

HEARING LOSS

DONALD HENDERSON, PhD
Hearing Research Laboratory
Department of Communicative Disorders
and Sciences
State University of New York at Buffalo
Buffalo, New York

ROGER P. HAMERNIK, PhD
Auditory Research Laboratory
State University of New York at Plattsburgh
Plattsburgh, New York

High-level noise exposures present special challenges to the auditory system. The mammalian ear has evolved so that it can detect sounds with displacements in the sub-angstrom range while, at its upper limits, it can faithfully encode sounds 10^6 units above threshold, or over a dynamic range of 120 dB SPL. However, with repeated exposure to sounds in the ear's upper range (above 85 dB SPL), the auditory periphery, or cochlea, progressively deteriorates.¹⁴ The damage caused by noise is pervasive and affects virtually all of the cellular subsystems of the inner ear (sensory cells, nerve endings, vascular supply). Sounds such as gunfire and certain industrial impacts—peak levels greater than 125 dB—are especially hazardous to the cochlea because they cause direct mechanical damage.¹² In short, the ear was not designed for exposures to the high-level noises found in contemporary environments.

This chapter reviews the biologic response of the ear to excessive noise and relates the cochlear pathology to changes in hearing function. To appreciate the ear's response to noise, it is necessary to understand the normal structure and function of the peripheral auditory system.

The auditory periphery (Fig. 1) can be divided into three systems: the external ear, consisting of the pinna and the external auditory meatus (EAM); the middle ear (ME), containing the three ossicles (malleus, incus, stapes), the two muscles (stapedius and tensor tympani),

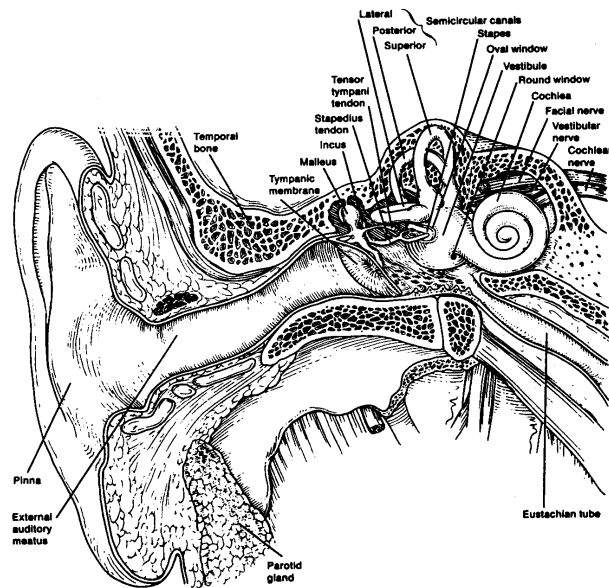


FIGURE 1. The major components of the right temporal bone and the external, middle, and inner ear.

sensory/neural elements. Each plays a role in the response of the auditory system to noise.

THE EXTERNAL EAR

The geometry of the EAM is that of a tube, closed at one end by the tympanic membrane (TM). Tubes open at one end have acoustic resonant properties that are described as follows: resonant frequency (f) = speed of sound / 4 \times length of EAM. Because the average human EAM is approximately 25 mm long, the resonant frequency of the average human ear is around 3,200 Hz. Figure 2A illustrates how EAM resonance can “amplify” sound as it passes from the entrance of the EAM to the tympanic membrane. Depending on the direction and frequency of the sound source, the sound pressure amplification can be as great as 20 dB in the midfrequency range. Thus, the resonant characteristics of the EAM help determine the acoustic energy delivered to the cochlea. For example, industrial noise typically has a broad spectrum; however, as it travels through the EAM, acoustic energy in the midfrequency range resonates or is amplified, creating a band pass noise centered at 3200 Hz at the TM (Figs. 2B and 2C).

There are two clinical implications of the acoustic resonance characteristics of the EAM. First, the acoustics of the EAM are primarily responsible for the appearance of the “4 Hz notch” that is typically seen in audiograms of patients with noise-induced

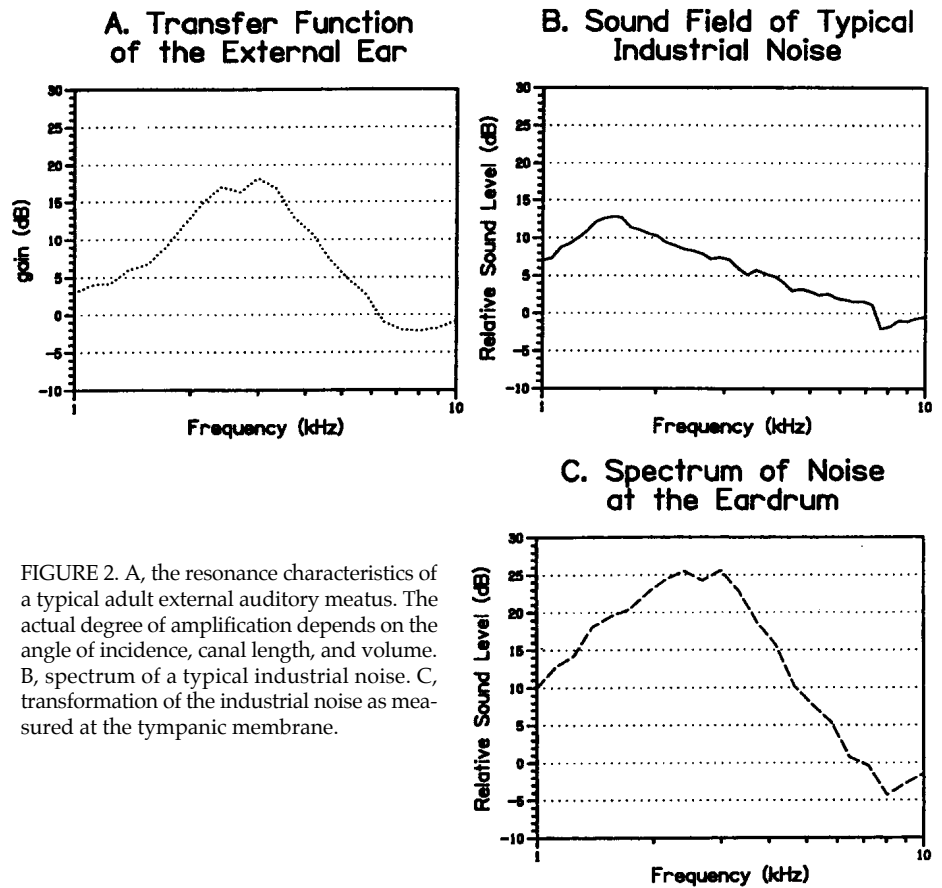


FIGURE 2. A, the resonance characteristics of a typical adult external auditory meatus. The actual degree of amplification depends on the angle of incidence, canal length, and volume. B, spectrum of a typical industrial noise. C, transformation of the industrial noise as measured at the tympanic membrane.

observed at ± 1 octave above the center frequency of the noise. Furthermore, studies of basilar membrane vibration show that the point of maximum displacement occurs half an octave above the frequency of stimulation. Since most industrial noises are relatively broad band, the transformation of the EAM creates a bandpass noise centered at 3 kHz; thus, the familiar 4 kHz notch is the result of the half-octave shift of the fundamental EAM resonance (see Fig. 2C) rather than a reflection of any inherent weakness in the 4 kHz region of the cochlea.

Second, the large variability in peoples' responses to noise may be partially related to the variation in acoustic transfer characteristics of the EAM. Hellstrom et al.¹¹ measured the length and volume in many subjects with normal hearing and NIHL and correlated them with quiet threshold and temporary threshold shifts (TTS) following exposure to noise. They reported that the transformation from the sound field to the tympanic membrane depends on the length and volume of the canal. The actual resonant frequency varies markedly across normal subjects. Subjects in the study by Hellstrom et al.¹¹ were categorized on the basis of whether their sound transfer functions (STF) showed greater gain at 2 kHz or 4 kHz. Specifically, subjects with greater (8 dB or more) gain at 4 kHz than at 2 kHz were classified as "high STF" subjects. Subjects with gain at 4 kHz that was no more than 3 dB greater than the gain at 2 kHz were classified as "low STF." Not only did the differences in STF classification correspond to differences in quiet threshold of about 6-8 dB between 2.5-5 kHz (the region of maximal resonance), but the STF classification correlated with the location and magnitude of hearing loss following exposure to either a 2- or 4-kHz narrow band noise (NBN) at 97 SPL dB for 10 minutes. The 5- to 8-dB differences between subjects shown in Figure 3 will produce an even greater hearing loss following a noise exposure because, for each dB of noise above the threshold for hearing loss, TTS or permanent threshold shift (PTS) increases at a rate of 1.7 dB/dB increase in noise level.¹⁴ The results reported by Hellstrom et al.¹¹ agree with earlier work by Ciazzo and Tonndorf,⁴ who showed that an artificially controlled EAM length had a direct effect on the level and frequency location of TTS. The general implication of the acoustic studies of the EAM is that variations in the normal geometry of the EAM can have a significant impact on the NIHL an individual ear develops.

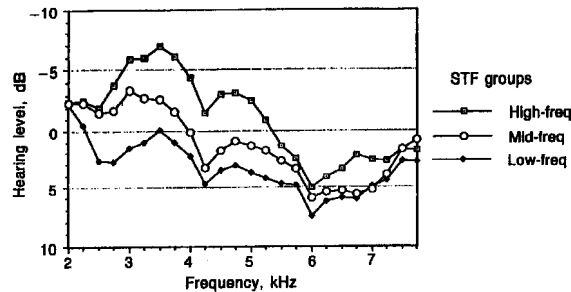
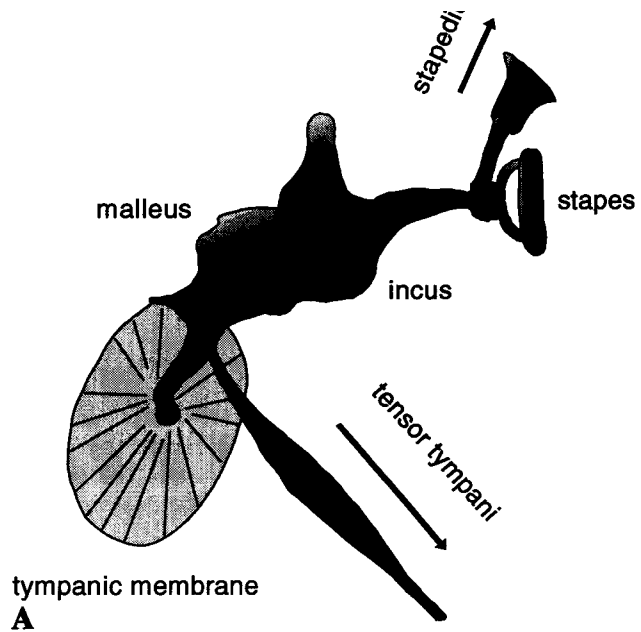


FIGURE 3. The relation between the sound transfer function and hearing levels of subjects following exposure to narrow band noise. The STF depends on volume and length of EAM. Hearing levels are shown relative to preexposure thresholds.

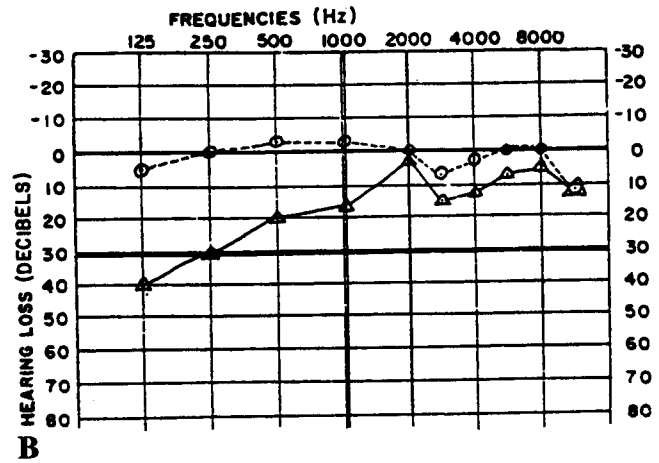
THE MIDDLE EAR

Figure 1 shows that the middle ear system is bound laterally by the tympanic membrane, which in turn attaches to the malleus, incus, and stapes. The stapes footplate is attached to the oval window by the annular ligament. The primary function of the ME is to serve as an impedance matching transformer to partially compensate for the approximately 40 dB transmission loss that occurs when airborne sound is introduced to the fluid-filled cochlea. The system normally behaves like a low pass filter with an approximate cutoff of 1,200 Hz. The attenuation at high frequencies (above 4 kHz) partially explains the poorer



A

THE MIDDLE EAR



B

FIGURE 4. A, the ossicles and muscles from a dorsal perspective. B, the attenuation provided by the AR in subjects who had voluntary control of middle ear muscle contraction. (Adapted from Reger SN: Effect of middle ear muscle action on certain psychophysical measurements. *Ann Otol Rhinol Laryngol* 69:1179-1198.)

audiometric performance at high frequencies. Variations exist in the size of the ossicles and middle ear space, but there are virtually no direct data regarding how individual variations in middle ear structure/mechanics contribute to susceptibility to NIHL. However, a clue that the ME mechanics may be important comes from a comparison of ME impedances in humans versus chinchillas. At 220 Hz and 660 Hz, the human impedance is two to three times²⁶ greater than the chinchilla impedance, and chinchillas are 10-15 dB more susceptible to NIHL.¹⁴

The acoustic reflex (AR) can be an important variable in the ear's response to noise exposure. The stapedius muscle is attached to the head of the stapes and is controlled by the facial (VII) cranial nerve; the tensor tympani muscle is attached to the malleus near the tympanic membrane and is controlled by the trigeminal (V) cranial nerve. When both muscles are activated, the tensor pulls the tympanic membrane toward the middle ear, thereby stiffening the TM, and the stapedius pulls the stapes almost perpendicular to the normal axis of movement in the oval window (Fig. 4A). Even though the planes of contraction are antagonistic, the action of the two muscles is complementary: their joint contraction decreases the transmission of sound through the ME by increasing the stiffness of the ME system. The attenuating action, however, is limited to sounds below 2,000 Hz (Fig. 4B).

A number of experiments show the contribution of the stapedius to protection from noise exposures that cause TTS. Zakrisson et al.²⁷ showed that when patients with Bell's palsy (unilateral paralysis of the facial nerve) were exposed to moderate levels of noise, the ear on the paralyzed side developed substantially more TTS than the ear on the normal side. As patients recovered, US became more symmetrical. The investigators extended their research to study the acoustic reflex action on PTS. The facial nerve was sectioned in a group of rabbits, and the subjects were given a noise that would normally produce PTS. Subjects without the stapedial reflex had substantially more low-frequency PTS and, interestingly, slightly less high frequency PTS (Fig. 5).

By contrasting the permanent hearing losses in workers with "efficient" and "sluggish" acoustic reflexes, Colletti et al.⁵ have provided insights into the importance of the AR effects on exposed populations in industrial settings. Subjects were assessed and categorized on the basis of reflex threshold, strength of contraction, resistance to adaptation, and latency. Workers with highly efficient reflexes (i.e., low threshold, strong contraction, free of adaptation) developed substantially less PTS over years of work experience than workers with sluggish reflexes. Although the study is interesting, it had few subjects. Also, because of the

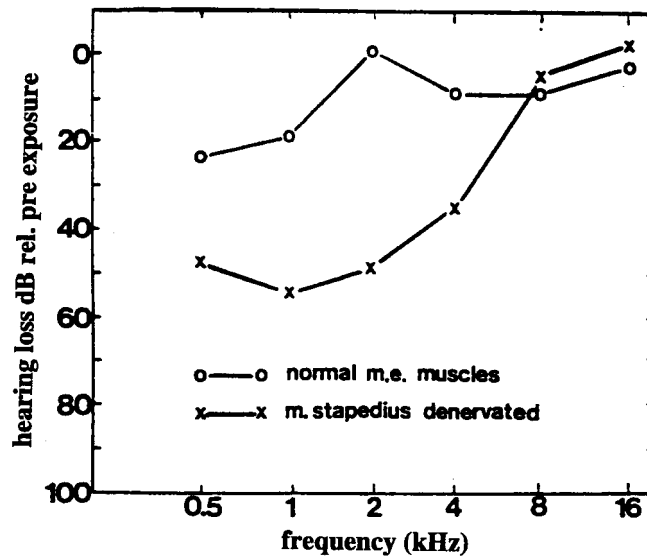


FIGURE 5. The protection provided by acoustic reflex. With stapedius sectioned, subjects developed 20-40 dB more permanent threshold shift. (Adapted from Zakrisson JE, Borg E, Liden G, Nilsson R: Stapedius reflex in industrial impact noise and fatiguability and role for temporary threshold shifts (TTS). Scand Audiol Suppl 12:326, 1980.)

the ear from exposure to impulse or impact noise.

The EAM and ME are responsible for the transmission of acoustic energy from air to the cochlea. The combined action of these systems transforms the input in a relatively predictable manner that is governed by laws of acoustics and mechanics. As energy enters the cochlea, the process becomes more complicated and involves a number of biologic processes.

THE INNER EAR

Sensory Epithelium

A cross-sectional schematic of the cochlea shown in Figure 6A illustrates the location of the organ of Corti (the basilar membrane and sensory/supporting cell complex) and stria vascularis and the overall arrangement of the fluid filled channels (scalae) of the membranous labyrinth. Figure 6B is a 1μ thick stained section from a chinchilla cochlea showing the organ of Corti at high magnification. The inner hair cells (IHCs) and the outer hair cells (OHCs) are responsible for the initial transduction of the mechanical stimulus (the traveling wave) into a receptor potential, which, in turn, activates the eighth nerve's neurons. The three rows of OHCs and single row of IHCs parallel each other along the entire basal to apical extent of the basilar membrane. Following a traumatic noise exposure, the metabolism of these cells is changed, the cellular architecture is damaged, and normal function is altered.

When sound is transmitted to the inner ear via the movement of the stapes footplate, it initiates a traveling wave that moves from the oval window to some point of maximum vibration along the basilar membrane (BM). The actual vibration patterns on the basilar membrane are determined by the physical characteristics of stiffness and mass (Fig. 7A). The mechanical (impedance) characteristics of the organ of Corti change systematically along its length. For instance, the base is 100-200 times stiffer than the apex, and the cross-section

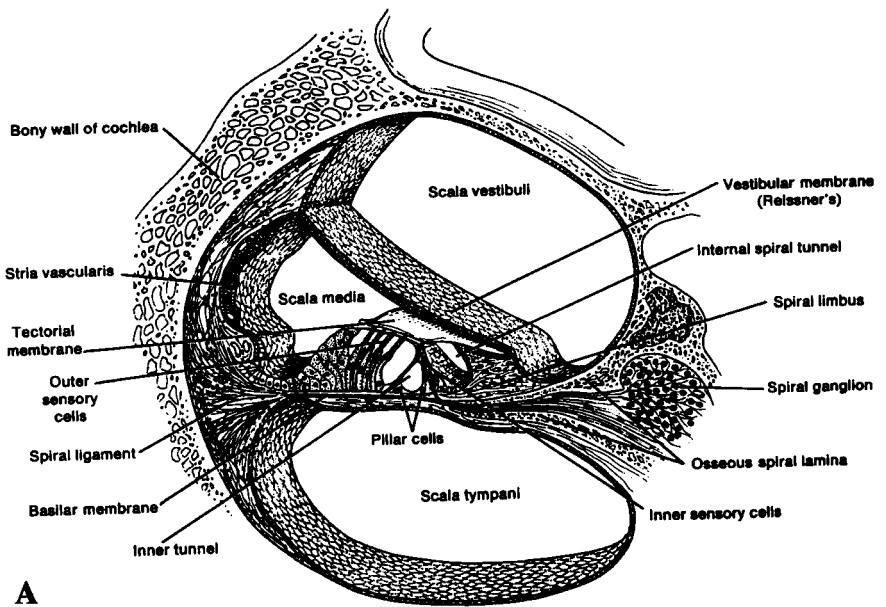


FIGURE 6A. The cochlea, including the organ of Corti (basilar membrane and sensory/supporting cell complex) and the overall arrangement of the cochlear elements. (Continued on next page)

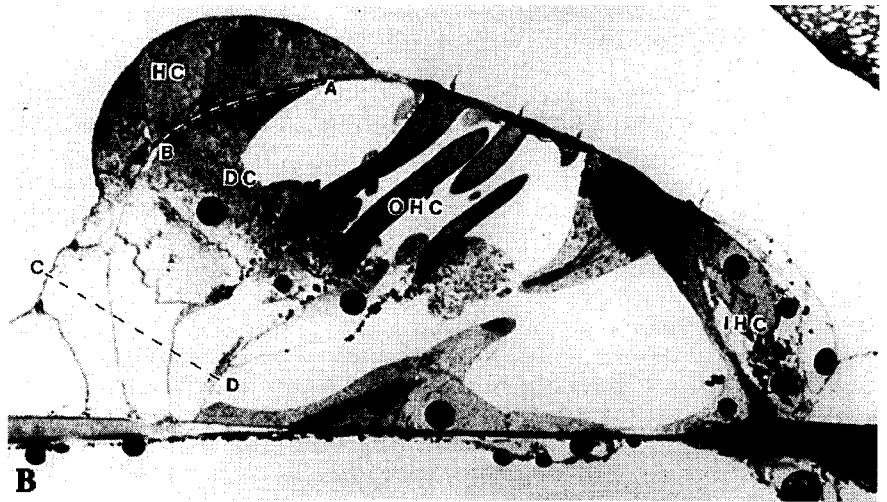


FIGURE 6B. A section 1μ thick through the organ of Corti (chinchilla). The tectorial membrane has been retracted to the top right of the micrograph. OHC, outer hair cells; IHC, inner hair cells; DC, Deiter cells; HC, Hensen cells. A-B and C-D indicate zones of the organ of Corti that are particularly susceptible to fracture or tearing following severe acoustic trauma.

or unit volume of the organ of Corti increases by a factor of about 203 from the base to apex. This impedance gradient along the length of the basilar membrane leads to a tonotopic encoding of the frequency of the incoming sound. That is, the peak of the traveling wave is

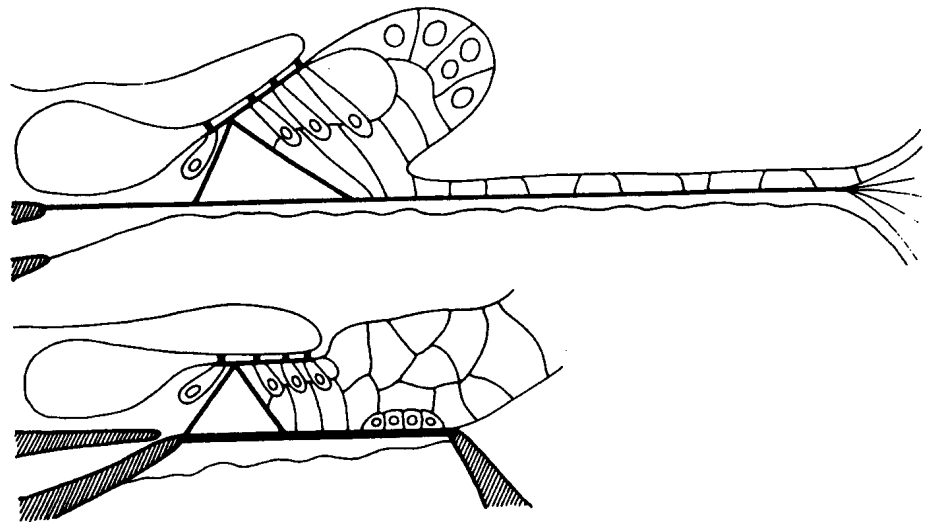


FIGURE 7. Cross-section of the basilar membrane at the base (top) and apical (bottom) region of the cochlea. (Adapted from Spoenclin HH: Anatomical changes following noise exposures. In Henderson D, Hamernik RP, Dosanjh DS, Mills JH (eds): Effects of Noise on Hearing. New York, Raven Press, 1976, pp 69-90.)

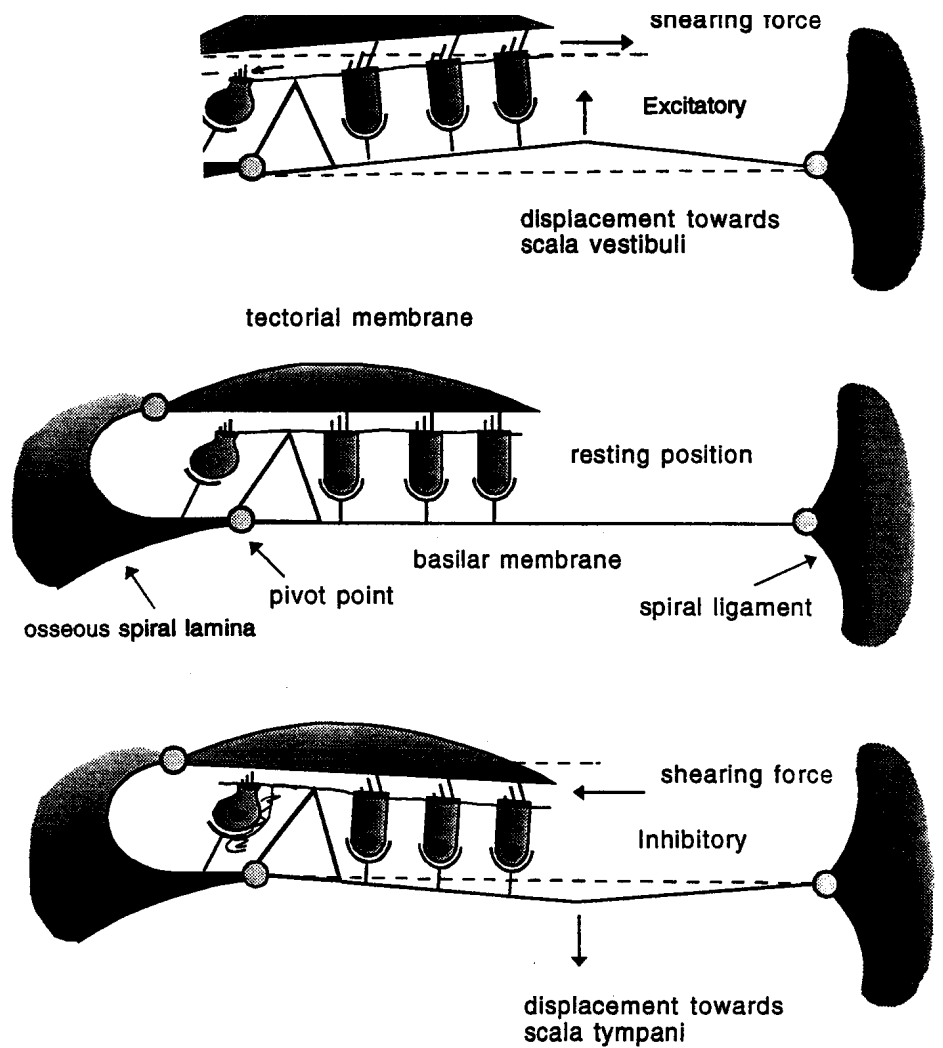


FIGURE 8. The organ of Corti, including the mechanism for generating shearing force at the stereocilia. Note that OHCs are connected to tectorial membrane and that there are possible differences in vertical displacement between OHCs and IHCs.

distributed such that high frequency stimuli cause maximal disturbance near the oval window, and as the frequency is lowered, the peak of basilar membrane motion systematically shifts apically.

The traveling wave induces shearing between the tectorial membrane, reticular lamina, and organ of Corti, which causes the final mechanical motion of significance: movement of the stereocilia. This mechanical process (Fig. 8) initiates a series of electrochemical events within the hair cells. Displacement of the basilar membrane toward scala vestibuli and bending of the stereocilia toward the spiral ligament is excitatory, leading to increased release of chemical transmitter at the basal surface of the hair cell. Conversely, displacement of the BM

leading to decreased release of neurotransmitter. The process of noise-induced hearing loss involves not only the sensory cells, but also supporting cells, nerve fibers, and vascular supply.

Figure 9, a scanning electron micrograph (SEM) of the surface of the organ of Corti shows, at various levels of magnification, the regular arrangement of the inner and outer hair cells and the orderly arrangement of the cilia on each of these classes of sensory cell. In contrast, the SEM in Figure 10A shows a segment of the organ of Corti from a noise-damaged cochlea, collected 10 days following exposure. Clearly visible are a loss of sensory cells, disturbance of the cilia, and the presence of considerable cellular debris. Sensory cell loss is often quantified in the form of a cochleogram,⁸ which shows the percent of missing IHCs and OHCs as a function of location (and hence frequency) along the basilar membrane. The cochleogram from this animal (Fig. 10B) indicates loss of both inner and outer hair cells

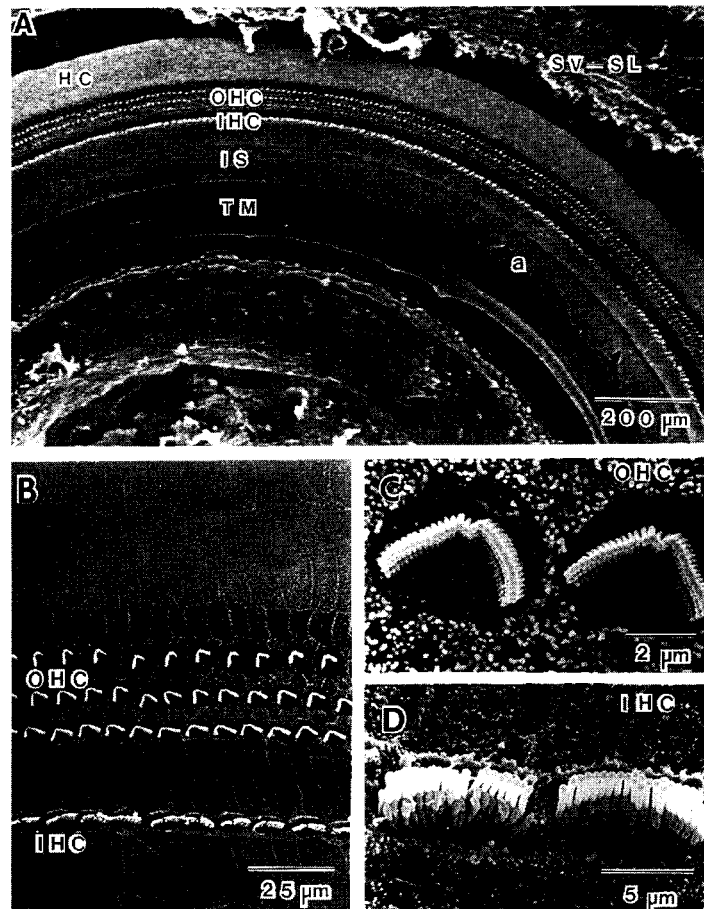


FIGURE 9. A, a low magnification view of about a quarter of a turn of the normal chinchilla organ of Corti showing the uniform appearance of the surface of the sensory epithelium. B, a higher magnification view of a section of the same specimen. The complex mosaic of cellular junctions and the individual cells are clearly visible. C and D, the orderly arrangement of the cilia on the outer and inner hair cells, respectively. SV, stria vascularis; SL, spiral ligament; HC, Hensen cells; OHC, outer hair cells; IHC, inner hair cells; IS, inner sulcus; TM, tectorial membrane; a, artifact.

to approximately 1-2 kHz.

Following a traumatic noise exposure, damage or loss of OHCs is invariably greater than that of the IHCs (Fig. 10). Several factors contribute to the greater vulnerability of OHCs to damage. First, OHCs experience a direct shearing force at their stereocilia, whereas the IHC stereocilia are stimulated by viscous drag (see Fig. 8). Secondly, the OHCs have most of their long axis "unprotected" from mechanical stress, whereas the IHCs are "supported" on all surfaces with supporting cells (see Fig. 6). Finally, the OHCs are closer to the point of maximal basilar membrane traveling wave displacement than are IHCs.

Early noise-induced changes in the sensory cell body involve essentially all the organelles (e.g., the mitochondria, the subsurface cisternae, and the smooth endoplasmic reticulum) and the nucleus, along with an increase in the number of lysosomes and other membrane bound structures such as Hensen bodies. Membrane alterations may impair the cell's ability to regulate its ionic composition, resulting in swelling and an evagination or herniation of cell contents at structurally weakened locations. Such herniation is frequently seen at various locations along the reticular lamina, a region characterized by a mosaic of tight cell junctions that seals off the subreticular space from spaces of Corti. Figure 11 shows examples of such herniation in the area of the IHCs of a sheep cochlea exposed to a high-level impulse noise (Fig. 11A) and from a noise-damaged section of a chinchilla organ of Corti (Fig. 11B) in which cellular material extrudes through the line of tight cell junctions between the Hensen cells and the outer most row of Deiter cells. In this region, the tight cell junctions have failed. Such changes ultimately lead to sensory cell loss in restricted regions. Even if the reticular lamina remains intact, the loss of sensory cells can itself be the cause of further progressive deterioration of the sensory epithelium by virtue of the intermixing of cochlear fluids.² In the course of sensory cell degeneration, holes frequently appear in the matrix of tight cell junctions of the reticular lamina, which allow the subreticular fluids to intermix with the fluid in the spaces of Corti (Fig. 12). The loss of sensory cells resulting from the intermixing of fluids can proceed over a prolonged time following the initial noise trauma. A correlate of this may be the growth of threshold shift often measured after a severe noise exposure.⁹ That is, threshold shift several hours after exposure can be much larger than that immediately



FIGURE 10A. Scanning electron microscopy of the sensory epithelium from a noise-damaged cochlea. The specimen was collected 10 days following exposure. Most of the outer hair cells in the first and second rows have been destroyed, and the remaining ones have severely disrupted cilia. The inner hair cells are all present, but there are numerous cilia disruptions; considerable cellular debris is present. O, outer hair cells; D, cellular debris. (Continued on next page.)

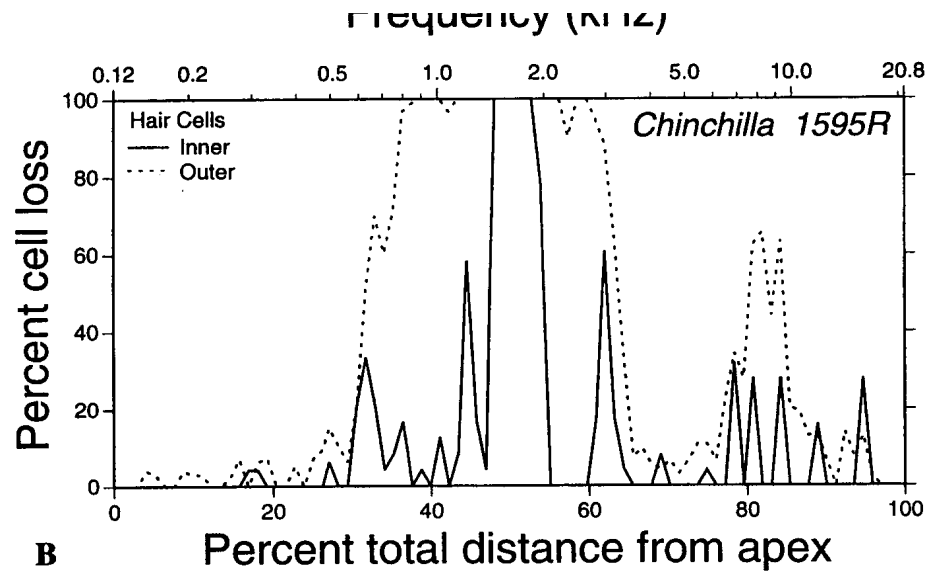


FIGURE 10B. A cochleogram from a noise-damaged cochlea. The cochleogram quantifies the extent of the sensory cell damage throughout the cochlea.

following the exposure. At the location of missing sensory cells, a scar eventually is created from the remaining supporting cells. Depending on the severity of the lesion, the scar may involve only Deiter cells in the case of OHC losses, or the Claudius and inner sulcus cells in

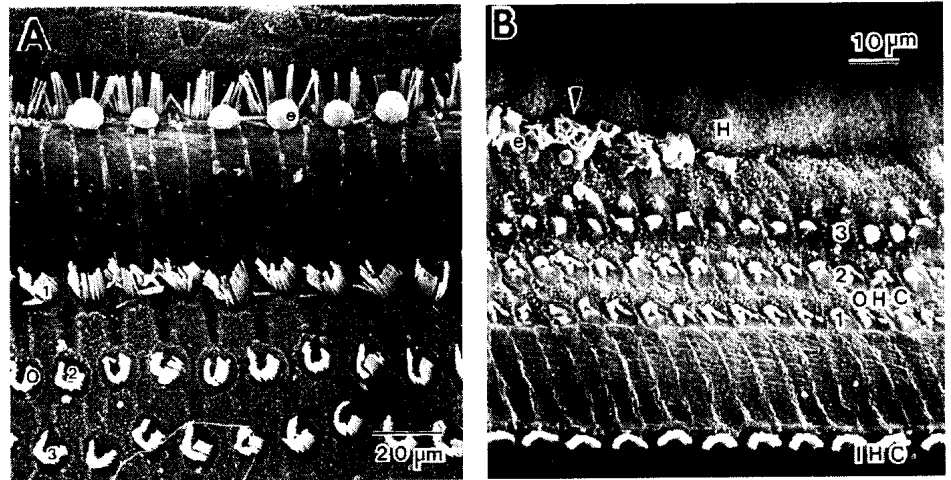


FIGURE 11. A, scanning electron microscopy from a noise-exposed sheep cochlea, showing disruption of inner and outer hair cell cilia and the extrusion (e) of cellular material through the reticular lamina. B, a failure of the tight cell junctions (arrow head) between the Hensen cells (H) and the last row of Deiter cells resulting in the extrusion (e) of cellular material. The inner hair cells are in good condition, but the outer hair cells show signs of damage. The specimen was collected from a noise-exposed chinchilla immediately after exposure.

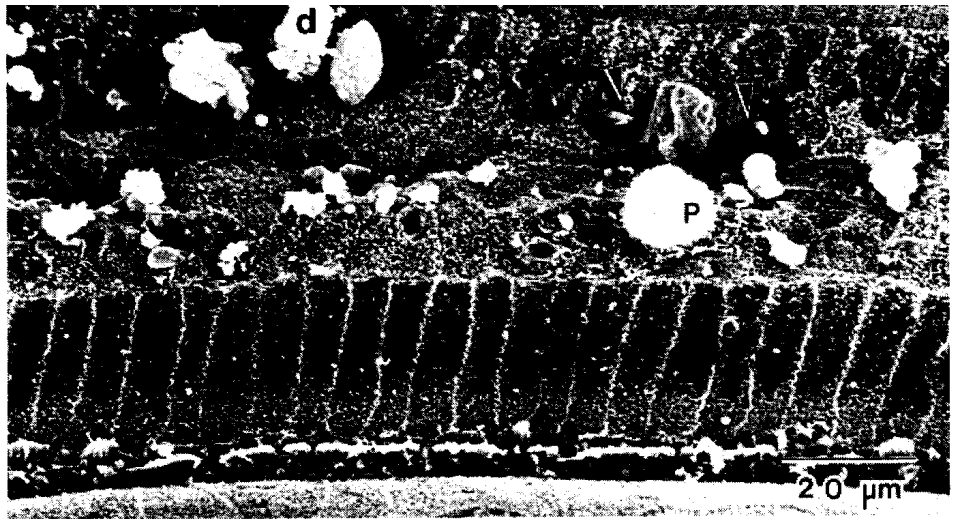


FIGURE 12. Scanning electron microscopy of the surface of a noise-exposed chinchilla organ of Corti, 1 day following exposure, showing a complete loss of outer hair cells. Several holes in the reticular lamina are visible (arrow heads), debris (d) is plentiful, and phagocytic cell types (P) can be found.

the case of near total loss of the organ of Corti.¹⁰ Examples of scarred portions of the organ of Corti are shown in Figure 13. The organ of Corti that is severely damaged to the extent that supporting cells are also lost is eventually covered with a thin undifferentiated squamous epithelium (Fig. 12B) but over a considerable extent of the basilar membrane. In some cases, a relatively mild pathology affects supporting structures such as the inner pillar cells, but few sensory cells may be implicated in altered function at locations distant to the locus of damage.¹⁸

As noise levels increase, a shift occurs from a primarily metabolic route of hair cell loss to a primarily mechanical route of loss. Acute high-level acoustic trauma results in widespread fracture of the tight cell junctions of the organ of Corti and in the separation of entire segments of cell populations from their basilar membrane attachments. The stained micron-thick cross-section of the organ of Corti shown in Figure 6 illustrates, with the use of dashed lines, the cellular connections that are most susceptible to fracture and dislocation. Examples of immediate postexposure pathologies involving these connections are shown in Figure 14. Since the OHCs and their efferent innervation are instrumental in the modulation of basilar membrane mechanics, such widespread damage affects not only the sensory/neural transduction process but the micromechanics of the basilar-tectorial membrane system, which, in turn, can affect the regions of the cochlea where there is little or no damage.

Stereocilia

Stereocilia, delicate hair-like structures arranged in staggered rows on the apical surface of the sensory cell, are bounded by a continuation of the plasma membrane of the cell. The arrangement of the cilia on the IHCs and OHCs differs, with IHCs having a more or less linear arrangement of cilia and OHCs having a "V"- or "W"-like configuration (see Fig. 9). The height of the cilia is graded by location, with the shortest cilia in the row facing the modiolus. The tallest of the OHC cilia are in contact with the tectorial membrane, while the cilia of the IHCs may not have such attachments. The morphology of the cilia and their interconnections is complex.²⁰ The cilia contain a lengthwise arrangement of actin filaments with transverse interconnections, resulting in a stiff, hair-like structure. Adjacent cilia are interconnected by tip links thought to be instrumental in the gating process during trans-

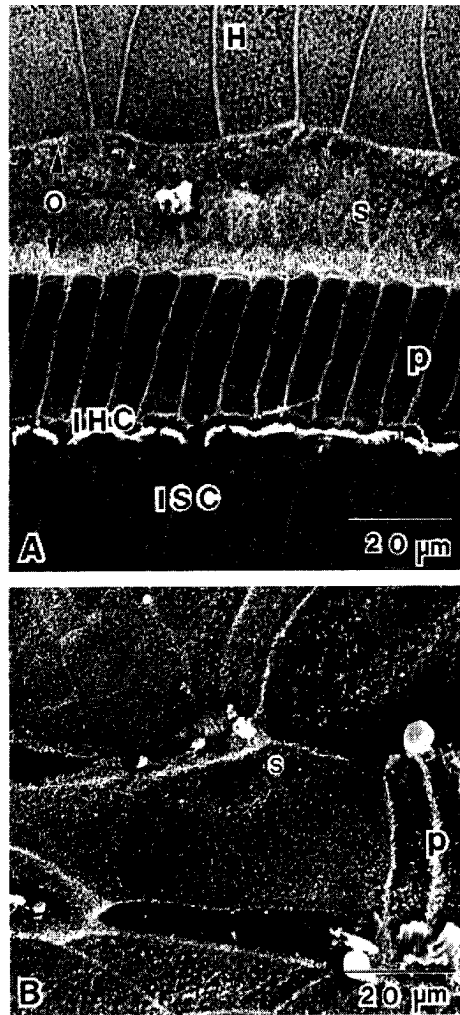


FIGURE 13. A, scanning electron microscopy of a region of scars (S) formed in an area of total outer hair cell loss (< 0 by the Deiter cells. Note the intact pillar cells (p), inner hair cells (IHC), and Hensen cells (H) B, a region of the organ of Corti where the sensory cells, border cells, Deiter cells, pillar cells, and Hensen cells have been destroyed and replaced by a scar (S) formed by the Claudius and inner sulcus cells (ISC).

duction and by intracilia glycocalyx specializations that interconnect the entire set of cilia on each sensory cell.

Some of these subtleties of cilia morphology are visible in the transmission electron micrographs (TEM). Each cilium is securely anchored into the cuticular plate with an actin rootlet (Fig. 15); the cilia of the OHCs also show delicate filamentous connections to the tectorial membrane. These displacement-detecting structures, with their very delicate interconnections, are among the first elements of the transduction sequence to be damaged by excessive displacement due to sound stimulation. OHCs are subject to damage from excessive shear between the tectorial membrane and the cuticular plate via the ciliary connections. This is one reason why OHCs are more susceptible to noise damage than IHCs. The SEMs and TEMs in Figures 16 and 17 are examples of cilia damaged by impulse noise exposure. The various panels show wrinkles and blebs in the encompassing membrane; fused cilia; disordered or splayed cilia indicating an obvious disruption of cilia interconnections; floppy cilia as a result of changes in the actin core; and cilia fractured at the root. These and other cilia changes have been implicated in both temporary and permanent changes in hear-

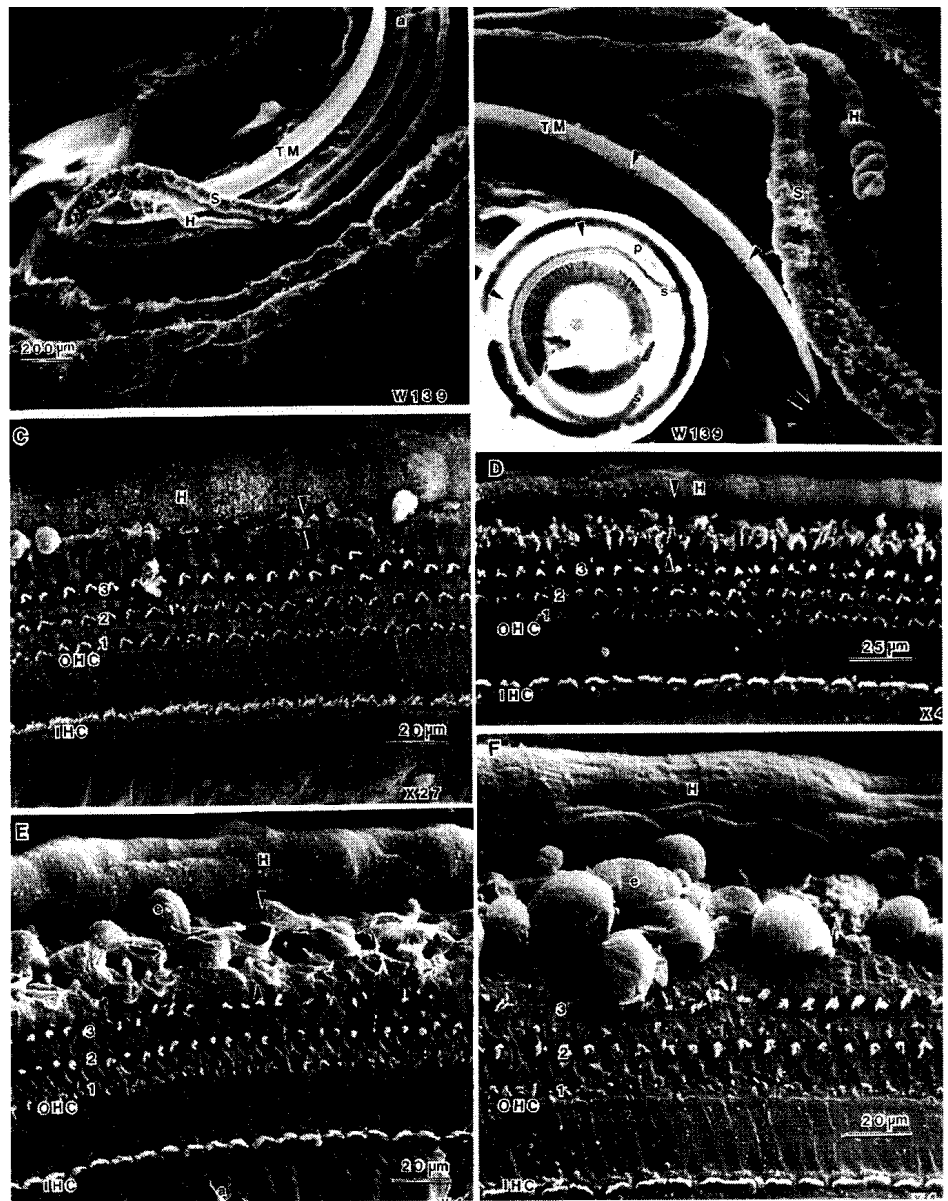


FIGURE 14. A, low magnification SEM of severe mechanical disruption of the organ of Corti showing the tearing of the strip of Hensen cells (H) and a separate strip of the sensory epithelium (S) from the basilar membrane. B, a higher resolution SEM of the specimen shown in A from a different perspective. The inset shows the appearance of the same region of the cochlea during the initial stages of dissection. The strip of Hensen cells is separated over a much longer segment (inset arrow heads) than is the strip of the sensory cells (S). C-F, examples of the open fracture of the tight cell junctions between the Hensen cells (H) and the last row of Deiter cells (arrow heads). Note the extrusion (e) of cell bodies and contents and the clearly disturbed appearance of the outer hair cells (OHC).

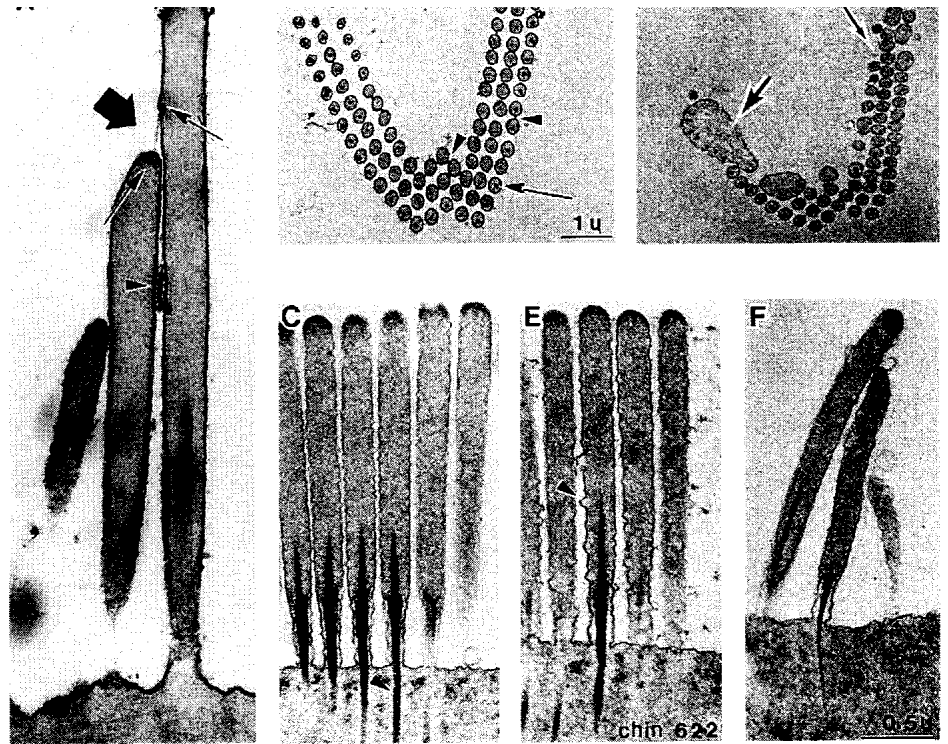


FIGURE 15. A, transmission electron micrograph of ruthenium red stained OHC cilia showing the cilia tip connection or filament (bold arrow) and the intracellular densities (arrows) typically associated with the filament connections at the cilia surface. Also visible is the intercalia glycoalkyx specialization (arrow head). B, a transverse section through an OHC cilia bundle showing the core of the rootlet (arrow) and intercalia connections (arrow heads). C, section through normal OHC cilia showing the actin rootlet and its insertion into the cuticular plate (arrow head). D, section through a noise-damaged OHC cilia bundle showing fused cilia; in this clump (large arrow) the rootlets of six cilia can be seen. The smaller arrow points out location of missing cilia. E, following noise exposure, the cell membrane encompassing the cilia is often wrinkled and blebed (arrow head). F, the structural rigidity of the cilia is often lost following a noise exposure, and the cilia tend to become floppy.

ing. The cilia appear to be the weakest link in the mechanical transduction process. There is evidence that some of these ciliary changes are reversible and may represent one of the underlying changes associated with recovery from temporary threshold shift.^{19,20} The fate of the sensory cell with severely damaged cilia is uncertain (Fig. 18). Sensory cells frequently survive a severe noise exposure with only a cilia pathology evident. Some changes in cilia configuration have been clearly linked to changes in individual nerve fiber activity.¹³

Vascular System of the Cochlea

Since it was generally assumed that changes in the metabolism of the cochlea were at least partly responsible for reduced function following noise exposure, correlates of this altered function were sought in the cochlea vasculature. A complex arrangement of arterioles, capillaries, and venules supply and drain the spiral lamina and the lateral wall structures of the spiral ligament, spiral prominence, outer sulcus, and stria vascularis. There is an order to this intertwined meshwork of vessels, and a number of vascular variables can be used to quantify the status of the blood supply to the cochlea.²² The overall impression from

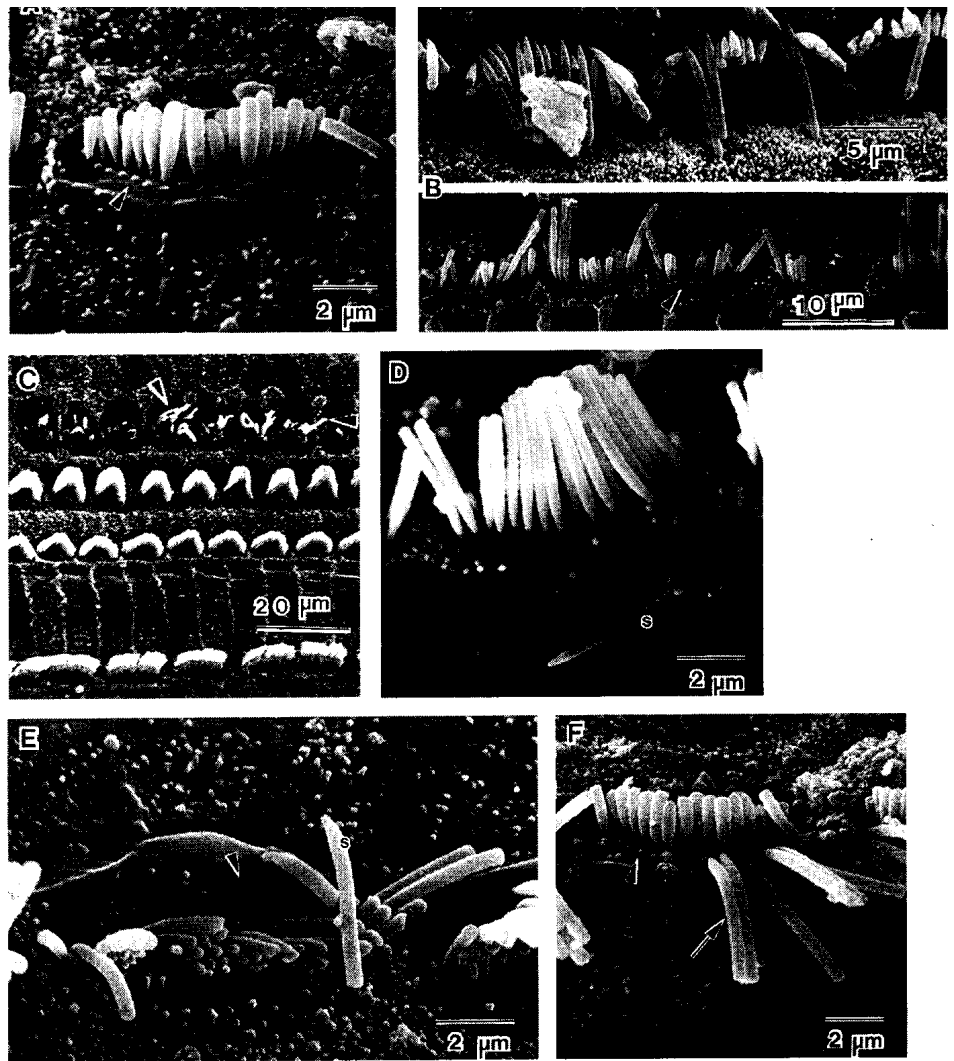


FIGURE 16. A series of SEMs illustrating some cilia pathologies. A, cilia in the tallest row of IHCs in the chinchilla have been broken off at the level of their insertion into the cuticular plate (arrow head). The stumps of the cilia roots are visible. B, missing IHC cilia (arrow heads) seen in sheep following impulse noise exposure. C, the IHC and first two rows of OHCs are normal in appearance while the cilia in the third row of OHCs are severely disarrayed (arrow heads). D and E, fractured IHC cilia (S) lying on the surface of the reticular lamina. F, floppy and bent cilia.

numerous studies is that there is no systematic change in the vasculature that can be easily quantified and related unambiguously to alteration in cochlear function or hearing ability, at least for moderate exposure conditions. Some studies involving severe noise exposures, in addition to showing severe losses to the organ of Corti, show lesions of the stria vascularis. These vascular lesions may be the result of tears or separations of Reissner's membrane,²³ especially at its attachment to the lateral wall, and are probably an effect secondary to the intermixing of the cochlear fluids. While the stria vascularis is vital to the maintenance of the endolymphatic potential (EP), the relatively restricted region of strial damage found



FIGURE 17. Three rows of OHCs several days after noise exposure. All the sensory cells are present, but the cilia are fused and severely disturbed. The cells may survive, but their function will likely be compromised.

Following some noise exposures is probably insufficient to appreciably alter the EP and hence the transduction process. Following less traumatic noise exposures, a variety of more subtle changes can be found in structures such as the stria vascularis and spiral ligament.²¹ These changes amount to a reduction in the number of capillaries, an occlusion of vessels, and changes in red blood cell packing density. Attempts have been made to quantify such changes and to relate them to changes in postexposure hearing function. Figure 18 shows a light micrograph of a surface preparation of the lateral wall that illustrates some of these vascular changes.

PATHOLOGY

Eighth Nerve

One of the earliest signs of noise effects on the structure of the eighth (VIII) nerve are the changes in the synapses at the OHCs and IHCs. The findings of high densities of synaptic vesicles in the efferent endings on the OHCs and the swelling of dendritic terminals at the IHCs are suggestive of high metabolic/functional activity. As levels of noise increase and OHCs are lost, severe swelling and loss of dendritic endings may occur.²⁴ Recently, regeneration of some of these endings has been reported and linked to some of the recovery of threshold usually seen following an acute acoustic trauma. An excitotoxic mechanism involving the neurotransmitter glutamate has been implicated in the pathologic process.⁶ Following severe exposures that lead to losses of IHCs, with a concomitant loss of inner pillar cells, there is a retrograde degeneration of VIII nerve fibers reflected in losses of spiral ganglion cells and morphologic changes in the ascending neural pathways.¹⁵

Relation Between Cochlear Pathology and Hearing Loss

The relation between hearing function and cochlear pathology is complicated, and the implications of the pattern of sensory cell damage for function are not simple,⁹ given the striking difference in the innervation pattern of these types of sensory cells and their roles in

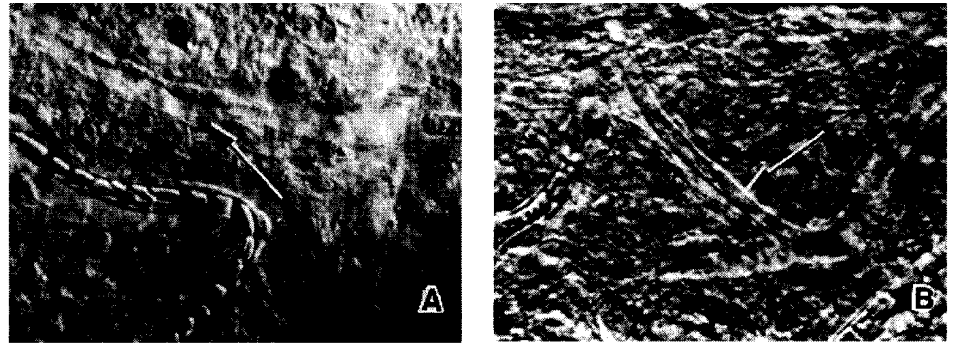


FIGURE 18. Examples of vascular pathology seen in lateral wall system. A, an avascular channel (arrow). B, a collapsed and empty capillary. These changes are found in spiral ligament and stria vascularis following traumatic exposures.

the transduction process. Basic science studies of individual VIII nerve fibers and hair cell changes show that subtle stereocilia defects on IHCs are associated with significant threshold shifts in the responses of individual VIII nerve fibers. Minor loss of OHCs can lead to threshold shifts and, more importantly, changes in tuning characteristics of VIII nerve fibers. On the other hand, stereocilia defects are not necessarily correlated with defects in hearing function.

The relation between cochlear pathology and hearing loss (HL) is not simple. With the loss of OHCs over a region spanning several millimeters, there is often a shift in quiet threshold of 30-50 dB, as measured behaviorally or by evoked potentials. A reasonable generalization is that the first 30-45 dB of hearing loss is often a clue to a loss of the active process or biologic amplifier associated with the action of the OHCs.³ Numerous studies report substantial hair cell losses (both inner and outer) with normal thresholds.¹² Thus, the consequences of hair cell loss on hearing function are difficult to predict with accuracy. Conversely, given a measurable hearing loss, it is difficult to predict the underlying pathology; i.e., the hearing loss may be attributable to sensory cell losses, stria vascularis degeneration, supporting cell failure, or neural degeneration. The fact that similar audiograms have quite different pathologic bases may account for the common clinical observation that individuals with essentially the same audiogram can have markedly different successes with the use of hearing aids.

Biologic Variability

Virtually every characteristic of the species has wide ranges of variability. Susceptibility to NIHL is no exception and, for ostensibly the same exposure to noise, the response can vary from no NIHL to large, debilitating losses. Perhaps one of the best demographic studies, by Taylor et al.,²⁵ examined hearing loss in a group of forge operators who had worked in the same factory for up to 40 years. Noise exposures were well-documented, and relatively accurate otologic records were available for each worker. Figure 19A shows the HL at 4 kHz for industrial workers exposed to the factory noise. Even with careful attention to the principles of demographic research, Taylor et al. found a large range of hearing loss for ostensibly the same exposure. Despite the efforts of Taylor et al.,²⁵ it is almost impossible to control all the relevant variables in a demographic study. Therefore, it is instructive to learn about "variability" in controlled laboratory experiments. Figure 19B shows the range of hearing loss for 5 chinchillas exposed to 161 peak dB impulse noise. Even though the subjects had similar preexposure thresholds, received the same noise exposures, and lived in the same environment, a range of variability still exists that is similar to variability found in demographic studies.

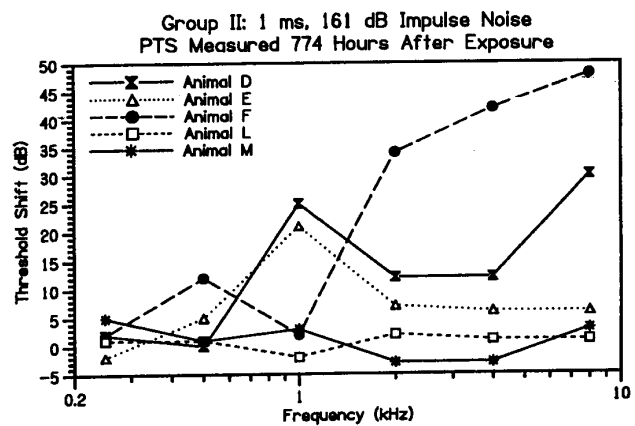
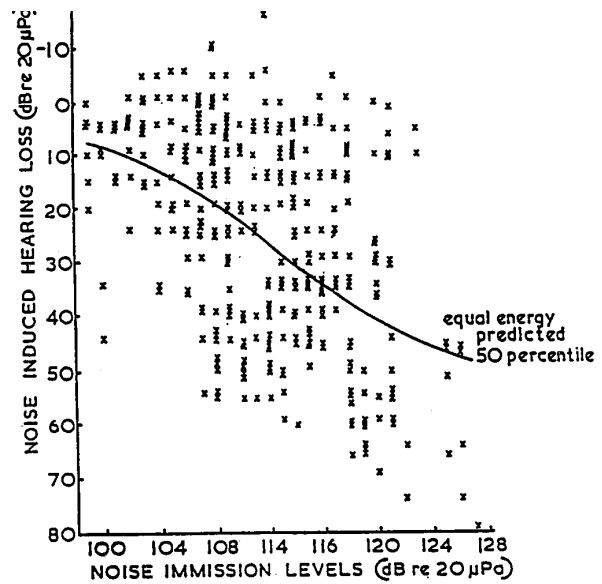


FIGURE 19. A, the range in hearing level at 4 kHz for workers exposed to noise in a forging plant. Note for most points on the horizontal axis (doses) there is a 40-70 dB range of thresholds for ostensibly the same exposure. B, the range in PTS for five chinchillas exposed to 161 dB impulse noise. Despite the rigorous experimental control, there is still enormous variability.

SUMMARY

The effects of noise are pervasive, and pathology can be found in the neural, sensory, supporting, and vascular cells of the cochlea after certain noise exposures. Several issues need to be addressed. First, what is the source of the wide range of susceptibility seen in people and animals that have similar noise histories but markedly different response to noise? Does this suggest a genetic factor in susceptibility to NIHL? Second, given the complexity of the biologic changes following noise exposures, it is difficult to predict the underlying pathology from standard audiologic tests. However, it would be desirable to distinguish sensory and neural and stria pathology because it is likely that the success of any aural rehabilitation will depend on knowledge of the type of pathology.

ever, one of the most exciting recent findings that undoubtedly will affect people with NIHL is that it is possible for sensory cells to regenerate.⁶ Research into the fundamental biochemical processes responsible for inducing regeneration is being pursued at a number of laboratories. The hope is to be able to repopulate the mammalian sensory epithelium with viable sensory cells.

REFERENCES

1. Axelsson A: The vascular anatomy of the cochlea in the guinea pig and in man. *Acta Otolaryngol Suppl* 243: 1968.
2. Bohne B, Rabbitt R: Wholes in the reticular lamina after noise exposure: Implications for continuing damage in the organ of Corti. *Hear Res* 11 :41-53, 1983.
3. Brownell R: Outer hair cell electromotility and otoacoustic emissions. *Ear Hear* 11 :82-89, 1990.
4. Ciazzo AJ, Tonndorf V: Ear canal resonance and TTS. *J Acoust Soc Am* 61:78, 1977.
5. Colletti V, Sittoni W: Noise history, audiometric profile and acoustic reflex responsivity. In Salvi R, Henderson D, Hamernik R, Colletti V (eds): *Basic and Applied Aspects of Noise Induced Hearing Loss*. New York, Plenum Press, 1985, pp 269-347.
6. Cotanche DA: Regeneration of hair cell stereociliary bundles in the chick cochlea following severe acoustic trauma. *Hear Res* 30: 18 I - 194, 1987.
7. Davis H, Morgan CT, Hawkins JE, et al: Temporary deafness following exposure to loud sounds and noises. *Acta Otolaryngol Suppl* 88:1-57, 1950.
8. Engstrom H, Ades HW, Andersson A: Structural pattern of the organ of Corti. Stockholm, Almqvist & Wiksell, 1966.
9. Hamernik RP, Patterson JH, Turrentine GA, Ahroon WA: The quantitative relation between sensory cell loss and hearing thresholds. *Hear Res* 38:199-212, 1989.
10. Hamernik RP, Turrentine G, Wright CG: Surface morphology of the inner sulcus and related epithelial cells of the cochlea following acoustic trauma. *Hear Res* 16: 143- 160, 1984.
11. Hellstrom PA: Individual differences in peripheral sound transfer functions: Relation to NIHL. In Axelsson A, Hamernik RP, Henderson D, et al (eds): *Noise Induced Hearing Loss*. New York, Thieme, 1995 (in press).
12. Henderson D, Hamernik RP: Impulse noise: Critical review. *J Acoust Soc Am* 80:569-584, 1986.
13. Liberman MC, Dodds LW: Single-neuron labeling and chronic cochlear pathology. III. Stereocilia damage and alterations of threshold tuning curves. *Hear Res* 16:55-74, 1984.
14. Mills JH, Gilbert RM, Adkins WY: Temporary threshold shifts in humans exposed to octave band noise for 16-24 hours. *J Acoust Soc Am* 65: 1238-1248, 1979.
15. Morest DK: Degeneration in the brain following exposure to noise. In Hamernik RP, Henderson D, Salvi RJ (eds): *New Perspectives on Noise-Induced Hearing Loss*. New York, Raven Press, 1982, pp 87-93.
16. Pujol R, Puel JL, Gervais d'Aldin C, Eyblin M: Physiopathology of the glutamatergic synapses in the cochlea. *Acta Otolaryngol Suppl* 476:32-36, 1993.
17. Reger SN: Effect of middle ear muscle action on certain psychophysical measurements. *Ann Otol Rhinol Laryngol* 69:1179-1198, 1964.
18. Salvi R, Hamernik R, Henderson D: Physiological bases of sensorineural hearing loss. In Tobias JV, Schubert E (eds): *Hearing Research and Theory*. San Diego, Academic Press, 1982.
19. Saunders JC, Cohen YE, Szymko YM: The structural and functional consequences of acoustic injury in the cochlea and peripheral auditory: A five year update. *J Acoust Soc Am* 90: 136-146, 1991.
20. Saunders JC, Flock A: Recovery of threshold shift in hair-cell stereocilia following exposure to intense stimulation. *Hear Res* 23:233-243, 1986.

- vascular system of the chinchilla cochlea. *Ann Otol Rhinol Laryngol* 94:87-92, 1985.
22. Shaddock LC, Hamernik RP, Wright CG: A morphometric technique for analysis of cochlear vessels. *Hear Res* 20: 109-117, 1985.
 23. Shaddock LC, Wright CG, Hamernik RP: A morphometric study of microvascular pathology following experimental rupture of Reissner's membrane. *Hear Res* 20:119-129, 1985.
 24. Spoendlin HH: Anatomical changes following various noise exposures. In Henderson D, Hamernik RP, Dosanjh DS, Mills JH (eds): *Effects of Noise on Hearing*. New York, Raven Press, 1976, pp 69-90.
 25. Taylor W, Pearson J, Main A, Burns W: Study of noise and hearing. *J Acoust Soc Am* 38:113-120, 1965.
 26. Woodford CW, Henderson D, Hamernik RP: Static and dynamic impedance of the chinchilla middle ear. *J Acoust Soc Am* 54:327, 1973.
 27. Zakrisson JE, Borg E, Liden C, Nilsson R: Stapedius reflex in industrial impact noise and fatigability and role for temporary threshold shifts (TTS). *Scand Audiol Suppl* 12:326, 1980.