Noninvasive Assessment of the Intralabyrinthine Pressure

A New Technique Applied to Patients With X-linked Progressive Mixed Deafness Syndrome With Perilymphatic Gusher During Stapes Surgery

Arne Ernst, MD; Ad F. M. Snik, PhD; Immanuel A. M. Mylanus, MD; Cor W. R. J. Cremers, MD, PhD

In this report, we describe the findings of a noninvasive assessment of the intralabyrinthine pressure in two patients from a family with X-linked progressive, mixed deafness syndrome in whom a perilymphatic gusher occurs during stapes surgery. The so-called tympanic membrane displacement measurement technique could be successfully applied in these two patients (from a total of five patients who were studied) because they still showed a stapedial reflex at 1 kHz, which is mandatory for application of the tympanic membrane displacement measurement technique. The findings were compared with those of age-related control subjects and indicated a significantly elevated intralabyrinthine pressure in the two patients who were observed. The results suggest that the tympanic membrane displacement measurement technique may serve as a screening test in audiological diagnostic studies of perilymphatic hypertension.


A mixed hearing loss in patients with an intact tympanic membrane has to be differentiated by audiological testing. One possible reason includes the X-linked progressive mixed deafness syndrome with the occurrence of a perilymphatic gusher during surgery. In such patients, stapes surgery can result in a severe hearing loss and even deafness in some patients. It is therefore important to detect preoperatively a potential stapes gusher in patients. Surprisingly, in patients who do not undergo operations and who suffer from the X-linked progressive mixed deafness syndrome with a stapes gusher, the stapedial reflex can frequently be elicited when the degree of hearing loss does not yet exceed certain limits. This enables the evaluation of the intralabyrinthine pressure by a noninvasive measurement technique, ie, the tympanic membrane displacement (TMD) measurement technique in patients with an X-linked progressive mixed deafness syndrome. The technique was developed in Great Britain and has been introduced in neurological and neuro-otological diagnostic studies recently. It is based on the finding that differences of the intralabyrinthine pressures lead to changed resting positions of the stapes footplate. On acoustical stimulation of the stapedial reflex, a changed reflex pattern in terms of TMD (± air volume displacement in the tightly occluded external ear canal) results. These changes are thoroughly controlled by a servoelectronically operated membrane within the headset to monitor these minute volume displacements (in nanoliters).

PATIENTS AND METHODS

Of a total of more than 30 patients (who all belonged to a family of patients with the genetically characterized X-linked progressive mixed deafness syndrome with a gusher during stapes surgery), five study participants still showed a stapedial reflex. Of this group, patients in whom the stapedial reflex could not be more greatly elicited and those in whom the hearing threshold was largely deteriorated usually underwent an operation for their stapes fixation; in these patients the well-known, intraoperative phenomenon of a stapes gusher was demonstrated.
For at least two of all of our patients who were studied, measurements could be successfully obtained, ie, the stapedial reflex could still be elicited at or below a 110-dB hearing level (HL) at 1 kHz. The remaining three patients only had a stapedial reflex at a 110-dB HL and could therefore not be included in the study.

Audiometry was performed in one (a 31-year-old man [patient 1]) of these two patients and showed symmetrical mixed hearing loss, with an average loss at 0.5, 1, and 2 kHz (the so-called pure-tone average) of an 80-dB HL. The mean air-bone gap at the same frequencies amounted to 27 dB at the right ear and 30 dB at the left ear. The tympanograms of both ears showed no abnormalities. In both ears, the contralateral stapedial reflex could be evoked at most test frequencies at a 100-dB HL.

The other patient was a 30-year-old woman (patient 2) who was a female carrier. As described by Marchbanks and Reid,9 female carriers also show some of the same characteristics as those of male carriers (these characteristics are also less severe). An asymmetrical hearing loss, predominantly of the conductive type, was found. The pure-tone average of the right ear was at a 48-dB HL with a mean air-bone gap of 15 dB; the pure-tone average of the left ear was at a 48-dB HL with a mean air-bone gap of 40 dB HL. Asymmetrical hearing loss in female carriers has been described before.9

After microscopic examination, the patients were studied with pure-tone audiometry and tympanometry; the stapedial reflex threshold was determined by using an acoustical impedance meter (Amplaid 720, Madsen Co, Copenhagen, Denmark) and ipsilateral as well as contralateral stimulation (pure tones of 0.5, 1, 2, and 4 kHz with a maximum stimulus at 120-dB HL). Twenty measurements were averaged. The patients were observed in sitting and supine positions because posture can decisively influence the TMD measurements that result from intracranial pressure changes.6,9 Moreover, changing the position allows one to draw conclusions on the patency of the cochlear aqueduct that connects the intracranial and cochlear spaces.9,11,12 The mean (Vm) and the maximum (Vi) inward displacement of the tympanic membrane (in nanoliters of displacement) were compared with the Vm and Vi of the age-related control subjects who had normal hearing and a normal stapedial reflex threshold at 1 kHz. The TMD measurement technique is based on the finding that a different intralabyrinthine pressure results in varying resting positions of the stapes footplate.9,11,12 As follows: (1) A low intralabyrinthine pressure leads to a resting position more within the oval niche; (2) an increased pressure shifts the stapes footplate more out of the oval niche.13

After stimulation of the stapedial muscle, the stapes footplate can move (with respect to the oval niche) more outward (point 1 above) or inward (point 2 above): this movement can be quantified in terms of air volume displacement (in nanoliters) within the tightly occluded external ear canal because the movement of the stapes footplate is transduced through the ossicular chain to the tympanic membrane. In some patients, a bidirectional movement that consists of an outward and inward movement is observed (point 2 above); this movement can be used to draw conclusions to the underlying relative intralabyrinthine and intracranial pressures, respectively. The Vm is the more reproducible parameter and, thus, the reliable parameter in this respect.8 The measurements are unspecifically influenced by, eg, the cardiovascular activity, head and body movements, and swallowing.2 The intracranial pressure (cerebrospinal fluid) is partially transferred to the cochlea in the case of a patent cochlear aqueduct that largely depends on age12 and has an impact on the measurements too. The patency of the cochlear aqueduct can be tested by changing the posture of the patients.9

In the present study, a statistical analysis was not performed because of the small number of patients (n=2) who showed a stapedial reflex threshold that was useful for investigation. However, the findings of the patients were compared with those of age-related control subjects (n=10).

RESULTS

Of five patients, only two who were both members of the same family and who suffered from the X-linked, progressive, hereditary mixed deafness syndrome (stapes gusher) could be studied because all of the other patients from this family did not have a stapedial reflex at 1 kHz and tolerable sound pressure level intensities (ie, below a 110-dB HL). It was proved that both patients had a significantly increased intralabyrinthine pressure compared with that of the age-related control subjects (Table). When their Vm and Vi values were compared with an average that was found in 10 age-related control subjects, the patients displayed a highly significant increase of the intralabyrinthine pressure as evidenced by the large negative Vm of the tympanic membrane on acoustical stimulation (Table) that corresponded to an inward displacement of the tympanic membrane (and the stapes footplate, respectively). At 10 dB above the stapedial reflex threshold (110-dB HL), the Vm of the right ear was −463 nL in patient 1 in a sitting position compared with +112 nL in the control subjects. No significant interaural differences were found in the results (Table). Another surprising finding in this patient was the fact that the physiological increase in the intracranial and intralabyrinthine pressures, after changing posture from a sitting to a supine position, did not occur (Figure). The Vm value (acoustical stimulation at 10 dB above the stapedial reflex threshold, right ear) changed from −463 nL (sitting position) to +40 nL. In the control subjects, the normal increase in the intralabyrinthine and
Comparison of the TMD Measurements in Two Patients With Those of Age-Related Control Subjects of Normal Hearing

<table>
<thead>
<tr>
<th>Stimulation</th>
<th>Sitting</th>
<th>Supine</th>
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<tr>
<td>Patient 1</td>
<td></td>
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<tr>
<td>SRT</td>
<td>R</td>
<td>-268 (±13)</td>
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<tr>
<td>SRT</td>
<td>L</td>
<td>-140 (±12)</td>
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<tr>
<td>SRT+10 dB</td>
<td>R</td>
<td>-463 (±19)</td>
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<tr>
<td>SRT+10 dB</td>
<td>L</td>
<td>-395 (±15)</td>
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<td>Control subjects (n=10)</td>
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<tr>
<td>SRT</td>
<td>R</td>
<td>+75 (±12)</td>
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<tr>
<td>SRT</td>
<td>L</td>
<td>-89 (±6)</td>
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<tr>
<td>SRT+10 dB</td>
<td>R</td>
<td>+112 (±13)</td>
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<td>SRT+10 dB</td>
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<td>-25 (±8)</td>
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<td>Patient 2</td>
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<tr>
<td>SRT</td>
<td>R</td>
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<td>SRT</td>
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<td>SRT+10 dB</td>
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<td>-866 (±26)</td>
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<tr>
<td>SRT+10 dB</td>
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<td>-1098 (±37)</td>
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* TMD indicates tympanic membrane displacement; Vm, mean displacement of the tympanic membrane; Vi, maximum inward displacement of the tympanic membrane; and SRT, stapedial reflex threshold at 1 kHz of the investigated ear in decibels of hearing level (dB HL). Patient 1, SRT of both ears at a 10 dB HL; patient 2, SRT at a 10 dB HL (right ear) and at a 110 dB HL (left ear). The stimulus was a tone burst at 1 kHz, with a rise/fall time of 500/60 milliseconds, delivered ipsilaterally at the SRT or SRT+10 dB HL. Data are given in nanoliters of displacement (±SD).

Intracranial pressures was found, i.e., the Vm value of the right ear changed from +112 nL (sitting position) to −98 nL (supine position).

In patient 1, an inverse reaction seemed to take place, i.e., the values correspond to a decline in the intralabyrinthine and intracranial pressures. The course and shape of the connections between the cochlea and the intracranial space—primarily the cochlear aqueduct that usually transfers the intracranial pressure change to the cochlea—are uniquely different in these patients. This anatomical situation might be responsible for the finding of an unexpected intralabyrinthine pressure decrease on posture change (from a sitting to a supine position).

Patient 2 only showed a moderate increase in pressure after being measured in the supine position (Table). The Vm values were −305 nL (right ear at a 100-dB HL) and −290 nL (left ear at a 110-dB HL). The recordings in the sitting position were insignificantly different from these parameters; they differed by about 25% compared with a 100% or even greater change in the control subjects (Table). The findings of both patients are contrasted by the expectedly lower intralabyrinthine pressure level in control subjects (sitting position) and the physiological increase in pressure on posture change (from a sitting to a supine position) in the control subjects (Figure); these findings were clearly evident under consideration of the generally smaller initial values. Therefore, the changes that were found from a sitting to a
supine position in control subjects were much more pronounced.

COMMENT

In general, the TMD measurements that were used to assess relative, intralabyrinthine pressure changes in the present study suggested a significant intralabyrinthine pressure elevation in the two patients who were observed. The air-bone gap and the stapedial reflex that were found in these patients are a matter of controversy: we hypothesized that they resulted from the anatomical malformations of the cochlea, the cochlear aqueduct, and the surrounding structures in these patients. Therefore, the audiovestibular system of these patients could have been more efficient in transducing skull vibrations into cochlear fluid motion, ie, the bone-conducting thresholds were better than expected, leading to the observed air-bone gap. This hypothesis has been confirmed by the findings of brain-stem audiometry in these patients. The stapedial reflexes that can be elicited in the initial phase of the disease disappear when the hearing loss continues.

The TMD measurement technique should become an interesting tool of audiological diagnostic studies to differentiate intralabyrinthine hypertension or hypotension further (eg, in a perilymphatic fistula or in the case of a clinically silent hydrocephalus that is accompanied by a hearing loss). In conventional otosclerosis, the test cannot be applied because of the lacking stapedial reflexes.

Our findings support the clinical finding that a stapes gusher is based on an intralabyrinthine overpressure that is released on opening the oval window at the footplate. The TMD measurement technique seems to be well suited to screen patients whose signs and symptoms are suggestive of a stapes gusher and perilymphatic hypertension. However, the large interindividual variations of the TMD measurement technique enable a relative assessment only so that the volume displacements that are measured (ie, Vm and Vi) can indicate an elevated intralabyrinthine pressure, but it cannot absolutely quantify. The TMD measurement technique that is based on previous experiments of Brask, and Densert et al seems to be sensitive enough for this audiological application.

However, the fact that the stapedial reflex can still be elicited in some of these patients should be reason enough not to have these patients undergo operations.

The TMD measurement technique might help to prevent complications in stapes gusher by preoperatively evaluating the intracranial and intralabyrinthine pressures, respectively. Our results confirm that the application of this noninvasive technique is helpful in screening patients for an increase in the intralabyrinthine and intracranial pressures.

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REFERENCES