The Audiology of Otosclerosis

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KEYWORDS

• Carhart notch • Immittance measurement and otosclerosis • Reflectance
• Wideband acoustic immittance and otosclerosis • Middle ear muscle reflex
• Power absorbance and otosclerosis • Hearing aids and otosclerosis
• Tinnitus and otosclerosis

KEY POINTS

- For most patients with otosclerosis, audiologic biomarkers include reduced middle ear compliance as revealed by tympanometry, and a 10- to 15-dB reduction in sound transmission via bone conduction most often in the vicinity of 2000 Hz (known as Carhart notch).
- Wideband acoustic immittance is an effective technique in identifying middle ear pathologies, such as otosclerosis; it can provide all the useful information that could be obtained from conventional and multifrequency tympanometry and additional information on the transfer of energy into the middle ear system across much wider range of frequencies.
- Middle ear resonance frequency shifts to higher frequency regions in most of the otosclerotic ears.
- In addition to middle ear ossicular surgery, hearing aids and implantable hearing devices are alternative approaches for the management of hearing loss in patients with otosclerosis.
- Tinnitus sound therapy and cognitive behavioral therapy are successfully used for the management of tinnitus in the otosclerotic population.

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INTRODUCTION

For many otologists and audiologists, otosclerosis is not a puzzling condition anymore. Advances in diagnostic and therapeutic procedures have provided a vast number of patients with otosclerosis with proper management. This article is designed in a fashion that enables otologists in better diagnosis and management of otosclerosis with the use of audiologic procedures. The accuracy of audiometric air-bone gap, appropriate use of masking techniques, and immittance measurements can completely influence the decisions made by otologists for the surgical management of otosclerosis. Otologists rely on the precision of the audiologic results and determination of the degree of the conductive component. Therefore, a precise audiologic work-up is a crucial part of the diagnostic protocol for otosclerosis. This article reviews the audiologic diagnostic test battery and the audiologic management of auditory effects of otosclerosis.

AUDIOMETRIC PATTERNS

As with other middle ear disorders, otosclerosis reduces sound-related energy passing from the tympanic membrane to the inner ear. Fixation and resultant stiffening of the ossicular chain almost always produces a hearing loss, particularly for lower-frequency sounds. The characteristic pattern of hearing loss in otosclerosis is useful in diagnosing the disease.[1–3] The diagnostic value of hearing assessment is enhanced when such test procedures as pure tone audiometry, tympanometry, and acoustic reflexes are combined into a test battery. Indeed, for most patients with otosclerosis, a unique pattern of findings for an appropriate collection of auditory tests almost always contributes to early and accurate diagnosis. Basic hearing test findings in patients with otosclerosis are summarized in **Table 1**.

Table 1
Patterns of basic auditory findings in patients with the diagnosis of otosclerosis

Procedure	Findings
Pure tone audiometry	
Air conduction	Hearing loss greater for low frequencies.
Bone conduction	Apparent decrease in bone conduction thresholds sometimes with a notching deficit at 2000 Hz (Carhart notch). Actual bone conduction hearing is typically normal.
Audiometric Weber test	Perception of low-frequency pure tone stimuli in the ear with conductive hearing loss.
Sensorineural acuity level test	Presence of an air-bone gap and confirmation of normal bone conduction hearing.
Acoustic immittance measures	
Tympanometry	Shallow type A tympanogram reflecting increased stiffness of the ossicular chain (see immittance measurement section for further discussion).
Acoustic reflexes	Absence of stapedial acoustic reflex activity even in patients with minimal air-bone gap and conductive hearing loss. Atypical acoustic reflex pattern in patients with very early subclinical otosclerosis.
Otoacoustic emissions	Otoacoustic emissions cannot be detected in patients with otosclerosis and conductive hearing loss. Recovery of detectable otoacoustic emissions is possible in patients following microtraumatic stapedotomy.

Representative pure tone audiometry findings for one ear of a patient with otosclerosis are shown in Fig. 1. There is a conductive hearing loss with considerably poorer hearing sensitivity for air versus bone conduction hearing. Bone conduction hearing is generally normal with the exception of a distinct notch-like decrease in bone conduction thresholds in the audiogram region of the 2000 Hz. The horizontal dotted line indicates actual bone conduction hearing whereas the bone conduction thresholds reflect an apparent deficit in sensory function. "Mechanical modifications" and effects of middle ear resonant frequency in bone conduction hearing associated with stapes fixation in patients with otosclerosis have been appreciated since the 1940s (discussed later). Indeed, normal bone conduction hearing sensitivity is unusual for patients with otosclerosis. Multiple theories have been offered for the negative effect of middle ear abnormalities on the response to bone conduction stimulation. However, evidence and agreement in support of a single mechanism are lacking.

Named after the well-known audiologist who first described it in detail,[1,4] Carhart notch has for more than 60 years been one of the most recognizable audiometric features of otosclerosis. Carhart[1] described an average decrease in bone conduction thresholds of 5 dB at 500 Hz, 10 dB at 1000 Hz, 15 dB at 2000 Hz, and 5 dB at 4000 Hz, as illustrated in Fig. 1.

More recent studies raise three general questions about the diagnostic value and specificity of a notching deficit in bone conduction thresholds at 2000 Hz.[5–8] First, bone conduction hearing thresholds are often decreased also at other test frequencies in patients with the diagnosis of otosclerosis. Researchers have observed for patients with the diagnosis of otosclerosis the possibility of a notching deficit in bone conduction thresholds in the low-, mid-, and high-frequency region, not just at 2000 Hz.[9,10]

Second, Carhart notch at 2000 Hz is not invariably observed in patients with the diagnosis of otosclerosis or fixation of the ossicular chain. A group of scientists reported a 2000-Hz notch in bone conduction thresholds for only 31% of 102 patients

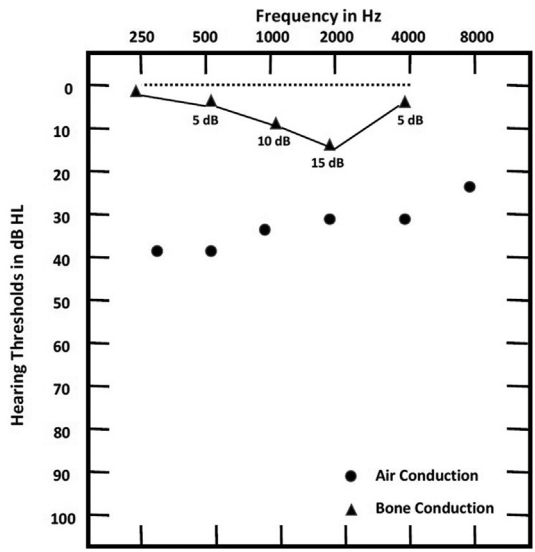

Fig. 1. Typical air and bone conduction hearing threshold patterns for a patient with otosclerosis. Notice the appearance of Carhart notch in bone conduction hearing at 2000 Hz. The *dotted line* indicates true bone conduction hearing or "cochlear reserve." (*Courtesy of* James W. Hall III, PhD, Salus University, Elkins Park, PA.)

with stapes fixation.[7] Finally, related to this second point, patients with etiologies for conductive hearing loss other than otosclerosis may show a notching deficit in bone conduction thresholds at 2000 Hz. Studies have reported the presence of Carhart notch in one-third of a series of 75 patients with congenital aural atresia.[8] Carhart notch pattern was shown only for 30% of patients with malleus or incus fixation and for 26% of 19 patients with detachment or discontinuity at the malleus and incus joint.[7] The resonant frequencies of the middle ear and particularly the ossicular chain seem to be in the vicinity of 2000 Hz, which is potentially why a reduction at 2000 Hz is seen in a good number of patients with stapes fixation as seen in otosclerosis.[4,7,8] In advanced cases of otosclerosis, conductive hearing loss develops into mixed hearing loss. Additionally, in cases with cochlear otosclerosis, moderate to profound sensorineural hearing losses are commonly observed in clinical practice.

Two additional pure tone hearing tests deserve mention because they sometimes contribute to the accurate assessment of auditory status in patients with otosclerosis. One is the audiometric Weber test and the other procedure is the sensorineural acuity level or sensorineural acuity level test.[2,3,11,12] The sensorineural acuity level technique provides valuable clinical information and plays a unique role in clinical audiology when performed with insert earphones and used as a supplement to conventional bone-conduction measurements for confirming ear-specific information on sensory hearing thresholds (see **Table 1**).

As summarized in **Table 1**, three other auditory findings are consistent with fixation of the ossicular chain and typical of patients with otosclerosis, in addition to the conductive hearing loss and Carhart notch. One is a shallow type A tympanogram, referred to as type As, which reflects abnormal restriction of the ossicular chain (discussed later).[3,13] A second typical finding is the absence of normal acoustic reflex activity, even for patients who have little evidence of conductive hearing loss with pure tone audiometry.[2,3] Indeed, the presence of acoustic reflex activity at expected intensity levels, that is, about 85 dB for pure tone stimuli, essentially rules out fixation of the ossicular chain and otosclerosis. Third, word recognition scores in quiet are good or excellent in most patients with otosclerosis, even in those with some apparent deficit in bone conduction hearing thresholds.

We conclude this discussion of auditory findings in otosclerosis with a few comments about the possible application of otoacoustic emissions (OAEs). There is a general consensus that OAEs are not recordable in patients with middle ear dysfunction including those with fixation of the ossicular chain and otosclerosis. However, several recent published papers describe a potential role for OAEs in the evaluation of auditory function following "microtraumatic stapedotomy."[14,15] Although results are inconsistent among studies and variable among patients, there are reports of the emergence of detectable OAEs in the frequency region of 1000 to 1500 Hz perhaps associated with normalization of the resonance frequency of the middle ear following microtraumatic stapedotomy.

MIDDLE EAR ANALYSIS IN OTOSCLEROTIC EARS

For clinicians, middle ear analysis is the most important diagnostic component of otosclerosis. Many have encountered cases with a conductive pathology and normally appearing tympanograms where the nature of underlying pathology is not clear. Simply put, not all of the ears with otosclerosis show a reduced tympanometric compliance and not all of the tympanograms with reduced compliance are caused by otosclerosis. The following section describes the science behind differentiation of the underlying middle ear pathologies with the use of immittance measurements.

Immittance Measurements

Immittance measurement has been used for several decades in the assessment of middle ear disorders. Immittance measurement consists of tympanometry and middle ear muscle reflex (MMR). Individuals with otosclerosis typically present a conductive hearing loss, sometimes type As or normal type A tympanograms,[16,17] absent MMRs, and normal otoscopic results. The normal otoscopy with conductive hearing loss is not distinctive to otosclerosis.[18] Similar patterns have been observed in cases of superior canal dehiscence and ossicular discontinuity.[19] However, MMRs are present in superior canal dehiscence and a type Ad tympanogram is observed in ossicular chain discontinuity. Differentiation of middle ear pathologies with the use of immittance measurements can sometimes be paradoxic.

Tympanometry

Tympanometry is a safe and quick method for assessing middle ear function. In this technique, a pliable probe is sealed in the external ear canal. Then a sound is presented while the air pressure is changed within the ear canal. The sound pressure level monitored at the probe tip provides an index of the ease with which acoustic energy flows into the middle ear system, which is referred to as acoustic admittance (Ya). Currently, tympanometry is mainly conducted at a conventional low probe tone frequency. Tympanometry performed at conventional low probe tone frequency (226 Hz) cannot identify most of the lesions that specifically affect the ossicular chain. For example, information provided by a conventional 226-Hz tympanogram is typically inadequate for distinguishing a normal middle ear from otosclerotic (stapes fixation) ears.[20–26]

Different parameters can be obtained from a conventional low probe tone frequency tympanogram. Two absolute parameters, static admittance (Y_{tm} – admittance at the level of the tympanic membrane) and tympanometric width in daPa, are most often derived from conventional low probe tone frequency tympanometry. Several studies have compared Y_{tm} in healthy and otosclerotic ears.[16,25–30] These studies have consistently shown that, on average, Y_{tm} tends to be lower in otosclerotic ears. However, the extensive overlap in the distributions of Y_{tm} for these two groups at conventional low probe tone frequency severely limits the diagnostic utility of this measure.

It has been shown that an abnormality is most obvious when the probe tone frequency approaches the frequency at which middle ear vibrates most readily.[26,31–33] This frequency is called the resonant frequency. Middle ear pathologies, such as otosclerosis, affect the resonant frequency of the middle ear system. The greatest impact of middle ear pathology on the Y_{tm} is at frequencies close to the resonant frequency.[26,34] Therefore, Y_{tm} measured in the vicinity of the resonant frequency may provide the most useful information regarding the differential diagnosis of middle ear pathologies. Several clinical and laboratory studies have reported prominent differences between healthy and otosclerotic ears[25,26,33,35–37] when Y_{tm} recorded using higher probe tone frequencies or resonant frequency were compared between healthy ears and otosclerotic ears.

Tympanometric shape has also been reported to be affected by otosclerosis. A measure that is, most commonly used to index the sharpness of the tympanometric peak at conventional low probe tone frequency is the tympanometric width. Some studies have reported narrower tympanometric peaks in otosclerotic ears than healthy ears.[25,38–40]

The appearance of multifrequency devices has made it possible to derive immittance subcomponents, susceptance (B) and conductance (G), and to perform tympanometry across a wide range of probe tone frequencies. Recent studies suggest

that identification of otosclerosis (or stapes fixation) is improved using measures derived from multifrequency tympanometry or by combining tympanometric variables in specific ways.[25,26,31,33,41,42]

One potentially useful parameter that is derived from multifrequency tympanometry is an estimate of the middle ear resonant frequency. The resonant frequency of the middle ear system may be shifted higher or lower compared with healthy ears by various pathologies. One major effect of otosclerosis is to increase the stiffness of the middle ear system resulting in a shift of the middle ear resonant frequency to the higher values. In the case of otosclerosis, the resonant frequency has been shown to be significantly higher than healthy ears.[21,25,26,43–47]

Middle Ear Muscle Reflex

MMR is measured by monitoring the change of the immittance, either a decrease in admittance or an increase in impedance, in the ear canal in response to a sufficiently loud sound. Stapedial muscles contract in both ears simultaneously in response to a sufficiently loud sound presented to one or both ears. This contraction is recorded in either ear by monitoring the change of immittance, which is time-locked to the stimulus presentation. To elicit stapedial muscle contraction in response to a loud sound, middle ear (conductive system), cochlea, 8th cranial nerve, and stapedial branch of the 7th cranial nerve should be intact. MMR is an excellent tool in conjunction with tympanometry to detect the presence or absence of the middle ear disorders including otosclerosis.

Normally, MMR is absent in presence of a modest conductive hearing loss of only 20 dB.[48] Typically, in cases of unilateral otosclerosis, MMR is absent in the ipsilateral mode (stimulus and probe tone are presented to the affected side-probe ear). However, MMR is elevated or absent in contralateral mode depending on the severity of the conductive hearing loss. It should be noted that contralateral MMR in the unaffected side is also absent when the probe is placed in the affected side (probe effect). The reason for the absence of MMR in the probe ear is that it is not possible to monitor the changes in immittance as a result of the stapedial contraction likely because of the stiffening of the ossicular chain, which prevents stapedial muscle to evoke a measurable change in the immittance.[48]

It should be noted that in early stages of otosclerosis a biphasic reflex response (an on-off effect also known as a diphasic response) has been observed.[49] This effect has been observed even before the commencement of an air-bone gap in the otosclerotic ears.[50] The biphasic middle ear reflex response is characterized by a sudden increase in admittance (a paradoxic response, as stapedial muscle contraction, should result in a decrease in admittance) by switching the stimulus on and off, which surrounds a central plateau at 0.

Wideband Acoustic Immittance

Wideband acoustic immittance (WAI) (**Fig. 2**) is a new middle ear assessment technique that has enabled researchers and clinicians to quantify the reflected, or the absorbed energy in the ear canal across a wide range of frequencies typically between 250 and 8000 Hz.[48] Power absorbance (PA) is a ratio of absorbed power over the incident power and varies between 0 and 1. A value of 0 means all sound energy has been reflected back and a value of 1 means all sound energy has been absorbed by the middle ear system.[48]

WAI has several potential advantages over conventional tympanometry. The technique measures over a large range of frequencies (250–8000 Hz). It is also very fast, taking only a couple of seconds to perform. Additionally, the magnitude of the PA

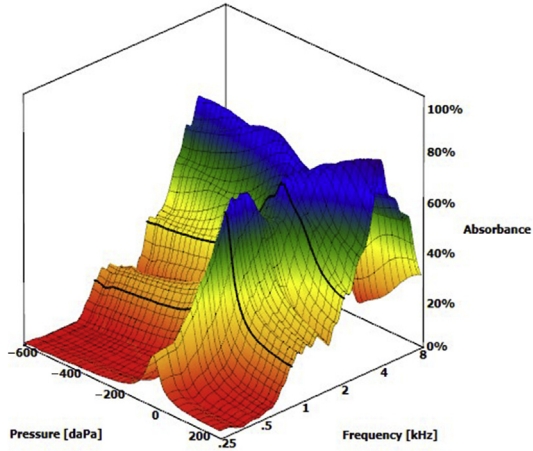

Fig. 2. Wideband acoustic immittance tracings showing three-dimensional multifrequency evaluation and power absorbance of the middle ear. (*Courtesy of* Interacoustics Audiology Solutions, Middelfart, Denmark; with permission.)

does not depend on the distance between the probe tip and the eardrum and so the location of the probe in the ear canal is not as critical as it is in tympanometry in children and adults.[51] Finally, WAI can be run at ambient pressure and does not require pressurization of the ear canal.[52] It is, however, possible to run a pressurized WAI measurement by varying the pressure in a manner identical to tympanometry.[53] At ambient pressures, healthy adults show a pattern of low absorption in the low frequencies, which increases to a maximum between 1000 Hz and 4000 Hz before decreasing again at high frequencies.[54] **Fig. 3** demonstrates this pattern in a normal-hearing individual.

Growing body of the literature suggests that WAI is a good indicator of middle ear pathologies in neonates, children, and adults.[55–63] Compared with conventional 226-Hz tympanometry, WAI may provide for a more sensitive test in evaluating middle ear disorders and conductive hearing loss.[52,64,65] In contrast to conventional 226-Hz tympanometry, WAI is significantly more sensitive to ossicular pathologies.[18] Moreover, the patterns of absorbance vary depending on the status of the middle ear and thus different pathologies result in different patterns of absorbance. Generally, a stiffening pathology results in decreased absorbance over a specific frequency range. For example, otosclerotic ears demonstrate significantly increased reflectance between 400 Hz and 1000 Hz.[18] Researchers found that PA was the most effective way of identifying ears with otosclerosis compared with 226-Hz tympanometry and multifrequency tympanometry.[18] PA was able to identify otosclerosis in 82% of their sample and had a false-positive rate of 17.2%. The research suggests that the use of PA in conjunction with other tools for assessment of middle ear function will improve the identification of otosclerotic ears in a clinical setting.[18] **Fig. 4** demonstrates an example of PA in surgically confirmed otosclerotic ears. The PA was obtained before the surgery and fixation of the stapes was confirmed during the surgery.

AUDIOLOGIC INTERVENTION FOR OTOSCLEROSIS

Auditory complications of otosclerosis include hearing loss and tinnitus. Involved patients rarely complain about sound sensitivity disorders, such as hyperacusis, and the

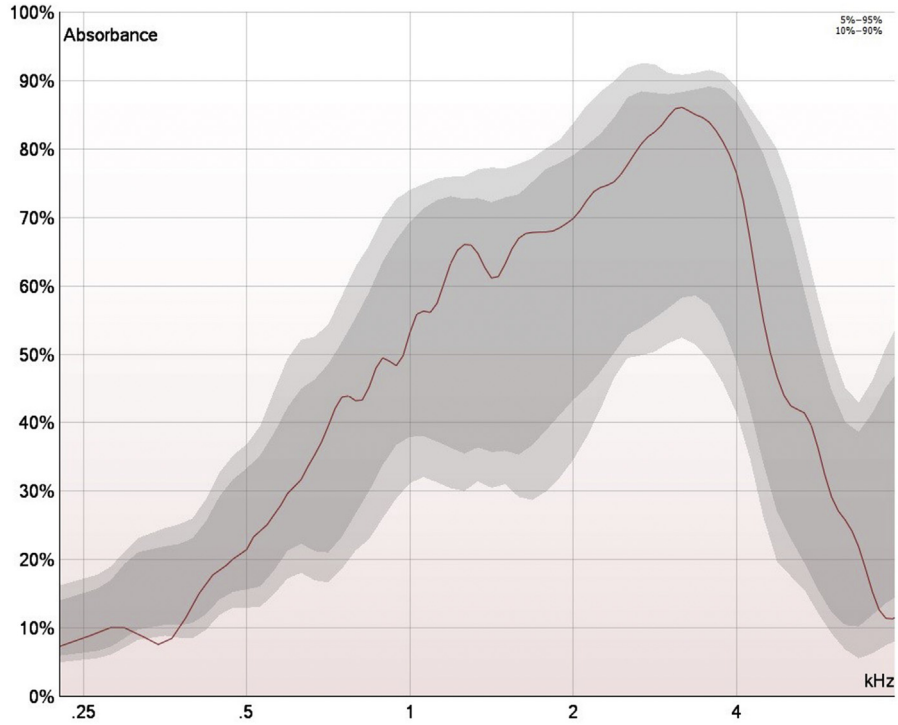

Fig. 3. Power absorbance in a normal-hearing adult. The y-axis is absorbance in % and the x-axis is the frequency in Hz. The shaded areas represent 80% (*dark gray*) and 90% range (*light gray*) of the normative data. (*Courtesy of* Navid Shahnaz, PhD, University of British Columbia, Vancouver, BC, Canada.)

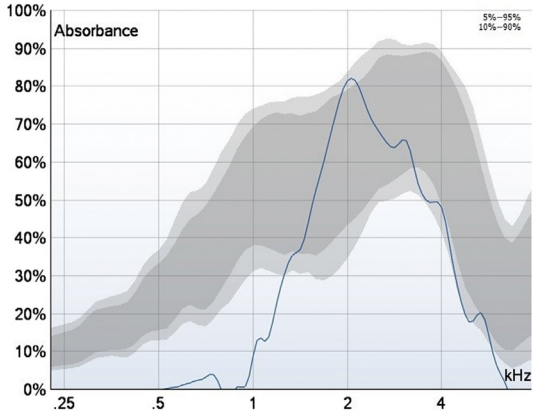

Fig. 4. Power absorbance in a surgically confirmed otosclerotic ear. Note that the absorption less than 1500 Hz is significantly reduced compared with the 90% range for normal individuals (*shaded area*). The y-axis is absorbance in % and the x-axis is the frequency in Hz. (*Courtesy of* Navid Shahnaz, PhD, University of British Columbia, Vancouver, BC, Canada.)

reports of vertigo and balance disorders are not common in presurgical otosclerotic ears. Many patients with otosclerosis are treated with otologic surgery; however, occasionally patients may choose amplification instead of surgery for medical complications, such as stapes gusher or dehiscence of anterior semicircular canals, which may be revealed by high-resolution computed tomography scans.[66] A survey of 184 otologists indicated that hearing aids are advised before surgery.[67]

Hearing Aids and Implantable Auditory Devices

Hearing aid evaluation should always be discussed and offered to have a well-informed consent before to surgery. Occasionally there are patients who do not select surgical management as a solution for their hearing loss and choose amplification. Additionally, because of some unforeseen circumstances, patients' hearing sensitivity does not improve or even worsen after surgical intervention[68] and patients are advised to use hearing aids. Use of hearing aids has been helpful in the management of hearing loss postsurgery.[69–71] Hearing loss caused by otosclerosis can also exacerbate because of sensorineural involvement[72] and this similarly heightens the inclusion of amplification and hearing management protocols in this population.

Modern hearing aids are highly advanced and small.[73] **Fig. 5** demonstrates contemporary hearing aids. Most of today's hearing aids are digital and have the ability of super computation and signal processing. The employment of wireless technology, such as Bluetooth, has enabled users to stream their telephone conversation and music or news to their hearing aids reducing the stigma of hearing aid use. Otologists should encourage their patients to use hearing aids and emphasize the role of neuroplasticity and enhancement of auditory function for the hearing impaired. There is ample evidence in the literature that supports the improvement of auditory function with amplification not only in patients with sensorineural hearing loss but also in those with conductive pathology.[74–77]

Those patients who choose amplification should receive ample amount of time for rehabilitation, orientation, verification, and validation by their audiologists. Successful hearing aid users usually are the ones who communicate effectively with their audiologists about their hearing aids. The advanced clinical standards and practice guidelines emphasize the role of real ear measurements in proper amplification and

Fig. 5. Modern hearing aids. These hearing aids can stream acoustic signals from electronic devices, such as cell phones and tablets, directly to the hearing aids. (*Courtesy of* Starkey Laboratories, Eden Prairie, MN; with permission.)

verification of hearing devices. Unfortunately, most of the over-the-counter hearing aids lack such standards of care.

The amplification management of hearing loss in otosclerosis can also be accomplished with the use of implantable technology. Auditory prosthesis and implantable devices, such as bone-anchored hearing aids[78] and Bonebridge,[79] have been used as alternatives for conventional hearing aids. These implantable devices provide direct signal transmission through bone conduction (ie, bypassing the middle ear with a conductive pathology, such as otosclerosis), which results in direct stimulation of the cochlea. The bone-anchored hearing aid approach also has been used in pathologies, such as congenital atresia and chronic otitis media.[78] In far advanced otosclerosis, profound or total hearing loss can be detected. These cases of far advanced otosclerotic ears have been managed by cochlear implants with great success.[80,81]

Tinnitus Management in Patients with Otosclerosis

For a large number of patients with otosclerosis, tinnitus can be as annoying as hearing loss. Tinnitus caused by otosclerosis has been associated with reversible modifications in the central auditory pathway because of conductive hearing loss.[82]

In many cases of patients with otosclerosis, tinnitus may improve following surgical intervention.[83] Current research indicated tinnitus improvement of 85% of cases within 6 months following stapedectomy.[83] A recent study has shown improvement of low-frequency tinnitus following stapedectomy; however, the researchers of the same study also reported that high-frequency tinnitus persists following surgical intervention.[84] The improvement of tinnitus also has been reported in patients with stapedotomy.[85]

Tinnitus improvement following stapedectomy is age-related. It has been shown that younger patients with otosclerosis may get relief from tinnitus following surgical intervention when compared with the older subjects.[86] In some cases, tinnitus may persist or even become louder after the surgery. Therefore, tinnitus management and intervention is an important component of the postsurgical management of otosclerosis.

Tinnitus management for patients with otosclerosis falls in the line of tinnitus management protocols that are used for those with sensorineural hearing loss caused by such conditions as noise-induced hearing loss or degenerative changes of the auditory system caused by aging. Tinnitus management includes such approaches as counseling, sound therapy, acoustic enrichment, and use of amplification.[87,88] Many patients report no significant perception of tinnitus with the use of hearing aids. The masking effect of amplification diminishes the patient's awareness of his or her tinnitus. In some cases, particularly in those patients with no significant hearing loss, cognitive behavioral therapy has been used for better coping with tinnitus.[89,90] The cognitive behavioral therapy and potentially sound therapy have been supported as effective methods for tinnitus management by practice guidelines presented by academic and clinical professionals.[91] Many patients habituate to their tinnitus with help and guidance from professionals who are specialized in tinnitus management. Attention to the patients' annoyance from tinnitus is important and clinicians should never dismiss the problem by downplaying tinnitus. Clinicians should avoid such statements as "nothing can be done." Tinnitus is a manageable condition!

Balance Disorders and Otosclerosis

Vertigo is rare in nonoperated otosclerotic ears; however, presence of vertigo may suggest inner ear malformation[92] or vestibular hair cell loss.[93] There is also evidence that otosclerosis is associated with endolymphatic hydrops and use of

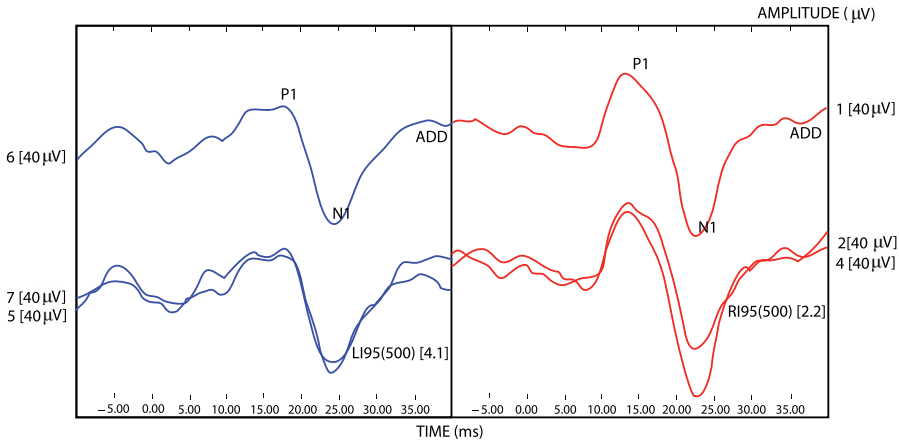

Fig. 6. Cervical vestibular evoked myogenic potential recordings in a confirmed case of superior canal dehiscence. Note the enhanced amplitude on the right side. (*Courtesy of* Ali A. Danesh, MS, PhD, Florida Atlantic University, Boca Raton, FL.)

electrocochleagraphy for better diagnosis has been reported.[94] Occasionally, patients with otosclerosis develop vertigo and dizziness following surgery.[95–97] A recent report has included secondary endolymphatic hydrops following operative interventions for otosclerosis as a pathologic finding.[98] Extensive evaluation and providing vestibular therapy is important for those who suffer from vertigo or other balance disorders. The vestibular evaluation test battery may include such procedures as videonystagmography, caloric tests, video head impulse test, and cervical and ocular vestibular evoked myogenic potential assesments.[99,100] These evaluations, particularly vestibular evoked myogenic potential studies, are helpful in the better diagnosis of such conditions as superior canal dehiscence, which potentially is associated with otosclerosis in some cases.[100–102] In general, low vestibular evoked myogenic potential thresholds and enhanced amplitudes in cases with superior canal dehiscence are expected (**Fig. 6**).[101–103] For patients with otosclerosis who develop balance disorders, vestibular therapy, balance exercises, and vestibuloadaptive therapy have been shown to be effective in managing their symptoms.[104]

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