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Clinical Investigation and Mechanism of Air-Bone Gaps in Large Vestibular Aqueduct Syndrome

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Abstract

Objectives—Patients with large vestibular aqueduct syndrome (LVAS) often demonstrate an air-bone gap at the low frequencies on audiometric testing. The mechanism causing such a gap has not been well elucidated. We investigated middle ear sound transmission in patients with LVAS, and present a hypothesis to explain the air-bone gap.

Methods—Observations were made on 8 ears from 5 individuals with LVAS. The diagnosis of LVAS was made by computed tomography in all cases. Investigations included standard audiometry and measurements of umbo velocity by laser Doppler vibrometry (LDV) in all cases, as well as tympanometry, acoustic reflex testing, vestibular evoked myogenic potential (VEMP) testing, distortion product otoacoustic emission (DPOAE) testing, and middle ear exploration in some ears.

Results—One ear with LVAS had anacusis. The other 7 ears demonstrated air-bone gaps at the low frequencies, with mean gaps of 51 dB at 250 Hz, 31 dB at 500 Hz, and 12 dB at 1,000 Hz. In these 7 ears with air-bone gaps, LDV showed the umbo velocity to be normal or high normal in all 7; tympanometry was normal in all 6 ears tested; acoustic reflexes were present in 3 of the 4 ears tested; VEMP responses were present in all 3 ears tested; DPOAEs were present in 1 of the 2 ears tested, and exploratory tympanotomy in 1 case showed a normal middle ear. The above data suggest that an air-bone gap in LVAS is not due to disease in the middle ear. The data are consistent with the hypothesis that a large vestibular aqueduct introduces a third mobile window into the inner ear, which can produce an air-bone gap by 1) shunting air-conducted sound away from the cochlea, thus elevating air conduction thresholds, and 2) increasing the difference in impedance between the scala vestibuli side and the scala tympani side of the cochlear partition during bone conduction testing, thus improving thresholds for bone-conducted sound.

Conclusions—We conclude that LVAS can present with an air-bone gap that can mimic middle ear disease. Diagnostic testing using acoustic reflexes, VEMPs, DPOAEs, and LDV can help to identify a non-middle ear source for such a gap, thereby avoiding negative middle ear exploration.

A large vestibular aqueduct may act as a third mobile window in the inner ear, resulting in an air-bone gap at low frequencies.

Keywords

air-bone gap; audiometry; conductive hearing loss; large vestibular aqueduct syndrome

INTRODUCTION

The term “large vestibular aqueduct syndrome” (LVAS) was introduced by Valvassori and Clemis¹ in 1978 to describe a group of patients with an abnormally large vestibular aqueduct (VA) measuring more than 1.5 mm in diameter on polytomography. A large VA is often associated with a sensorineural or mixed hearing loss. An air-bone gap at the low frequencies has been reported to occur in 15% to 100% of patients with LVAS by various authors.^{1–10}

The mechanism of an air-bone gap in LVAS remains unclear. Some investigators have suspected fixation, loosening, or discontinuity of the ossicles.^{7,11–13} However, the middle ear and the ossicular chain have been reported to be normal on exploratory tympanotomy.^{1,2,5,14} Therefore, speculation has focused on abnormalities of inner ear mechanics as being the cause of the apparent conductive hearing loss. Putative explanations have included 1) a decrease in stapes mobility due to increased perilymphatic or endolymphatic pressure^{2,6}; 2) an unspecified disturbance of cochlear mechanics⁵; 3) a selective facilitation of bone-conducted hearing because of the large VA acting as a third window^{8,9}; and 4) an impairment of air-conducted hearing combined with facilitation of bone-conducted hearing because of the large VA acting as a third window in the inner ear.¹⁵

In this report, we describe investigations into the mechanics of air-bone gaps in LVAS using a variety of diagnostic tests such as tympanometry, acoustic reflexes, measurements of umbo velocity by laser Doppler vibrometry (LDV), vestibular evoked myogenic potentials (VEMPs), and distortion product otoacoustic emissions (DPOAEs). These various noninvasive tests are helpful in the differential diagnosis of conductive hearing losses, including the diagnostic separation of middle ear causes from inner ear causes of an air-bone gap.^{16–27} Our results support the idea that the large VA causes an air-bone gap by acting as a mobile third window within the inner ear. We believe that the large VA affects both air conduction and bone conduction, but in a different manner for each pathway. We present a qualitative model to explain the air-bone gaps observed in LVAS.

MATERIALS AND METHODS

This study was approved by the Institutional Review Board of the Massachusetts Eye and Ear Infirmary. It was a prospective study of patients with LVAS derived from the clinical practices of 3 of the authors (S.N.M., J.B.N., and D.J.L.); the patients had been referred to the Middle Ear Research Unit of the Infirmary for LDV. The diagnosis of LVAS in each case was based on standard radiologic criteria (diameter of VA greater than 1.5 mm) on multidetector computed tomography (CT) scans of the temporal bone.^{1–10} The scans were performed with 0.5-mm collimation and without intravenous contrast.

Each patient had a detailed otolaryngological history-taking and examination. Audiological tests, including pure tone and speech audiometry, tympanometry, and DPOAE testing, were performed by standard clinical procedures.²⁷ The presence or absence of the acoustic reflex for 1,000 Hz or broadband noise was noted for a high-intensity signal between 80 and 100 dB hearing level (HL).

Measurements of umbo velocity were made with LDV as previously described.^{16,17} A sound was introduced into the ear canal, and the umbo velocity produced in response to the sound was measured in a noninvasive manner by LDV in the span of a few minutes. The measured umbo velocity was normalized (divided) by the sound pressure measured in the ear canal, and compared with that in normal ears. In this article, the umbo velocity is described as “normal” if its magnitude was within 1 standard deviation of the mean for normal ears, and as “high normal” if it was larger than 1 standard deviation above the mean but still within the normal range.

The VEMP response was measured by a technique that has also been previously described.¹⁸ The VEMP thresholds were measured with tone bursts of 250, 500, 750, and 1,000 Hz. The VEMP thresholds were categorized as “normal” if they were 70 to 80 dB, as “low” if they were 65 dB or less, and as “elevated” if they were greater than 90 dB HL.

RESULTS

We studied 8 ears from 5 individuals with LVAS. Brief clinical summaries are presented for each patient. The audiological, LDV, and CT scan data are also shown in Figs 1–5. The case reports are followed by a summary of the test results, and implications for the mechanism of the air-bone gap.

Case 1

This 22-year-old patient had had bilateral, stable hearing loss since birth. She had no otalgia, otorrhea, tinnitus, or vertigo. Her history was also notable for bilateral preauricular pits, a branchial cleft cyst or sinus in her neck on both sides, and unilateral renal agenesis. Exploratory tympanotomy of the left ear at 8 years of age had revealed intact and mobile ossicles with a patent round window niche. Evaluation at the time of presentation (age 22 years) showed normal tympanic membranes on otoscopy. The Rinne tests were negative bilaterally. A clinical diagnosis of branchio-oto-renal syndrome was made. Audiometry showed bilateral, large (15 to 65 dB), low-frequency air-bone gaps (Fig 1). Word recognition scores and tympanometric findings were normal bilaterally. Acoustic reflexes were present on the right and absent on the left. The VEMPs were present and normal on each side, with thresholds of 70 dB HL on the right and 80 dB HL on the left. The LDV showed the umbo velocity to be high normal bilaterally, approximately 2 standard deviations above mean normal (Fig 1). The CT scan showed large VAs on both sides (Fig 1). Both cochleas showed 2 rather than 2½ turns. The inner ears did not show dehiscences of any of the semicircular canals or other abnormalities. The middle ears were normal.

Case 2

This 9-year-old boy was discovered to have hearing loss during screening at school. He denied otalgia, otorrhea, and vertigo. There was no family history of hearing loss. Otoscopy revealed normal ear canals and tympanic membranes bilaterally. Audiometry revealed anacusis on the right and a low-frequency, 15- to 35-dB air-bone gap on the left with a word recognition score of 96% (Fig 2). Tympanometric findings were normal bilaterally. Acoustic reflexes and DPOAEs were absent on the right and present on the left. The VEMP responses were present on both sides with low thresholds (65 dB on the right and 55 to 60 dB on the left). The LDV testing showed the umbo velocity to be normal except for a slight dip at 2,000 Hz (Fig 2). The CT scan showed an enlarged VA on both sides with otherwise normal middle and inner ears (Fig 2).

Case 3

This 48-year-old man gave a history of hearing loss since the age of 3 years. He had worn amplification since the age of 5 years. The hearing loss had been slowly progressive over time. He had no otalgia, otorrhea, or vertigo. There was no history of middle ear infections. Otoscopy revealed normal tympanic membranes on both sides. The Rinne test was negative on the right and positive on the left. Audiometry revealed a mixed hearing loss on both sides, with large air-bone gaps (20 to 70 dB) at the low frequencies (Fig 3). Word recognition was poor on both sides (2% on the right and 32% on the left). Tympanometric findings were normal bilaterally. The LDV showed the umbo velocity to be high normal below 600 Hz, and normal at 600 Hz and higher frequencies in both ears (Fig 3). The CT scan showed bilaterally enlarged VAs and minor under-segmentation of the apical and middle turns of the cochleas (Fig 3). The middle and inner ears were otherwise normal.

Case 4

This 42-year-old patient had congenital hearing loss on the left. She had normal hearing on the right until the age of 40 years, when she suffered sudden, idiopathic sensorineural hearing loss without vertigo. She had no history of middle ear infections. Otologic examination revealed normal ear canals and tympanic membranes bilaterally. The Rinne test was positive on the right and equivocal on the left. The Weber test lateralized to the left. Audiometry showed a somewhat flat, purely sensorineural hearing loss on the right and a mixed hearing loss on the left with a low-frequency air-bone gap of 10 to 40 dB (Fig 4). Word recognition scores were 62% on the right and 4% on the left. The LDV revealed the umbo velocity to be close to mean normal on the right, and to be higher than mean normal (by approximately 1 standard deviation) on the left for frequencies below 1,000 Hz (Fig 4). The CT scan showed normal middle and inner ears on the right without enlargement of the VA, whereas on the left, there was an enlarged VA (Fig 4) with a slight dilation of the cochlea at the junction of the basal and middle turns. No other radiologic abnormalities were identified in the middle and inner ears.

Case 5

This 42-year-old woman presented with a long-standing left-sided hearing loss, as well as tinnitus and aural fullness that had been present for about a year. She did not have any vestibular complaints. She had no individual or family history of middle ear disease. Otologic examination revealed normal external auditory canals and tympanic membranes bilaterally. The Rinne test was positive on both sides, and the Weber test lateralized to the left. Audiometry showed normal hearing on the right and a mild, mixed hearing loss on the left with an air-bone gap of 5 to 30 dB at the low frequencies (Fig 5). Word recognition scores were normal on both sides. Tympanometric findings were normal, and acoustic reflexes were present in both ears. The DPOAEs were present on the right but absent on the left. The LDV showed the umbo velocity to be close to mean normal on both sides (Fig 5). A CT scan showed a large VA only on the left (Fig 5). The inter-scalar septum within the left cochlea was inconspicuous. No other abnormalities were identified in the middle or inner ears on the CT scan. A magnetic resonance imaging (MRI) scan showed an enhancing 4-mm-diameter lesion in the central portion of the internal auditory canal on the left, consistent with an acoustic neuroma.

Summary and Interpretation of Results

Eight of the 10 ears studied had a large VA on CT scan. Of these 8, there was anacusis in 1 ear and a low-frequency air-bone gap in the remaining 7. The gap was only seen at frequencies of 1,000 Hz and lower, with the average gap being 51 dB at 250 Hz, 31 dB at 500 Hz, and 12 dB at 1,000 Hz. Measurement of umbo velocity by LDV is helpful in the differential diagnosis of an air-bone gap in ears with an aerated middle ear and an intact tympanic membrane. Our previous work,^{16,17,19,20} as well as that of others,²¹ has shown that LDV measurements in

this setting can differentiate among various disorders such as malleus fixation, stapes fixation, ossicular discontinuity, and superior canal dehiscence (SCD). When compared to measurements in normal ears, ossicular discontinuity results in a marked increase in umbo velocity (greater than 2 standard deviations above normal), ossicular fixations show velocities that are lower than mean normal, and SCDs demonstrate umbo velocities in the normal or high normal range. It has also been shown that an SCD leads to an air-bone gap by acting as a third mobile window in the inner ear that affects both air and bone conduction thresholds.^{15,20,22–25} In the present study, all 7 ears with an air-bone gap and a large VA showed umbo velocities that were in the normal or high normal range, which are not consistent with middle ear disease such as ossicular fixation or discontinuity. There are also some finer details in the LDV data of our patients that showed similarities to LDV data from patients with SCD, such as a tendency for a reduction in the resonance frequency around 700 to 1,000 Hz (seen in Figs 2–4), as well as changes in phase (data not shown). The similarity of these LDV results to those observed in SCD is consistent with the idea that the air-bone gap is due to the large VA acting as a third mobile window in the inner ear.

Additional evidence that the air-bone gap in these ears with a large VA was not the result of middle ear disease comes from the findings that tympanometry was normal in all 6 ears tested, acoustic reflexes were present in 3 of the 4 ears tested, VEMP responses were present in all 3 ears tested, DPOAEs were present in 1 of the 2 ears tested, and exploratory tympanotomy in 1 case showed a normal middle ear. Acoustic reflexes, DPOAEs, and VEMP responses are typically absent when there is true middle ear disease causing a conductive hearing loss.^{15,22,27}

Case 5 is worthy of comment, as there were 2 lesions in the left ear — a 4-mm acoustic neuroma and the enlarged VA. It is difficult to explain the air-bone gap on the basis of the small intracanalicular acoustic neuroma. The presence of acoustic reflexes and LDV data argued against middle ear disease being responsible for the air-bone gap. We believe that the most logical explanation for the gap in this case was the large VA.

Minor developmental abnormalities of the cochlea were noted on CT scan in 6 of the 8 ears with LVAS. Although it is possible that these anomalies could produce the observed air-bone gaps by some as yet undescribed mechanism, the third window mechanism is consistent with the observed gaps.

DISCUSSION

Our observations indicate that the air-bone gap in all 5 patients with LVAS was not the result of a lesion within the middle ear sound transmission system, and are consistent with evidence presented in the literature. For example, acoustic reflexes have been noted to be present in patients with LVAS despite the air-bone gaps.^{6,7} Other investigators have demonstrated that the resonant frequency of the middle ear is significantly reduced in LVAS (and the resonant frequency is increased in ossicular fixation).^{7–9} The VEMP responses have also been reported to be present (sometimes with low thresholds) in ears with air-bone gaps and LVAS.^{7,26} Finally, several investigators have surgically explored the middle ear in LVAS and have not found lesions of the ossicular chain or oval or round windows.^{1,2,5,14}

Clinical Implications

Large vestibular aqueduct syndrome (along with SCD) should be considered in the differential diagnosis of conductive hearing loss in the presence of an intact tympanic membrane and an aerated, otherwise healthy middle ear. A typical scenario would involve a pediatric patient with a unilateral or bilateral, congenital, conductive or mixed hearing loss showing air-bone gaps at the low frequencies. A heightened awareness of the diagnostic possibility of LVAS should

serve to reduce the number of patients who undergo negative middle ear explorations. We recommend inclusion of acoustic reflex testing as part of the diagnostic workup prior to middle ear exploration. Acoustic reflexes are typically absent in true middle ear disease, but would be present in LVAS. If available, VEMP testing, measurement of DPOAEs, and LDV testing can also assist in the diagnosis. When suspected, a CT or MRI scan will help to make a definitive diagnosis of LVAS.

The clinical appreciation of a low-frequency air-bone gap highlights the necessity of obtaining bone conduction threshold data at 250 Hz. This is not a common practice in some clinics, as thresholds at 250 Hz are sometimes characterized as “not reliable.” This impression may have arisen partly because of cases such as those presented here (as well as patients with SCD), in which no middle ear lesion can be found in the presence of a persistent air-bone gap. Bone conduction thresholds at 250 Hz should be routinely reported, particularly in light of new mechanisms that explain these air-bone gaps,^{15,22,23} and because of the benefit in the eventual diagnosis. Testing to threshold (ie, below 0 dB HL; eg, case 1) is also indicated in light of possible hypersensitivity by bone conduction, and is particularly important for calculation of adequate masking in these cases.

The acoustic reflex performs a nontraditional function in disorders such as LVAS and SCD. The object of the test in these examples is to differentiate disease from normality of the ossicles. With this end in mind, the traditional approach is not indicated, wherein tonal thresholds are determined for ipsilateral and contralateral stimulation in order to assess the status of the cochlea and the reflex arc involving the cochlear and facial nerves.²⁷ Any single clear change in admittance, from any source or stimulus, is sufficient to rule out ossicular disease. One suggested approach is to stimulate with high-level (90 dB HL) broadband noise ipsilaterally. High-level broadband noise is the stimulus most likely to elicit a strong reflex, and if absent, this is a clear indication of fixation without lengthy searching for thresholds. If, on the other hand, the contralateral ear is better, the better ear could also be stimulated (contralateral reflex) to show the capability for movement of the ossicles in question. Searching for threshold by reducing the stimulus may speak to cochlear or neural factors, but a strong response above threshold will provide the clearest indication of normality of the ossicles. It is worth noting that both VEMP and LDV testing can be performed when hearing is too poor to expect a reflex, and are therefore robust additional tests.

Mechanism of Air-Bone Gap Resulting From LVAS

We can gain insight into the mechanics of sound transmission in LVAS by analogy to the air-bone gap in SCD. Our laboratory has conducted a series of studies to investigate the mechanisms by which an air-bone gap arises in SCD, including theoretical model analyses,²³ experimental measurements in a cadaveric human temporal bone preparation,²⁰ experiments in a chinchilla model of SCD,^{23–25} and measurements of middle ear sound transmission in patients with SCD.^{20,23} These investigations have suggested that an SCD results in an air-bone gap by acting as a third mobile window in the inner ear that results in elevation of thresholds for air-conducted sounds and reduction of thresholds for bone-conducted sounds. We have previously hypothesized that a similar mechanism may also explain the air-bone gap in LVAS.¹⁵

A schematic representation of our conceptual mechanism of LVAS is shown in Fig 6. Air-conducted sound stimuli enter the vestibule through motion of the stapes (Fig 6A). We hypothesize that the enlarged VA allows for a portion of this acoustic energy to be shunted away from the cochlea, thus resulting in a loss of hearing by air conduction (Fig 6B). The effect of the enlarged VA on bone conduction thresholds can be understood as based on the compressional mechanism of bone conduction. In the normal ear, compression of inner ear fluid by bone-conducted sound results in a hearing percept because of an inequality in the

impedance between the scala vestibuli side and the scala tympani side of the cochlear partition (Fig 6C), which in turn is due to a difference between the impedance of the oval and round windows, respectively. This inequality leads to a pressure difference across the cochlear partition, resulting in motion of the basilar membrane that leads to perception of bone-conducted sound. An enlarged VA increases the difference between the two sides of the cochlear partition by lowering the impedance on the vestibuli side, thereby improving the cochlear response to bone conduction (Fig 6D). Supranormal bone thresholds may be evident in some instances, such as the -5 -dB thresholds seen at 250 Hz in cases 1 and 2. Many patients, however, have accompanying true sensorineural hearing loss in LVAS, which can mask improved bone conduction thresholds due to the third window mechanism. In our previous model analysis of SCD,²³ it was shown that the hearing loss would be restricted to frequencies below 2,000 Hz, on the basis of the physics of sound and the anatomic dimensions of the human labyrinth. We believe that a similar situation exists with LVAS, so that the observed air-bone gap is generally confined to the lower frequencies.

The third window hypothesis in LVAS, as we envision it, is somewhat different from that proposed by other authors.^{8,9} These other authors believe that the enlarged VA results in an air-bone gap because of improved thresholds for bone-conducted sound via the brain and the cerebrospinal fluid pathway.^{8,9} We believe that there is a worsening of thresholds for air-conducted sounds and improved bone-conducted hearing (the latter via the compressional pathway of bone conduction).

The model we have espoused is a qualitative one. Future work could involve development of a more quantitative model, as well as experimental simulations in cadaveric temporal bones or animal preparations. Research is also needed to explain the variability regarding the presence or absence of an air-bone gap, as well as the variability in the size of the gap from one ear to another, in LVAS. Potential factors that may explain such variability that need to be investigated include the physical dimensions of the LVAS, especially at the junction between the VA and the labyrinth, as well as the ratio between the cochlear impedance and the middle ear impedance in a given ear. It is to be noted that the data in the present study were generated primarily from adults; only 1 case was in a child. Although we believe that the mechanisms of the air-bone gap are similar in children and adults, this belief needs to be investigated and verified in future studies.

Nomenclature of Air-Bone Gap and Hearing Loss in LVAS and Other Third Window Lesions

Various terms such as pseudoconductive loss, inner ear conductive loss, cochlear conductive loss, etc, have been used to describe the air-bone gap seen in patients due to abnormalities of the inner ear such as LVAS or SCD. Some of these terms can be ambiguous. We recommend that clinicians and researchers adopt terms that are more explicit and less confusing.

We believe that the term “air-bone gap” is appropriate, because it accurately conveys the audiometric finding of a difference between air and bone conduction thresholds, regardless of the source of the air-bone gap. One can go on to further characterize an air-bone gap by describing the location of the lesion causing the gap (external, middle, or inner ear) or the audiometric artifact (a collapsing ear canal or improper headphone placement)

The term “pseudoconductive” hearing loss carries some degree of ambiguity. It implies that the observed hearing loss is not conductive in nature. By default, then, such a loss has to be sensorineural. However, in patients with LVAS or SCD, the air-bone gap is not the result of a lesion of the sensory or neural structures of the cochlea. Similarly, the term “cochlear conductive loss” is also ambiguous. The term was originally used to refer to a pure sensorineural hearing loss (as determined by an audiogram) wherein no abnormalities were observed in the inner ear on light microscopy that could explain the hearing loss.²⁸ However,

the term “cochlear conductive loss” has also been used to refer to an air-bone gap (as determined by an audiogram) that is believed to result from a mechanical problem in the cochlea.⁵

Therefore, we recommend that the terms pseudo-conductive loss and cochlear conductive loss should not be used. Hearing losses should be classified as sensorineural (no air-bone gap on audiometry) or conductive (presence of an air-bone gap on audiometry). Conductive hearing losses should be subclassified according to the site of lesion, such as the external, middle, or inner ear. Thus, one would characterize LVAS as resulting in a conductive hearing loss of inner ear origin.

SUMMARY AND CONCLUSIONS

We have presented 7 ears from 5 individuals with LVAS and air-bone gaps at the low frequencies. Audiological testing combined with acoustic reflex, VEMP, DPOAE, and LDV analyses in these ears indicated that the hearing loss was not the result of middle ear disease. A model was presented to explain the hearing loss, based on a large VA acting as a third mobile window in the inner ear. We hypothesize that the air-bone gap is produced by shunting of air-conducted sound away from the cochlea through the large VA that elevates air conduction thresholds. We also propose that the third mobile window associated with a large VA increases the difference in impedance between the scala vestibuli side and the scala tympani side of the cochlear partition, improving thresholds for bone-conducted sound. Clinicians should include LVAS in the differential diagnosis of congenital conductive hearing loss. An accurate diagnosis can be made on the basis of acoustic reflex, DPOAE, VEMP, and LDV testing, as well as CT or MRI scanning, thereby avoiding negative middle ear explorations.

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Fig 1.

(Case 1) Female patient, 22 years old. Audiogram shows bilateral air-bone gap at low frequencies. Laser Doppler vibrometry (LDV) data are plotted as magnitude of umbo velocity (normalized by ear canal sound pressure) versus frequency. Shaded area represents normative data: mean $\pm$ 1 SD. Umbo velocity was “high normal” on both sides (approximately 2 SD above mean normal). Computed tomography (CT) scan was similar on two sides with bilaterally enlarged vestibular aqueducts (VAs). Axial cut from right ear is shown (arrow points to VA).

Fig 2.

(Case 2) Male patient, 9 years old. Data from left ear are shown. There was low-frequency air-bone gap on audiometry. LDV testing showed umbo velocity to be normal except for slight dip around 2,000 Hz. Axial cut from CT scan with enlarged VA (arrow) is shown.

Fig 3.

(Case 3) Male patient, 48 years old. He had bilateral mixed hearing loss with large air-bone gaps at low frequencies. Umbo velocity was in high normal range at low frequencies and normal at frequencies 600 Hz and higher. CT scan was similar on two sides with bilaterally enlarged VAs. Axial cut from right ear is shown (arrow points to VA).

Fig 4.

(Case 4) Female patient, 42 years old. Data from left ear are shown. Audiometry showed mixed hearing loss with low-frequency air-bone gap. Umbo velocity was normal (within 1 SD) except at 3 kHz and higher. Axial cut of CT scan with enlarged VA (arrow) is shown.

Fig 5.

(Case 5) Female patient, 42 years old. Data from left ear are shown. Audiometry showed mild, mixed hearing loss with low-frequency air-bone gap. Umbo velocity was normal. Axial cut of CT scan with enlarged VA (arrow) is shown.

Fig 6.

Schematic representations of mechanism of air-bone gap in large vestibular aqueduct syndrome (LVAS). **A)** Normal ear, air conduction. Air-conducted sound stimuli enter vestibule through motion of stapes. There is pressure difference between scala vestibuli and scala tympani, resulting in motion of cochlear partition. Volume velocities of oval and round windows are equal in magnitude but opposite in phase. **B)** Enlarged aqueduct, air conduction. It is hypothesized that large VA allows portion of acoustic energy entering vestibule through motion of stapes to be shunted away from cochlea. Shunting occurs primarily at low frequencies, resulting in hearing loss by air conduction. **C)** Normal ear, bone conduction. Compression of inner ear fluid by bone-conducted sound results in hearing percept because of inequality in impedance between scala vestibuli side and scala tympani side of cochlear partition. This inequality is primarily due to difference in impedance between oval and round windows. As result, there is pressure difference across cochlear partition, resulting in motion of basilar membrane that leads to perception of bone-conducted sound. **D)** Enlarged aqueduct, bone conduction. Enlarged VA increases difference between impedance on scala vestibuli side and scala tympani side of cochlear partition by lowering impedance on vestibuli side, thereby improving cochlear response to bone conduction. Supranormal bone thresholds may be evident in some instances, such as -5-dB thresholds seen at 250 Hz in cases 1 and 2. Many patients, however, have accompanying true sensorineural hearing loss in LVAS, which can mask improved bone conduction thresholds due to this mechanism.