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REVIEW

Can a Passive Mechanical Traveling Wave be Generated in Superior Canal Dehiscence Syndrome, Cochlear Implant, Otosclerosis or Atresia of Windows?

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Air conducted and bone conducted auditory stimulation both activate the inner ear by a similar mechanism since responses to air conducted stimulation can be cancelled by a bone conducted stimulus with appropriate phase and intensity. This has been taken as evidence that both air conducted and bone conducted auditory stimulations induce equal but opposite phase displacements of the two windows, the oval and round windows. These give rise to pressure differences across the basilar membrane, which initiate a passive traveling wave propagating from the base to the apex, as suggested by the studies of von Bekesy. However, in certain clinical conditions, this does not seem likely. For example, in superior semicircular canal dehiscence, the dehiscence is a third window, but nevertheless, the air conduction loss is small, while bone conducted thresholds are often better than normal. In cochlear implantation, residual low frequency acoustic hearing at the apex of the cochlea is preserved even though the implantation procedure often causes damage to the basilar membrane. In treating otosclerosis by stapes surgery (stapedectomy or stapedotomy), bone conducted thresholds prior to the surgical treatment (when there is only one mobile window) and following the surgery (two mobile windows) are similar in most cases. In addition, rare cases of congenital atresia of the round window have been reported, and in such patients, bone conduction thresholds were close to normal. These findings in the patients support the suggestion that the outer hair cells can be excited during both air conduction and bone conduction stimulation directly by intra-cochlear fluid pressures, without the mediation of a passive basilar membrane traveling wave.

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Auditory stimulation can activate the inner ear by air-conduction or by bone-conduction. This is thought to involve a passive mechanical traveling wave propagating along the basilar membrane which initiates active mechanics, leading to sound sensation.

Air Conduction

Air borne sound stimuli impinging on the tympanic membrane (air conduction stimulation) induce vibrations of the stapes footplate in the oval window. Since the inner ear is a series of sealed channels which are filled with an incompressible fluid and has only two windows (the oval window and the round window), the fluid volume displacements of the stapes footplate in one direction must be accompanied by equal fluid volume displacements (proportional to sound intensity) in opposite phase of the round window. This gives rise to a difference in fluid pressure across the basilar membrane, inducing its movement [1-3]. Since the basilar membrane at the base has a lower mass and greater stiffness than that of the basilar membrane at the apex, the basilar membrane displacements begin at the base, are larger at the base in response to higher sound frequencies, and progress toward the apex, where the magnitude of its displacements is greater for lower frequencies. Thus the basilar membrane displacements appear as a traveling wave propagating from the base toward the apex. This mechanical wave was observed and described by von Bekesy [4] in human cadavers using stroboscopic illumination, therefore requiring high sound intensities (about 130 dB SPL). These basilar

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membrane displacements are passive, i.e. the energy for them comes from the sound stimulus and they were observed in cadavers. It is generally assumed that this mechanism, seen to induce the passive basilar membrane displacements at high sound intensities, is the same as that acting at low sound intensities. In the normal cochlea, these passive basilar membrane displacements activate the outer hair cells (OHCs) by opening ion channels, leading to modulation of the electrical potential of the OHCs (i.e. mechano-electrical transduction). The electrical changes, in turn, activate the motor protein prestin in the OHCs so that they undergo mechanical length changes (shortening and lengthening) (i.e. electro-mechanical transduction) [5,6]. This is the cochlear amplifier. The synchronous mechanical active motility of a group of neighboring OHCs produces active basilar membrane displacements, which sum with the passive ones. The energy for these active displacements comes from metabolism, providing the electro-chemical gradients required for mechano-electrical and electro-mechanical transduction). The next stage involves activation of the inner hair cells which synaptically excite the auditory nerve fibers.

Bone conduction

The inner ear can also be stimulated by bone conduction, achieved by applying a bone vibrator to the skull. The induced skull vibrations reach the inner ear by indirect mechanical pathways without directly involving the tympanic membrane and the ossicular chain of the middle ear. It is generally assumed that the mechanism of bone conduction excitation of the inner ear also involves a sequence of events beginning with induction of passive basilar membrane displacements in the form of a traveling wave, followed by initiation of the active displacements. Evidence for this has come from the fact that bone conduction sound stimulation (bone vibrator applied to the skull) elicits a pitch sensation which is identical to that induced by air-conduction sound (for example, an earphone) of identical frequency. Furthermore, von Bekesy [4,7] was able to cancel the sensation of a 400 Hz bone conducted tone by means of an air-conducted stimulus at that frequency, by properly adjusting the intensity and phase of the tone. In addition, Lowy [8] and later Wever and Lawrence [9] were able to obtain cancellation of the cochlear microphonic potentials induced in experimental animals in response to tones presented by air conduction when they presented identical frequency bone conducted stimuli, with appropriate adjustment of intensity and phase. Furthermore, a human listener hears his own voice by the fusion within his inner ear of an air-conducted and a bone-conducted component. These results and findings have been taken as evidence that both types of auditory stimulation (air- and bone-conduction) induce similar patterns of passive basilar membrane traveling waves in the cochlea. During bone-conduction stimulation, three major mechanisms have been suggested for the transmission of skull vibrations to the inner ear: a translatory or inertial mode (relative motion between the bony shell of the inner ear and the ossicular chain[9-12], a compressional mode or distortional mode (distortion of the wall of the inner ear with fluid displacement in and out of the two windows [12-14] and an occlusion mode (when the external canal is blocked, skull vibrations induce changes in air pressure within the cavity formed in the external canal which acts on the tympanic membrane as in air-conduction). Each of these requires, in one way or another, mobility of the two windows on either side of the basilar membrane. Thus, in final analysis, the same cascade of events and the same sensory receptor (hair) cells are involved in both air- and bone-conduction. Further evidence that bone-conducted and air-conducted stimuli interact mechanically in the inner ear comes from recent experiments which showed that the distortion product otoacoustic emissions at 2f1-f2 (a tone at frequency 2f1-f2, not audible to the subject, generated by the active cochlear amplifier in response to the presentation of two audible tones, f1 and f2) can be elicited in human subjects by presenting f1 by air conduction (earphone) and f2 by a bone vibrator applied to several sites on the head[15].

Clinical Conditions

We will now select several clinical conditions and assess, using both air and bone conducted stimulation, whether the responses are consistent with the involvement of a passive basilar membrane traveling wave in their generation.

Superior Semicircular Canal Dehiscence Syndrome

In 1929 Tullio [16] described a condition bearing his name (Tullio phenomenon), in which loud sounds induce vestibular symptoms of vertigo and oscillopsia (oscillation of objects in the visual field); i.e. in this condition, sound stimuli can activate the vestibular end organs. It has recently been shown by Minor [17] that in many of such cases of vestibular hypersensitivity to sound, there is usually a dehiscence (an opening) of the superior semicircular canal which has introduced a low impedance pathway between the small fluid volume of the inner ear and the much larger fluid volume of the cranial cavity (CSF). In spite of this dehiscence ("third window") open to CSF, the air-conducted loss is relatively small, and there is even an improvement in bone conduction threshold, giving the impression of an air-bone gap in the absence of an actual mechanical middle ear conductive lesion [18]. The small air-conducted loss, coupled with a small bone conduction gain, are incongruent with mechanisms requiring the generation of a traveling wave along the basilar membrane, since the inner ear is no longer sealed due to the presence of a low impedance pathway (dehiscence) to the cranial cavity.

Furthermore, both air-conducted and bone conduction auditory stimuli can activate the vestibular end organs and this is used clinically to elicit the vestibular evoked myogenic potential (manifestation of a vestibular reflex pathway to the sterno-cleido-mastoid muscle [19]. Thus, here too air-conducted and bone conduction sound stimuli can activate the vestibular hair cells in the inner ear, even though there is no basilar membrane (and hence no traveling wave) in the vestibular end organs [20].

Cochlear Implant

An additional clinical condition for consideration here is the use of cochlear implantation in cases of severe to profound sensori-neural hearing loss. In the past, patients with residual low frequency acoustic hearing were not considered to be good candidates for implantation since it was thought that the implant insertion procedure would cause damage at the basal turn and compromise the residual low frequency hearing from the apical turn and also because the implant electrode itself takes up a relatively large volume of scala tympani, reducing the effective fluid volume. Recently, however, it has been shown that in many cases the insertion of the implant does not compromise the initial residual low frequency hearing, i.e. the residual low frequency hearing is preserved following implantation [21-25] and the use of cochlear implantation is being extended even to cases with residual hearing at low frequencies. Thus the implant provides higher frequency hearing by electrical stimulation at the basal turn, while the residual low frequency hearing is acoustically (perhaps with assistance from a hearing aid) elicited at the apical turn [26]. On the other hand, Tien and Linthicum [27] examined post-mortem the temporal bones of 11 implantees and found damage to the osseous spiral lamina or to the basilar membrane in eight of them. Insertion trauma to the vestibular end organs has also been reported [27,28]. Experimental studies (cochlear implantation in cadaver temporal bones) have also shown that the implant insertion procedure itself often causes damage to the basilar membrane at the basal turn (e.g. elevation and rupture of the basilar membrane, fracture of the osseous spiral lamina and the presence of the electrode in scala vestibuli) [29-32]. In such cases (preservation of the residual low frequency hearing following cochlear implantation), the basal turn basilar membrane is often damaged, but nevertheless the apical hair cells can still be acoustically excited [33].

Otosclerosis

A third clinical condition of interest is the hearing loss accompanying otosclerosis, in which there is fixation

of the stapes footplate in the oval window, causing its immobilization, i.e. in such cases there is only one mobile window, the round window. The condition is treated by stapedectomy (replacement of the immobile stapes footplate) or stapedotomy in which a perforation is made in the stapes footplate, and a prosthesis is introduced between the long process of the incus and the perforation in the footplate. In a large series of 2,525 patients operated on by the same surgeon using the same technique [34], it was found that the mean pre-operative air- and bone-conduction thresholds at four frequencies (0.5, 1, 2 and 4kHz) were 51.4 dB HL and 25.8 dB HL respectively. The mean post-operative air- and bone-conduction thresholds were 27.2 dB HL for air and 25.5 dB HL for bone-conduction. In other words, in the presence of a relatively immobile stapes footplate pre-operatively, the mean bone-conduction threshold was 25.8 dB HL. Post-operatively, with two mobile windows, mean post-operative threshold was 25.5 dB HL, i.e. there was no improvement in bone-conduction threshold; the only improvement seen was that for air conduction (and hence reduction of the air-bone gap). Thus, even though it has been assumed that a passive traveling wave is also initiated during bone conduction stimulation, such a wave requires two mobile windows for inner ear fluid volume displacements (whether to allow for the translatory - inertial mode, for the compressional mode or the occlusion mode of bone-conduction) and this is not the case pre-operatively in otosclerosis. In fact, surgical mobilization of the footplate was not accompanied post-operatively by an improvement in bone-conduction threshold. In addition, before the introduction of stapes surgery for alleviation of otosclerosis, fenestration of the horizontal semicircular canal was used to reduce this hearing loss [35]. While the stapes surgery procedures are an attempt to reconstruct the normal mechanism of cochlear activation, the fenestration procedure provided an alternative mobile window on the scala vestibuli side of the basilar membrane (replacing the immobile stapes footplate). Together with the mobile round window, pressure differences could thus be initiated across the basilar membrane, with improvement in hearing. Interestingly, here too, the immediate post-fenestration bone conduction threshold (with two mobile windows) was usually the same as that before the fenestration (with only one mobile window) [9].

Window Atresia

Finally, rare cases of apparently congenital atresia of the round window have been reported [36-38] and even of the oval window [39]. Nevertheless, bone conduction thresholds were close to normal. This of course is incompatible with the assumption that bone conduction stimulation also involves a passive traveling wave propagating along the basilar membrane, since a prerequisite for such a passive traveling wave is the presence of two mobile windows. In fact, Borrmann and Arnold [38], based on their review of 13 cases of non-syndromic round window atresia, state: “the hearing test results of the effect of round window atresia-cannot be thoroughly explained by current theories of sound transmission”. In addition, Linder et al [37] state “Analysis of these two cases and the literature review present puzzling findings, which contradict some of the current concepts of hearing function. Implying that there is no extra pressure release mechanism of the cochlea in these patients, we conclude that the cochlea is also sensitive to an alternative way of stimulation”.

Alternative Mode of Activation

Such an alternative mode of bone conducted stimulation has been demonstrated in several studies on human subjects and experimental animals. These studies have shown that vibratory stimuli delivered to several sites on the head can elicit auditory responses in the absence of actual skull vibrations. It was concluded that a major route for transmission of skull vibrations to the inner ear is by a fluid pathway from skull contents (CSF), by means of fluid communications between the CSF and the inner ear fluids, to the inner ear [15,40,41], exciting the OHCs directly. In several of these studies, other modes of stimulation based on a passive traveling wave were experimentally excluded [40,42].

Furthermore, with respect to air-conduction stimulation, additional analyses have been conducted in which holes, about the same size as the round window, were introduced at several sites in the wall of the inner ear. In these conditions, the inner ear was no longer sealed. Nevertheless, auditory thresholds to air- and bone conducted stimulation were not elevated [43-45]. Based on these findings, and others, one can suggest that low intensity auditory stimulation, whether air-conducted (leading to stapes footplate vibrations) or bone-conducted (fluid pressures from the CSF to inner ear fluids), induces alternating condensation/rarefaction fluid pressures at the frequency of the sound stimulus in inner ear fluids. These fluid pressures can directly activate the stretch sensitive ion channels in the lateral walls of the OHCs [46-48]. In this way, a passive traveling wave along the basilar membrane is not required to activate the OHCs.

Conclusion

Such a mechanism (direct fluid pressure activation of the OHCs) can excite the inner ear during both air- and bone-conducted stimulations, even in the presence of clinical conditions in which the generation of a passive basilar membrane traveling wave is likely compromised, such as in superior canal dehiscence syndrome, cochlear implant, otosclerosis or atresia of windows.

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