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Tympanometric Findings in Patients With Enlarged Vestibular Aqueducts

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Objectives: **The purpose of this study was to study systematically some relationships between the resonance frequency of the middle-ear transmission system and the volume of the endolymphatic duct and sac in patients with an enlarged vestibular aqueduct (EVA).** ***Study Design:*** **Prospective study.** ***Methods:*** **Thirteen patients (24 ears) with EVA, 17 subjects (29 ears) with normal hearing, and 17 patients (21 ears) with sensorineural hearing loss without EVA served as experimental subjects. Standard pure-tone audiometry, standard clinical tympanometry (using a 226-Hz probe tone), and multifrequency tympanometry were performed on each ear. Magnetic resonance imaging was used to determine the area of the cochlear modiolus and the volume of the endolymphatic duct and sac.** ***Results:*** **The audiometric configurations for most patients sloped downward from the low to the high frequencies. A significant air–bone gap was computed at each of these test frequencies. Multifrequency tympanometry yielded resonance frequencies for the patients with EVA that was significantly lower than those measured for the control subjects. In general, for patients with EVA, the resonance frequency of the middle ear system decreased as the volume of the endolymphatic duct and sac increased. This inverse relation was significant (correlation coefficient = −0.483, *P* = .0157). However, there was no correlation between resonance frequency and the degree of cochlea modiolar deficiency.** ***Conclusions:*** **Clinically, our findings suggest that EVA probably should be included in the differential diagnosis for a patient who presents with a moderate to severe mixed hearing loss, a normal tympanogram at 226 Hz, and a resonance frequency that is abnormally low.** ***Key Words:*** **Enlarged vestibular aqueduct, resonance frequency, multifrequency tympanometry.**

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INTRODUCTION

The vestibular aqueduct (VA) is a bony canal within the otic capsule. It extends from the medial wall of the vestibule to the posterior surface of the petrous pyramids. The endolymphatic duct traverses this canal. In 1978, Valvassori and Clemis[1] published the first radiographic study that focused on pathologic enlargement of the VA in humans. They identified 50 enlarged vestibular aqueducts in a group of 3700 consecutive patients who had been referred for polytomography of the inner ear. Although complete otologic and audiologic data were not available for all 50 patients, Valvassori and Clemis concluded that an enlarged VA (EVA) is usually "associated with a congenital or a possibly early acquired HL that may be purely sensorineural, or may be mixed in nature." Subsequent research has supported the work of Valvassori and Clemis and the relationship between EVA and hearing loss that typically is progressive and bilateral.[2–6] The air-conduction audiometric configuration usually slopes downward from the low frequencies to the high frequencies. A conductive component to the hearing loss has been reported in most cases of EVA.[7]

Tympanometry obtained with a single low-frequency probe tone provides useful diagnostic information for patients with disorders of the tympanum, for patients with disorders that affect the tympanic membrane, and for patients with eustachian tube dysfunction.[8] Govaerts et al.[6] used standard tympanometry (with a probe-tone frequency of 226 Hz) to investigate the cause of the conductive component in 6 patients with EVA. A normal (type A) pattern was generated for all patients. Multifrequency tympanometry is a more sensitive method for evaluating lesions that affect the ossicular chain and for detecting subtle anomalies of the peripheral auditory system.[8–13] This technique allows one to measure the resonance frequency (f_0) of the entire middle ear transmission system. Resonance occurs at the frequency where the stiffness and the mass elements of the ear are in balance. If the stiffness element of the ear increases, the resonance frequency is

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higher than normal. Conversely, if the mass element of the ear increases (or if the stiffness decreases), the resonance frequency is lower than normal.[14] We have reported that f_0 in patients with EVA is lower than in normal individuals. However, there is no pathologic evidence in the middle ear of patients with EVA.[15] The complex acoustic immittance at the lateral surface of the tympanic membrane and the resonance frequency of the middle ear are determined by the mass and the stiffness of the middle ear transmission system, by the volume and the pressure of air in the tympanic cavities, by the tonus of the middle ear muscles, and by the mechanical immittance of the cochlea. If enlargement of the VA produces an enlargement of the endolymphatic duct and sac and if this, in turn, increases the volume of endolymphatic fluids and reduces its impedance, then patients with EVA should have a resonance frequency that is lower than normal.

The purpose of this study was to examine some relationships between resonance frequency and the volume of the endolymphatic duct and sac in patients with EVA and between the resonance frequency and deficiency in the cochlea modiolar.

MATERIALS AND METHODS

Thirteen patients (24 ears) with EVA (5 males and 8 females; age range = 6–36 y; mean age = 20 y) were included in this study. These patients were referred to our tertiary care center by other physicians. Twelve patients had bilateral enlargement of the VA and 1 patient had unilateral enlargement. One ear was excluded from this study because of a perforation of the tympanic membrane. The control group consisted of two groups: A and B. Group A included studies n 29 ears with normal hearing for pure tones (17 subjects; 11 males and 6 females; age range = 6–34 y; mean age = 23 y). Group B included studies n 21 ears with sensorineural hearing loss without enlargement of the VA (17 subjects; 10 males and 7 females; age range = 8–30 y; mean age = 15 y). The mean and standard deviation (SD) values for the average hearing levels at three frequencies (500 Hz, 1 kHz, and 2 kHz) in group B were 71.0 ± 32.2 dB. The hearing level was matched to that of the patients with LVA syndrome (84.7 ± 21.6 dB). The control subjects of both groups were also approximately matched to the patients with EVA with respect to age. Otoscopic examination for all ears proved normal.

Standard tympanometry (using a 226-Hz probe tone) and multifrequency tympanometry were performed on each ear using a middle ear analyzer (Grason-Stadler, Model GSI 33, Version 2). Static acoustic admittance (at +200 da Pa), tympanometric peak pressure at 226 Hz, and resonance frequency were measured. The same audiometer (Rion, Model AA-61BN) was used in the same sound-insulated chamber for all pure-tone audiometry on all subjects. Air–bone gaps were computed for each test frequency (250 Hz, 500 Hz, 1 kHz, 2 kHz, 4 kHz, and 8 kHz).

Magnetic resonance imaging (MRI; heavily T2-weighted three-dimensional fast-spin-echo MRI) was used to determine the area of the cochlear modiolus and the volume of the endolymphatic duct and sac for both ears of each experimental patient. The VA itself is not visualized on MR images. Its contents, however, are clear.[16] Figure 1 depicts a transverse image of the right petrous temporal bone in a 29-year-old male subject. The cochlear modiolus is identified with an arrowhead. The distinct low-signal-intensity area of the cochlear modiolus was outlined where it was visualized at the maximum size, and the area was measured on the console.[16] The enlarged endolymphatic duct and sac is identified with an arrow. The sum of the areas of the endolymphatic duct and sac was multiplied by the thickness of the section (0.8 mm) to compute the total volume. This method has been described in detail previously.[17] An endolymphatic duct and sac is considered enlarged when the diameter at the midpoint between the common crus and the external aperture is greater than 1.5 mm on thin-section MR images.[16]

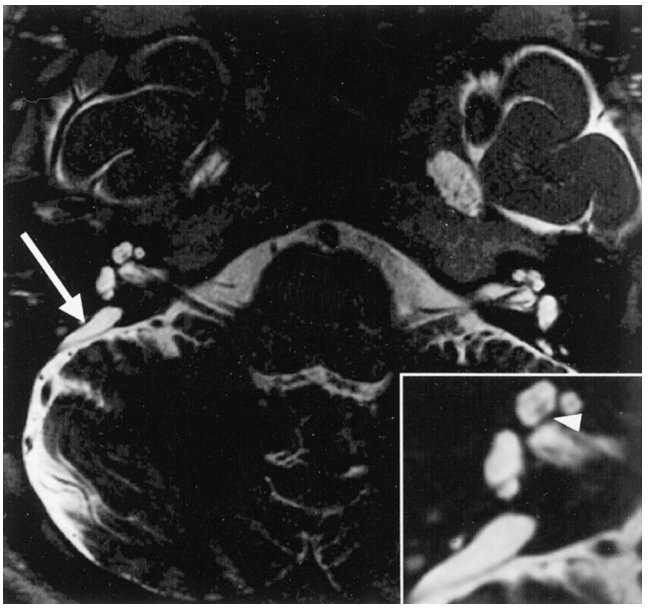

Fig. 1. Heavily T2-weighted transverse magnetic resonance image in a 29-year-old man with an enlarged endolymphatic duct and sac (arrow). Inset shows cochlear modiolus of the right ear (arrowhead).

RESULTS

Figure 2 depicts the mean air-conduction and bone-conduction thresholds for the 13 patients (24 ears) with EVA. Vertical lines through each mean datum point define ± 1 SD. Table I summarizes the air–bone gap data at three frequencies for the patients with EVA. If a patient did not respond to the maximum output of the bone vibrator at a given frequency, their data were excluded for this frequency. The audiometric configurations for most patients sloped downward from the low frequencies to the high frequencies. Consequently, hearing by bone conduction in the middle and in the high frequencies often could not be measured. The mean air–bone gap at 250 Hz was greater than at 500 Hz and 1000 Hz. These differences were significant (Fisher's Protected Least Significant Difference test, $P < .01$ at 500 Hz and $P < .001$ at 1000 Hz).

Standard tympanometry (using a 226-Hz probe tone) produced a normal (type A) pattern for all patients with EVA and for all control subjects. In contrast, multifrequency tympanometry produced results for the patients with EVA that differed from the results produced by both control groups.

The distributions of resonance frequencies (f_0) for the ears with EVA, for the ears in group A, and for the ears in group B are shown in Figure 3. Resonance frequencies for the patients with EVA ranged from 410 Hz to 1200 Hz, whereas those for groups A and B ranged from 670 Hz to 1340 Hz and from 410 Hz to 1410 Hz, respectively. The mean and SD values for f_0 were 777.3 ± 230.5 Hz in the

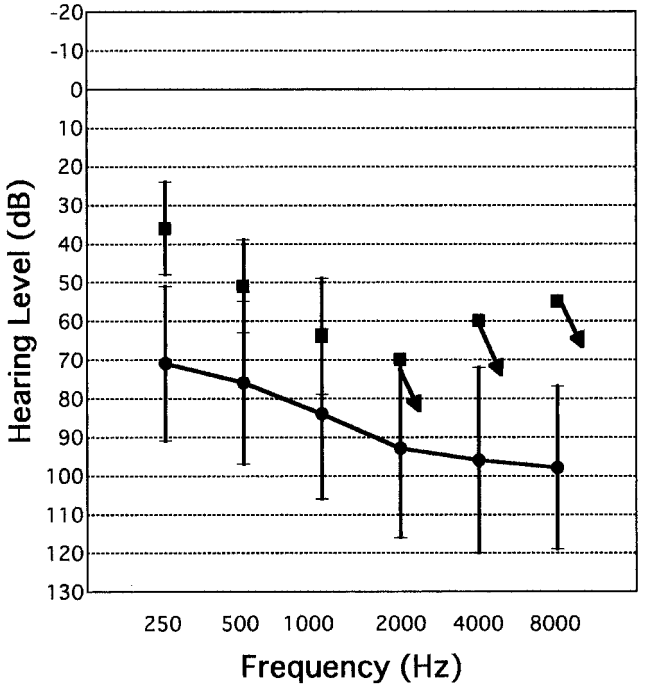

Fig. 2. Mean air-conduction (●) and bone-conduction (■) thresholds for 13 patients (24 ears) with EVA. Vertical lines through each mean datum point define ± 1 standard deviation. The arrow indicates that the average hearing level is beyond the maximum output at a given frequency.

ears with EVA, 956.2 ± 200.2 Hz for the ears in group A, and 944 ± 247.7 Hz for the ears in group B. The value of f_0 in the patients with EVA was significantly lower than both in group A and in group B (Mann-Whitney *U* test, $P = .0064$ for group A and $P = .0203$ for group B). Values of less than 800 Hz for f_0 in the ears with EVA were found in 14 of 24 ears, whereas values of more than 800 Hz in the control groups were found in 39 of 50 ears. The sensitivity and specificity of f_0 for the diagnosis of EVA were 58.3% and 78%, respectively.

Static acoustic admittance (Y_{tm}) was measured and expressed as an equivalent volume of air at 226 Hz. The mean and SD values for Y_{tm} were 0.56 ± 0.25 mL for the ears with LVA syndrome, 0.58 ± 0.27 mL for the ears in group A, and 0.63 ± 0.30 mL for the ears in group B. The difference between these mean values is not significant (Mann-Whitney *U* test, $P = .9359$ for group A and $P = .4193$ for group B).

The mean and SD values for air pressure at the peak of the tympanogram were −2.5 ± 11.7 da Pa in the ears

TABLE I.
Means, Standard Deviations, and Ranges for Air–Bone Gaps at 250Hz, 500 Hz, and 1 kHz.

	0250 Hz (dB) (n = 22)	500 Hz (dB) (n = 22)	1 kHz (dB) (n = 20)
Mean	34.3	25	19.5
Standard deviation	10.4	11.2	10.1
Range	5–45	0–50	0–35

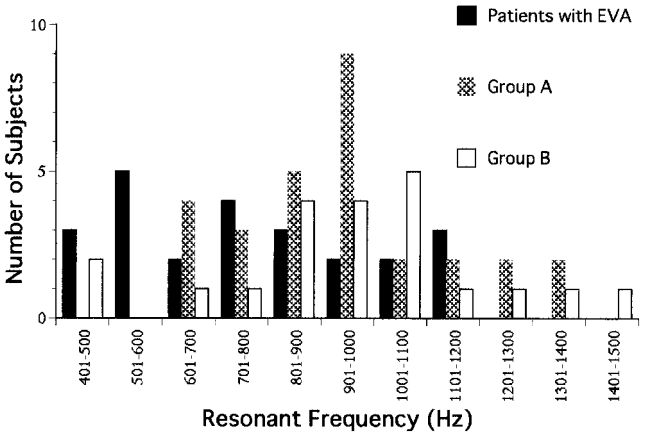

Fig. 3. Distribution of resonance frequencies for the ears with EVA, for the ears in group A, and for the ears in group B.

with EVA, 1.03 ± 4.89 da Pa in the ears of group A, and −2.4 ± 14.1 da Pa in the ears of group B, respectively. The difference between these mean values is not significant (Mann-Whitney *U* test, $P = .5259$ for group A and $P = .7587$ for group B).

The volume of the endolymphatic duct and sac for the patients with EVA ranged from 55.5 μL to 709 μL. The mean value was 368.2 μL with a SD of 204.4 μL. The correlation between the resonance frequency and the volume of the endolymphatic duct and sac is shown in Figure 4. In general, for these patients, the resonance frequency (f_0) of the middle ear system decreased as the volume of the endolymphatic duct and sac increased. This inverse relation was significant (correlation coefficient = -0.483, $P = .0157$). The area of cochlea modiolus for the patients with EVA ranged from 0.96 mm^2 to 5.86 mm^2 (mean ± SD, 2.37 mm^2 ± 1.28 mm^2). The correlation coefficient between the area of cochlea modiolus and resonant frequency was 0.117 ($P = .5892$).

Table II provides a summary of correlation coefficients for the patients with EVA. More specifically, the correlation coefficient between air–bone gap data and f_0

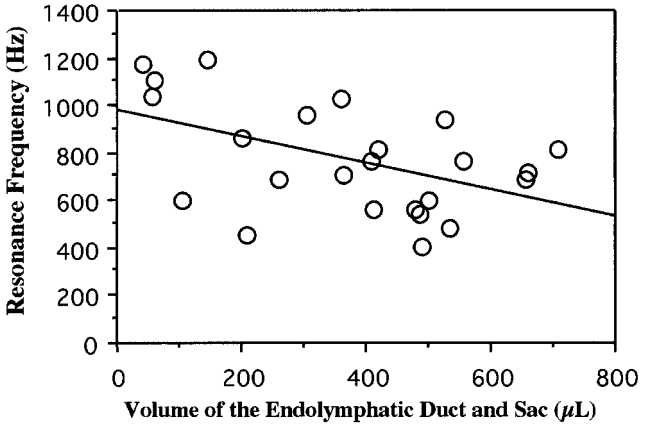

Fig. 4. Graphic representation of correlation between the resonance frequency and the volume of the endolymphatic duct and sac (r = −0.483). The line represents the best-fit regression line.

TABLE II.
Correlation Coefficients and Probability (p) Values Between Air–Bone Gap and Resonance Frequency Values, Between Air–Bone Gap and Cochlea Modiolar Deficiency, and Between Air–Bone Gap and Volume of the Endolymphatic Duct and Sac at 250 Hz, 500 Hz and 1 kHz.

	250 Hz	500 Hz	1 kHz
Resonance frequency	−0.063 (P = .7817)	−0.186 (P = .4115)	−0.016 (P = .9515)
Area of the cochlea modiolus	−0.388 (P = .0745)	−0.265 (P = .2374)	−0.220 (P = .9331)
Volume of the endolymphatic duct and sac	−0.410 (P = .0576)	−0.087 (P = .7039)	−0.122 (P = .6472)

values, between air–bone gap data and the area of cochlea modiolus, and between air–bone gap data and the volume of the endolymphatic duct and sac are tabulated. Probability (P) values also are listed. Superficially, it appeared that the air–bone gap at 250 Hz was larger for the patients with an increased volume of the endolymphatic duct and sac but no significant correlation was found between the air–bone gap and the volume of the endolymphatic duct and sac at any frequency. Furthermore, no correlation was found between the air–bone gap and frequency of resonance and between the air–bone gap and the area of cochlear modiolus.

DISCUSSION

The exact pathophysiology of the hearing loss that accompanies EVA is unknown. It has been reported that the size of the vestibular aqueduct or the size of the endolymphatic duct and sac is not associated with the degree of hearing loss in EVA.[17] A recent MRI and computed tomography study revealed that LVA syndrome is often associated with cochlea modiolar deficiency.[16,18] No correlation was found, however, between cochlea deficiency and hearing level.[16]

Regarding the air–bone gap observed in most cases of EVA, the results from the present study suggest that there is no significant relationship between the magnitude of the air–bone gap and the volume of the endolymphatic duct and sac. To date, no reports have described middle ear or ossicular chain anomalies during operations for inspection of the middle ear or during cochlea implant surgery on patients with EVA. In some cases, however, a profuse leakage of perilymphatic fluid has been observed.[6,15] Intracochlear micromechanical alterations may be considered as the cause of the air–bone gap in EVA. However, there was no correlation between air–bone gap and the degree of cochlea modiolar deficiency in the present study.

We have reported that the resonance frequency (f_0) in the patients with EVA is low compared with the f_0 measured for subjects with normal hearing.[7] In the present study, we have shown that the resonance frequency (in hertz) and the volume of the endolymphatic duct and sac (in microliters) are related inversely.

At least two mechanisms may be responsible for this finding. The first mechanism involves the volume of endolymph within the human inner ear. The total volume of endolymph within the normal inner ear is approximately 34 μL, but there is no measurable amount of endolymph in the normal endolymphatic sac.[19] In the present study, the volume of the endolymphatic duct and sac in the patients with EVA varied from 55.5 μL to 709 μL. Thus, we can assume that the volume of the endolymph was significantly larger in patients with EVA than in those with normal ears. Because the mechanical impedance at the footplate of the stapes is inversely proportional to the volume of fluid within the inner ear, an increase of the volume of inner ear fluid may be responsible for the reduction in f_0 that was measured during multifrequency tympanometry.

Evidence for a second mechanism comes from research in the area of hearing by bone conduction. Tonndorf and Tabor[20] investigated the compressional component of bone conduction that was first described by Herzog and Krainz.[21] Their findings supported the notion that the oval window and the round window are not the only release points for pressure within the cochlea. They used a term proposed by Ranke et al.,[22] "the third window," to refer collectively to the vestibular aqueduct, to the cochlear aqueduct, and to other vascular and neural channels around the cochlea. Enlargement of the vestibular aqueduct increases the size of this third ("window") pressure-release point of the cochlea, decreases the mechanical impedance of the inner ear, and thus reduces the resonance frequency of the entire system.

Theories for hearing by bone conduction also may help explain why we observed a significant air–bone gap for our patients with EVA. At least four mechanisms are responsible for hearing by bone conduction. In normal ears, the magnitude and the phase of each mechanism interact with frequency to yield "normal" bone-conduction hearing and the bases for audiometric standards. If, however, the volume of the endolymphatic duct and sac is abnormally large, then the relative contributions of: 1) the ossicular–inertial mechanism[23,24]; 2) the cochlear–inertial mechanism[25]; and 3) the compressional mechanism[21] are probably modified. This in turn could lead to an improvement in hearing by bone conduction and an air–bone gap in the absence of middle ear or ossicular chain anomalies.

As we have noted earlier, tympanometry with a single low-frequency probe tone "can provide useful diagnostic information for patients with disorders of the tympanum (effusion or abnormal air pressures within the middle-ear cavity), for patients with disorders that affect the tympanic membrane (atrophic scarring, retraction or perforation) and for patients with Eustachian tube dysfunction."[8] Low-frequency, single-component tympanometry, however, is relatively insensitive to lesions that af-

fect the ossicular chain and, from the present study, to lesions that involve enlargement of the endolymphatic duct and sac.

Finally, when a patient presents with a moderate to severe mixed hearing loss, the clinician typically will not have computed tomography or MRI studies available. Still, if the standard (226-Hz) tympanogram is normal, and if the resonance frequency is abnormally low, then the possibility of an enlarged vestibular aqueduct should be included in the differential diagnosis.

CONCLUSION

We have found that standard tympanometry (using a 226-Hz probe tone) produced a normal (type A) pattern for all patients with EVA and for the control subjects with normal hearing. In contrast, multifrequency tympanometry revealed that the resonance frequency for patients with EVA is significantly lower than normal. These findings provide support for our experimental hypothesis. Namely, if enlargement of the VA produces an enlargement of the endolymphatic duct and sac and if this, in turn, increases the volume of endolymphatic fluid and reduces its impedance, then patients with EVA should have a resonance frequency that is lower than normal. Clinically, our findings suggest that EVA should probably be included in the differential diagnosis for a patient who presents with a moderate to severe mixed hearing loss, a normal tympanogram at 226 Hz, and a resonance frequency that is abnormally low.

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