dapsit et al 1980, Causse et al 1983, Fox et al 1988). More recently, recording of body sway with postural platforms has been used to record the reaction to both pressure changes and loud sound stimulation of patients suspected of having PLF (Black et al 1990, Pyykko et al 1991).

In our experience, attempts to record nystagmus in patients with PLF have been frustrating. Most of our patients reported a subjective sensation of dizziness and nausea to sonic and subsonic stimuli but no nystagmus could be observed in the dark by means of an infra-red video system.

Kohut (1992) reported that symptoms of dizziness and nausea, rather than nystagmus, were present in the majority of his patients during "fistula tests". Black et al (1990) considered that the suppression of nystagmus in these patients is often replaced by a vestibulospinal reflex (body sway) as a compensatory mechanism.

We conducted a study to compare the effects of both a subsonic and a sonic stimulus in patients suspected of having PLF. The test relied on the patients' subjective report of dizziness and nausea and on our own observations of the vestibulospinal reflex.

The procedure was totally subjective as, at the time of the study, we did not have any means to record the vestibulospinal reflex. We considered, however, that the test procedure was no less reliable than subjective audiometry, which has been relied upon for more than 60 years for the diagnosis of ear pathologies.

Method and Procedures

Patients undergoing exploratory tympanotomy for PLF were tested pre-operatively and the test results were compared to the surgical findings. Three months later, the patients were surveyed in order to verify resolution and/or improvement of post-surgery symptoms.

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Two different stimuli were used to elicit the reactions. The first was a subsonic stimulus produced by systematically changing the pressure pump to an impedance bridge from +400 to -400 mmH_2O at 1 to 2Hz (Black et al 1990). This stimulus produced the equivalent of 160 dBSPL when delivered into the ear canal through an air tight sealed probe tip.

The sonic stimulus was chosen on the basis of Pyykko's recommendation that the lower the frequency of the sound at a high intensity the more effective the stimulation. We used a pure tone 128 dBSPL at 250 Hz as it was the highest SPL our audiometer would produce at a lower frequency. In order to avoid sound leakage, the sound was delivered into the ear canal using an EAR Tone 3A insert earphone.

The subjects were instructed to concentrate on their balance and asked to report any sensation felt during or immediately after the stimulation. The examiner observed the subject during the procedure and registered any head and/or body sway observed during the stimulus presentations.

A control group of 10 males and 10 females with no past history of ear pathology was tested. None presented any reaction to either subsonic or sonic stimuli. The sonic stimulus produced a startle reflex in 12 of the subjects.

A second control group with different ear pathologies was also tested. No reaction was observed in 40 ears with noise induced hearing loss, 20 ears with presbyacusis, 6 ears with acoustic neuromas and 2 ears with meningiomas. A positive reaction, however, was observed in 35% of our 21 ears diagnosed as having Meniere's disease.

During a period of 30 months, we collected a series of 40 patients who were undergoing exploratory tympanotomy for PLF. The decision for surgery was made by the surgeon's clinical judgment. Eight of these patients had an iatrogenic cause attributed to the PLF (7 post-stapedectomy and 1 post-mastoidectomy), 14 had a history of barotrauma (11 scuba diving, 1 free diving and 2 flying), 5 had a history of head injury, 3 had Meniere's disease, 9 had no defined cause, and 1 patient had acoustic trauma as the suspected cause.

Table 1 shows the pre-operative symptoms in the suspected ear of the 40 patients involved in the study. All patients were tested prior to the surgery with the procedure described above. The non-suspected ear was tested first, followed by the suspected ear. With the exception of one female, no others presented any reaction to the stimulation of the non-suspected ear. This patient, who presented a bilateral positive reaction, was found to have fistulae in both round window membranes due to scuba diving.

The subjective reactions described by our patients were: dizziness, unsteadiness, spinning in the head, moving sideways, moving back and forth, rocking, lightheadedness, and/or nausea. The test was considered positive if any of the above reactions was reported and/or if the examiner observed a vestibulospinal reaction during the stimulation.

In some instances, it was difficult to establish if a reaction to the sonic stimulus was due to the Tullio phenomenon or to a startle reflex. We considered it to be a startle response if the spinal reflex was not accompanied by a subjective vestibular reaction and was present only during the first stimulation, disappearing when the sound was presented for a second time.

Table 1.
Pre-operative symptoms in the suspected ear of all patients in the study.

	AGE	SEX	HEARING LOSS	TINNITUS	BALANCE	NAUSEA
1	35	f	mild flat*	o.ring.	c.float.	nil
2	42	m	mod.HF	c.ring.	c.rock.	nil
3	45	f	mild flat	c.ring.	c.float.	constant
4	39	f	nil	nil	c.off bal.	constant
5	40	m	mild HF	o.pop.	c.lighthead.	occasional
6	29	f	nil	nil	o.vertigo	constant
7	22	f	sev.HF	nil	o.vertigo	occasional
8	17	f	mod.HF	nil	normal	nil
9 #	26	f	mild	c.ring.	c.off bal.	nil
10	64	f	mod LF*	c.buzz.	c.off bal.	constant
11	42	m	mod flat	c.ring.	o.vertigo	occasional
12	72	f	mod/sev HF*	o.ring.	c.off bal.	constant
13	55	m	mod/sev HF*	o.ring.	c.off bal.	nil
14	64	f	mod HF	nil	c.off bal.	nil
15	32	m	mild MF	c.ring.	normal	nil
16	50	f	sev flat	c.buzz.	c.off bal.	constant
17	25	f	nil	nil	c.rock.	nil
18	38	f	mild*	o.ring.	c.off bal.	occasional
19	60	f	mod HF*	c.ring.	o.vertigo	occasional
20	33	f	sev flat*	c.ring.	c.rock.	constant
21	47	f	nil	o.ring.	o.off bal.	occasional
22	48	m	sev HF	o.ring.	c.off bal.	occasional
23	23	f	nil	o.buzz.	c.off bal.	occasional
24	49	m	mild HF	c.ring.	o.off bal.	occasional
25	52	m	mod/sev HF	c.ring.	c.lighthead.	occasional
26	19	f	mild flat*	o.ring.	c.off bal.	constant
27	72	f	mod LF	c.ring.	o.vertigo	occasional
28	48	m	mild flat*	o.ring.	o.off bal.	occasional
29	27	f	mod HF*	c.ring.	o.vertigo	nil
30	29	m	nil	c.ring.	o.off bal.	nil
31	25	f	sev flat*	c.buzz.	o.off bal.	nil
32	32	m	nil	o.buzz.	c.off bal.	constant
33	36	f	mild flat*	c.buzz.	o.vertigo	occasional
34	39	f	mod/sev HF	c.ring.	normal	nil
35	34	f	mod HF	c.ring.	c.float.	occasional
36	31	m	mod HF	c.ring.	o.vertigo	nil
37	29	f	nil	o.ring.	c.float.	constant
38	44	f	mild HF	nil	c.off bal.	nil
39	50	f	nil	o.humm.	o.vertigo	nil
40	27	m	nil	c.buzz.	o.vertigo	nil

c:constant
o:occasional
pop.:popping
buzz.:buzzing
humm.:humming
float.:floating
rock.:rocking
ring.:ringing
bal.:balance
lighthead.:light headed
HF:high frequency
LF:low frequency
mod:moderate
sev:severe
Patient had bilateral PLF
* Conductive component

Results

Thirty-four of the 40 patients undergoing surgery for PLF had a positive reaction to either subsonic and/or sonic stimulation in the pre-operated ear.

Table 2 shows the pre-operative test results for both the subsonic and the sonic stimuli in the suspected ear, the surgical findings and the probable aetiology according to the surgeon's notes.

All the patients had been operated on by either Dr. John Tonkin or Dr. Paul Fagan. Of this group of operated patients, 26 had a PLF identified in surgery. The remaining 14 had both windows grafted in a prophylactic procedure.

A questionnaire 3 months after surgery showed that 29 of the operated patients (including 4 whose PLF was not identified) had their symptoms improved or resolved. Table 3 shows the post-operative symptoms according to patients' questionnaire reply.

Although we realise the possible placebo effects of surgery, for the purposes of this study all patients who reported improvement or resolution of symptoms were

Table 2.

PLF test results for sonic and subsonic stimuli, operated ear, surgical findings and probable aetiology according to surgeon's notes.

	SONIC STIM.	SUBSONIC STIM.	SURGICAL FIND.	AETIOL.
1	posit.	posit.	PLF R. OW	iatrogenic
2	posit.	negat.	PLF R. RW	unknown
3	posit.	posit.	no R. PLF	barotrauma
4	posit.	posit.	PLF R. OW	head injury
5	posit.	posit.	PLF R. RW	unknown
6	posit.	posit.	PLF L. OW	head injury
7	posit.	posit.	PLF L. RW	head injury
8	negat.	posit.	PLF L. RW	barotrauma
9 #	posit.	posit.	PLF R.L.RW	barotrauma
10	negat.	posit.	PLF L. OW	iatrogenic
11	negat.	negat.	no L. PLF	Meniere's
12	negat.	negat.	no R. PLF	iatrogenic
13	negat.	negat.	PLF L. OW	barotrauma
14	posit.	posit.	no L. PLF	unknown
15	posit.	posit.	PLF R. OW	barotrauma
16	posit.	posit.	PLF L. OW	unknown
17	posit.	posit.	PLF R. OW	unknown
18	posit.	posit.	PLF R. OW	iatrogenic
19	negat.	posit.	no L. PLF	Meniere's
20	posit.	posit.	PLF R. OW	iatrogenic
21	posit.	posit.	PLF L. RW	head injury
22	posit.	negat.	no L. PLF	unknown
23	negat.	posit.	no L. PLF	barotrauma
24	posit.	negat.	no R. PLF	unknown
25	posit.	negat.	PLF L. OW	barotrauma
26	posit.	posit.	PLF R. OW	iatrogenic
27	negat.	negat.	no R. PLF	Meniere's
28	negat.	negat.	no R. PLF	barotrauma
29	posit.	posit.	PLF L. OW	barotrauma
30	posit.	posit.	PLF L. OW	barotrauma
31	posit.	negat.	no R. PLF	barotrauma
32	posit.	posit.	PLF R. RW	unknown
33	posit.	negat.	PLF L. OW	iatrogenic
34	posit.	posit.	no L. PLF	acoust.trauma
35	posit.	posit.	PLF L. OW	barotrauma
36	posit.	posit.	PLF R. RW	barotrauma
37	posit.	posit.	no L. PLF	head injury
38	negat.	negat.	no R. PLF	unknown
39	negat.	posit.	PLF L. OW	iatrogenic
40	negat.	posit.	PLF L. RW	barotrauma

R.:right OW:oval window acoust.:acoustic
L.:left RW:round window

Bilateral PLF

Table 3.

Post-operative symptoms according to patient's questionnaire results. The presence of PLF was either confirmed by surgery or by improvement of symptoms as shown in questionnaire.

	HEARING	TINNITUS	BALANCE	NAUSEA	PLF
1	same	same	cured	nil	yes
2	same	same	cured	nil	yes
3	better	same	cured	better	yes
4	same	nil	cured	cured	yes
5	worse due to post-op.		complications		yes
6	same	nil	better	better	yes
7	better	nil	better	better	yes
8	better	nil	nil	nil	yes
9	better	same	cured	cured	yes
10	same	same	better	better	yes
11	same	better	better	better	no (ESD)
12	same	same	same	same	no
13	same	better	cured	nil	yes
14	same	nil	same	nil	no
15	same	same	nil	nil	yes
16	same	better	cured	cured	yes
17	same	nil	better	nil	yes
18	same	better	better	better	yes
19	same	better	better	better	no (ESD)
20	better	better	better	better	yes
21	worse	better	cured	cured	yes
22	same	same	same	same	no
23	same	better	better	cured	yes
24	same	same	same	same	no
25	same	better	better	nil	yes
26	same	better	better	cured	yes
27	same	better	better	cured	no (ESD)
28	better	better	better	better	yes
29	same	better	cured	nil	yes
30	same	cured	better	nil	yes
31	same	same	same	nil	no
32	same	same	same	same	yes
33	same	better	cured	cured	yes
34	same	same	same	same	no
35	same	better	better	better	yes
36	same	same	same	same	yes
37	same	same	better	better	yes
38	same	nil	same	nil	no
39	same	better	cured	nil	yes
40	same	same	same	same	yes

(ESD): patients had endolymphatic sac decompression at the same time as the tympanotomy.

considered to have a PLF. One of the patients had his symptoms worsened due to post-operative complications but, as a PLF was reported in the surgical notes, its presence was considered positive for the purposes of this study.

Three of the operated patients where the presence of PLF was considered negative had endolymphatic sac decompression (ESD) as well as a graft of the cochlear windows during surgery. The improvement of their symptoms was attributed to the ESD as they all had Meniere's disease. Although it has been assumed that they did not have PLF for the purpose of this study, a question mark still remains.

There was, therefore, a total of 30 patients who were considered to have a PLF and 10 patients who did not have a PLF confirmed by surgical observation or by improvement of their post-operative symptoms.

Of the 30 patients who had PLF, 23 presented a positive reaction to the sonic stimulus and 25 presented a positive reaction to the subsonic stimulus. There was, therefore, a 76.6% hit rate with the sonic stimulus and 83.3% with the subsonic stimulus.

The false negative rate for the sonic stimulus was 23.3% and 16.6% for the subsonic stimulus, as 7 patients who had PLF did not react to the sonic stimulus and 5 did not react to the subsonic stimulus. There was also a false positive rate of 12.5% for the sonic stimulus and 7.5% for the subsonic stimulus in this group of patients.

Statistical analysis, using the Chi-square test, showed no significant difference between the expected and observed values at the 5% level. This means that a positive reaction to subsonic or sonic stimuli is a significant predictor of the presence of PLF.

When combining the results of sonic and subsonic stimulation for each patient, the hit rate for the procedure rises to 93%. This is due to the fact that 28 of the 30 patients with PLF presented a positive reaction to either sonic or subsonic stimuli.

Discussion

Although this study used a small group of patients, our hit rate combining the results for sonic and subsonic stimuli (93%) is very close to the best PLF predictor procedures reported in the literature. Black et al (1990) reported a 97% hit rate using a postural platform and a subsonic stimulus, and Gibson (1992) obtained a hit rate of 82% using EcochG to measure the changes in the AP before and after raising the intra-thoracic pressure.

The rate of false positive results in our study was fairly high, indicating that a positive reaction to the sonic and/or subsonic stimuli is not pathognomonic of PLF.

The sonic stimulus produced a higher rate of false negative and false positive results. Due to this and the fact that it is not always easy to distinguish a true Tullio

phenomenon from a startle reflex, we decided to abandon the sonic stimulus.

The subsonic stimulus also produced positive results in patients diagnosed as having Meniere's disease. These results raise two questions: 1) Is PLF a consequence of Meniere's disease? 2) Is endolymphatic Hydrops a consequence of PLF? As not all of these patients have been surgically explored for PLF its presence could not be verified.

Another explanation for the reaction observed in patients with Meniere's disease relies on the presence of vestibular fibrosis which occurs in the late stages of the disease. According to Nadol (1977), a fibrous attachment between the footplate and the saccule will cause instability due to excessive movement of the stapes during pressure changes in the ear canal. Schuknecht (1993) explains the reaction in a similar way, but believes that the fibrous attachment is between the stapes and the utricle.

The fact that our test procedure relies mostly on the patients subjective report has attracted some criticism. More recently, our clinic acquired a "Balance Master" force platform. This equipment provides objective measurements of the patients body sway. At this stage, however, we have not found an improvement in our negative rate using this platform. If the patient does not report a reaction during the stimulation while sitting down, no reaction is observed on the platform. However, the possibility of recording body sway is particularly useful for those patients who have difficulty in objectively reporting their reaction to the stimulation. It would seem reasonable to suggest that a larger number of patients ought to be tested on the force platform, and it is our expectation that the platform (with subsonic stimulus) will provide a documented and measurable result. This will be reported on in a future study.

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The ear, though the most important of all the senses, has hitherto claimed but little attention from the Profession. The diseases of every other organ are well understood, together with the modes of repairing their defects; but the imperfections of this sense having been little attended to by the regular Profession, the treatment has been, for the most part, confined to the hands of empirics: hence, obscurity and prejudice have prevailed in this branch of practice, and an apathy has taken place on the subject, highly injurious to the interests of society.

JOHN HARRISON CURTIS
London, 1817