Chapter 178: Sensorineural Hearing Losses of Adulthood

A. Julianna Gulya

Because cochlear function depends on the integrated action of the hair cells, cochlear neurons, stria vascularis, and neural network, all of which are critically dependent on a continuous blood supply, dysfunction or degeneration of any of these elements, alone, or in combination, can result in cochlear impairment and sensorineural hearing loss (SNHL). Various systemic disorders and local insults can precipitate, or colligatively augment, impaired cochlear function. This chapter is an overview of those disorders that have SNHL as their sole or predominant initial symptom in adults. An approach to the diagnosis and management of these disorders will be discussed. A more detailed discussion of several of the causative entities, such as perilymph leaks, sudden hearing loss, autoimmune disorders, cochlear trauma, and central auditory dysfunction can be found in other chapters of this volume.

Evaluation of the Adult With Sensorineural Hearing Loss

To determine the cause of SNHL the physician must consider and synthesize cochlea-based signs and symptoms with elements related to concurrent or past disease, hereditary considerations, and socioeconomic factors. The hearing loss should first be assessed and categorized according to whether one or both ears are involved, the rapidity of onset and progression, fluctuation, and any associated symptoms such as tinnitus, aural fullness, and pain. The handicap perceived by the patient should be assessed. The temporal bone environs of the cochlea should be explored, seeking evidence of infection and other causes of the patient's symptoms.

The general health status of the patient should be determined. Systematic questioning should ascertain the presence or absence of allergies, current or past drug usage, particularly with reference to potentially ototoxic medications, and whether the patient has systemic illnesses, such as endocrine, metabolic, autoimmune, or cardiovascular disorders. The family history should be reviewed for any suggestions of hereditary or familial hearing loss; similarly, the patient's work and recreational environments should be evaluated for possible adverse environmental, dietary, or habitual noxious influences.

Physical examination paralles the history, in that it expands from a focus on the ears and builds on clues contained in the history. Tuning fork tests can suggest a possible conductive component, and the whisper test gives an immediate, although crude, estimate of the severity of the hearing loss and the quality of speech discrimination. The presence of malodorous, purulent debris in the external auditory canal, with or without visualization of an inflammatory polyp or distinct cholesteatoma, raises concerns of a chronic infectious basis for the hearing loss. More often, however, the otologic examination is unrevealing. Tinnitus, particularly if unilateral or pulsatile, may be auscultated (Toynbee tube or bell stethoscope) and the effects of vascular compression or head manipulation evaluated.

The complete head and neck examination can detect other neurologic defects, the stigmata of hereditary dysfunction (eg, the white forelock of Waardenburg's syndrome), or signs of systemic disease (eg, xanthelasma of hypercholesterolemia).

The purposes of diagnostic testing are multiple and include (1) to establish severity, (2) to identify the site of lesion, (3) to determine the etiology, and (4) to determine curative or rehabilitative therapy.

A complete audiogram initiates the auditory and vestibular testing and suggests direction for further testing. Pure-tone thresholds are obtained for both air and bone, speech discrimination is assessed along with PIPB (performance-intensity function for phonetically balanced words) testing, and tympanometry is performed. As emphasized by Toner and Kerr (1987), an apparently profound SNHL may instead represent a severe SNHL with conductive overlay, a situation that may be differentiated only by complete audiologic evaluation. Auditory brainstem response (ABR) testing is performed if the audiogram suggests retrocohlear disease, if the SNHL is unilateral or asymmetric, or if there is evidence for other neurologic dysfunction such as dizziness or facial nerve paresis. Occasionally, the ABR may be used to validate the audiogram in difficult-to-test or malingering patients.

Electrocochleography (ECoG) comprises measurement of the cochlear microphonic, the summating potential (SP), and the action potential (AP) of the auditory nerve. Recently, an elevated SP/AP ratio has been regarded as suggestive of endolymphatic hydrops; in such patients, Ferraro et al (1985) reported that the combination of hearing loss and aural fullness was the strongest predictor of an elevated SP/AP ratio. ECoG is also useful in those instances where wave I of the ABR is poorly defined since the AP of the ECoG corresponds to wave I of the ABR.

The discovery of otoacoustic emissions (OAEs) has rapidly led to their use in the evaluation of cochlear function (Martin et al, 1990); OAEs reflect the micromechanical activity of outer hair cells, can be measured objectively and noninvasively from the external auditory canal, and operate at low levels of stimulation (Martin et al, 1990). Distortion product emissions (DPEs) are one type of OAE, and they are elicited by the simultaneous application of two different tones to the ear canal; DPEs are thought toarise from particular loci along the cochlea varying with, and predicted by, the frequencies of the two eliciting tones. Thus, it may be that DPEs can be used to monitor cochlear function at specified frequencies. DPEs seem sensitive toearly noise-induced SNHL and the fluctuant SNHL of Ménière's disease (Martin et al, 1990), and they may be helpful in the near future for the assessment of patients receiving otototoxic medications or even in general auditory screening.

Vestibular testing is obtained when vertigo or imbalance is present, or in selective cases of unilaterally or bilaterally asymmetric SNHL (Kumar et al, 1986).

Most cases of bilaterally symmetric SNHL will not require radiologic evaluation; however, if there is evidence of temporal bone or retrocochlear disease, the additional information provided by imaging modalities, such as computerized tomographic (CT) scanning and magnetic resonance imaging (MRI), can be helpful. In general, disorders confined to the temporal bone, for example, chronic otitis media, cholesteatoma, or developmental anomalies, are best evaluated by high-resolution, bone-algorithm driven, axially and coronally sampled, temporal bone CT scanning. MRI supplemented by gadolinium enhancement and/or gated acquisition techniques (MR angiography) has assumed a preeminent role in the evaluation of retrocochlear, craniocervical junction, and even vascular disorders of the head and brain.

Laboratory testing is obtained in accordance with the findings of the initial history and physical examination. Routine evaluation of hematologic/metabolic parameters such as electrolytes, glucose, cholesterol, lipids, and thyroid function may reveal abnormalities that more likely are coincidental, rather than closely related to, the patient's SNHL. Similarly, screening for autoimmune disorders indiscriminately does not appear warranted. Either the MHA-TP (Microhemagglutination test for *Treponema pallidum*) or FTA-ABS (fluorescent treponemal antibody absorption) are routinely obtained, because abnormal results are obtained in 6.5% of cases of unexplained SNHL (Zoller et al, 1978) and 7% of patients with Ménière's disease (Pulec, 1972). Additional Western blot analysis may prove helpful in determining activity of luetic infection (see below).

The diagnostic possibilities for SNHL in the adult include ototoxic medications; neurologic disorders; vascular or hematologic diseases; infectious, metabolic, or renal abnormalities; normal aging processes; endocrine dysfunction; disorders of bone metabolism; hereditary disorders; autoimmune diseases; and disorders of unknown etiology. These various possibilities will be reviewed as they are related to the type of SNHL they cause most frequently. If the onset of SNHL dates to childhood or infancy, different diagnostic considerations are more likely, and these are reviewed in Chapter 176.

Ototoxicity

According to Catlin (1985), a minimum of 96 agents are known to have ototoxic potential and have been grouped into 18 categories (see box). Those agents with cochleotoxic potential that will be discussed here include antibiotics, loop diuretics, antiinflammatory medications, cancer chemotherapeutic drugs, and hematologic agents.

Aminoglycoside antibiotics

The aminoglycoside antibiotics include the older agents streptomycine sulfate, dihydrostreptomycin, kanamycin, and neomycin, and the newer drugs gentamicin, tobramycin, amikacin, and netilmicin (Matz, 1990); their common bacteriocidal effect is mediated by inhibition of protein synthesis at the ribosome level. They are administered parenterally for the treatment of systemic, gram-negative aerobic infections; they are excreted unmetabolized; and they can be concentrated in the urine; thus, serum and therefore periolymph levels can be elevated in renal failure. Although their serum half-life approximates 80 minutes (Fee, 1980), the perilymph half-life is much longer, varying from 5 to 15 hours (Fee, 1980; Lerner and Matz, 1980), especially in renal failure (Matz, 1990). Aminoglycosides enter the hair cell by an energy-dependent transport process and can then interfere with other intracellular reactions (Matz, 1990). The potential ototoxicity of the aminoglycosides was recognized initially with streptomycin sulfate and dihydrostreptomycin (Schuknecht, 1974); further experience has revealed different relative cochleotoxicity and vestibulotoxicity for the various aminoglycosides. Aminoglycosides that are particularly cochleotoxic indlude kanamycin, tobramycin, amikacin (Catlin, 1985), neomycin, and dihydrostreptomycin (Schuknecht, 1974). The ototoxicity associated with the newer aminoglycosides is less, but similar in character to that seen with the older agents. It has been argued that reported ototoxicity rates are too low (Matz, 1990) because of failure to adequately test all treated patients or too high because the criteria for cochleotoxicity could conceivably fall within the test/retest variability of audiometric evaluation (Brummett and Morrison, 1990). Nonetheless, available information

from retrospective and prospective studies (Fee, 1980; Kahlmeter and Dahlager, 1984; Lerner and Matz, 1980; Matz, 1986) suggests cochleotoxicity rates of 3.9% to 9% with amikacin, 7% to 18% for gentamicin, 6.1% to 15.3% for tobramycin, and 2.4% with netilmicin.

Histopathologic correlates of cochlear intoxication include (Nadol, 1981; Schuknecht, 1974) hair cell loss, most pronounced basally and for the outer hair cells, with variable degeneration of cochlear neurons. The associated hearing loss involves the 1000 Hz to 8000 Hz range, with the high frequencies affected most commonly as would be predicted from histopathologic findings (Fee, 1980). Cochleotoxicity can occur despite maintaining drug levels within "safe" limits and in the absence of nephrotoxicity (Fee, 1980).

Delayed cochleotoxicity that begins even after discontinuation of the drug has been associated with neomycin (Schuknecht, 1974), dihydrostreptomycin (Schuknecht, 1974), gentamicin (Fee, 1980; Matz, 1986), tobramycin (Fee, 1980; Matz, 1986), amikacin (Matz, 1986), and netilmicin (Matz, 1986). With parenterally administered aminoglycosides one would expect symmetric cochlear damage; however, prospective studies have shown that from 50% to 91% (Fee, 1980) of cochleotoxic events are either unilateral or bilaterally asymmetric. The hearing loss may progress with continuation of aminoglycoside therapy, and in some cases, it may even progress despite cessation of therapy (Fee, 1980). Reversibility of SNHL has also been reported (Fee, 1980; Lerner and Matz, 1980; Matz, 1986) in up to 55% of the cases (Fee, 1980) and occurs from 1 week to 6 months after treatment. Recovery is less likely if (1) the SNHL is delayed in onset; (2) the SNHL is immediate in onset and progresses after drug discontinuation; (3) the SNHL is greater than 25 dB; or (4) further therapy with aminoglycosides is required despite the detection of immediate toxicity (Fee, 1980).

Risk factors for ototoxicity with aminoglycosides have been enumerated by Matz (1990) and include the presence of renal disease, longer duration of therapy, elevated serum peak and/or trough levels, advanced age, and concomitant use of other ototoxic medications, especially the loop diuretics furosemide and ethacrynic acid. Animal studies have shown potentiation of aminoglycoside ototoxicity by pretreatment with butathionine sulfoximine (Hoffman et al, 1988) believed to be mediated by glutathione depletion, or with concomitant vancomycin therapy (Brummett et al, 1990).

Measures that should be taken to minimize the risk of cochleotoxicity include repetitive monitoring of serum peak and trough antibiotic levels and increasing the interval between successive antibiotic doses or courses of therapy to allow clearance of the drug from perilymph, especialy in cases associated with renal failure (Matz, 1990). Ototoxicity can occur despite keeping serum levels within recommended guidelines (Fee, 1980), and genetic susceptibility has been thought to account for such SNHLs (Nadol, 1981). Alternatively, others have suggested, at least with respect to amikacin, that ototoxicity is related to total drug exposure independent of plasma levels; Beaubien and associates (1989) found that cochleotoxicity of amikacin in the guinea pig correlated with the total dose of amikacin administered *or* with the area under the curve describing plasma concentration versus time, and proposed that these parameters be used in monitoring the toxicity rather than peak and trough serum levels.

4

For now, once SNHL is detected during the course of aminoglycoside therapy, little can be done except to discontinue the drug. There is promising work in the animal model that suggests that concomitant administration of fosfomycin (a phosphonic acid antibiotic) may protect against aminoglycoside ototoxicity (Ohtani et al, 1985); human application has not yet been reported.

Vancomycin

The intravenous administration of vancomycin has been linked with development of SNHL when serum levels have been equal to or greater than 30 microg/mL. It is excreted by the kidneys, and renal failure can prolong vancomycin half-life (Hermans and Wilhelm, 1987). The drug is also nephrotoxic, and both ototoxicity and nephrotoxicity may be decreasing because of the availability of newer, more highly purified preparations of the drug; however, no hard data are available to confirm or refute these presumptions (Medical Letter, 1986).

Topical ear preparations

Otic preparations commonly used in the topical treatment of bacterial infections incorporate neomycin, polymyxin B, and propylene glycol; ophthalmologic preparations containing chloramphenicol are also used. Animal studies demonstrate cochlear damage by these various preparations (Leach et al, 1990; Morizono, 1988), but there are anatomic and pathologic differences between the animal model and man. Podoshin et al (1989) reported a small (approximately 6 dB) but statistically significant worsening of hearing in patients treated for chronic otitis media (COM) with ototoxic antibiotic drops compared with controls treated only with steroid-containing drops. When very concentrated solutions of aminoglycosides (streptomycin, gentamicin) are introduced into the middle ear cavities of patients with Ménière's disease (who have normal middle ears and presumably round window membranes) to unilaterally ablate peripheral vestibular function, significant cochlear ototoxicity can result in some of the patients (Bagger-Sjoback et al, 1990; Schuknecht, 1974). Even though these studies suggest that ototoxicity is possible or likely from the use of aminoglycoside ear drops, an incredibly large number of ears have received these topical medications over the many years of their use without the emergence of any obvious causal relationship of the drops to a clinically significant hearing loss. Nonetheless, the use of otic preparations, particularly with tympanic membrane perforations, should be carefully monitored by the physician. Fosfomycin seems to diminish the ototoxicity of polymyxin B in the chinchilla model when instilled simultaneously with the otic preparation (Leach et al, 1990); clinical trials in the human have yet to be accomplished.

Antimycotic preparations, such as M-cresul acetate, clotrimazole, tolnaftate, as well as propylene glycol and acetic acid (2%) appear to have some potential for ototoxicity in the guinea pig (Marsh and Tom, 1989) when allowed to enter the tympanic cavity. Although the correlation to the human is questionable, the same prudence should be observed in their use.

Loop diuretics

The loop diuretics, such as furosemide (Lasix), ethacrynic acid (Edecrin), and bumetanide (Bumex), constitute a class of drugs that act on the proximal renal tubule in the loop of Henle to promote diuresis; they have been linked to the development of a bilaterally symmetric SNHL, which may be sudden in onset (Arnold et al, 1981; Matz, 1990; Schuknecht, 1974). The SNHL caused by loop diuretics usually is transient, reversing within 30 minutes to 24 hours of administration (Koegel, 1985). Risk factors for developing ototoxicity include renal failure, rapid rate of administration, and concomitant aminoglycoside administration (Koegel, 1985).

The loop diuretics appear to interfere with enzymatic function in the stria vascularis, and histopathologically edema, thickening, and cystic degeneration is seen (Arnold et al, 1981; Koegel, 1985; Matz and Hinojosa, 1973; Schuknecht, 1974). Hair cell loss and endolymphatic hydrops have also occasionally been observed.

Alterations in the electrophysiologic response attributed to loop diuretics include a prolonged decrease in the cochlear microphonic (Schuknecht, 1974) and the endocochlear potential (Arnold et al, 1981), changes that could reflect dysfunction of the stria and/or hair cells (Koegel, 1985; Schuknecht, 1974).

Recent animal studies (Rybak and Whitworth, 1988; Rubak et al, 1986) have suggested that the use of intravenous quinine as a protective pretreatment for furosemide and ethoacrynic acid otototixicity appears to prevent the ototoxicity induced by the loop diuretics. Similarly, furosemide ototoxicity in animals can be ameliorated by pretreatment with intravenous sodium salicylate. Human correlation is yet to be established.

Ototoxicity from loop diuretics is best averted by using a slow rate by intravenous infusion (less than 15 mg/min) (Matz, 1990), decreasing the dosage in the presence of renal failure, and avoiding the concomitant use of aminoglycosides (Koegel, 1985). Once ototoxicity from these agents is established, the only management is drug cessation.

Cisplatin

Cisplatin (cis-diamminedichloroplatinum) is a cancer chemotherapeutic agent used to treat squamous cell carcinoma of the head and neck and other malignancies (Kopelman et al, 1988). In addition to dose-limiting nephrotoxicity, ototoxicity can occur, and it is manifested predominantly as an irreversible, bilaterally symmetric, progressive SNHL that initially ismost severe at the high frequencies (Kopelman et al, 1988; Matz, 1990) and is generally accompanied by tinnitus; vertigo and disequilibrium have also been reported (Schaeffer et al, 1981). The incidence of ototoxicity varies with the rapidity of infusion as well as the frequency bands tested. With bolus administration of high-dose cisplatin (150 to 225 mg), 100% of patients tested with ultra-high-frequency audiometry showed a loss of response to tones of 9000 Hz and above after one to two doses (Kopelman et al, 1988); in another study, 88% of patients treated with high-dose cisplatin had evidence of SNHL as measured by conventional audiometry (Myers et al, 1991). With additional cisplatin, the hearing deficit encompasses progressively lower frequencies and becomes progressively more severe (Myers et al, 1991).

Chemically, the toxicity of cisplatin is thought to involve inhibition of adenylate cyclase. It is significant that, similar to the aminoglycosides, cisplatin can be concentrated in the inner ear (Kopelman et al, 1988).

Pathologic examination has revealed outer hair cell loss that is most severe basally; with higher doses, inner hair cell, strial, and neural damage has also been reported (Kopelman et al, 1988).

The risk of ototoxicity appears to be related more to the size of each dose rather than the cumulative amount of administered drug; no ototoxic changes were found (by standard audiometry) if peak plasma concentrations remained below 1 microg/L (Laurell and Jungnelius, 1990).

Two promising avenues are being explored in animal models to reduce or eliminate this ototoxicity. The first involves the use of platinum analogues, which do not possess the ototoxicity and nephrotoxicity of cisplatin (Schweitzer et al, 1986b). The second involves the simultaneous administration of fosfomycin with cisplatin to lessen the resultant ototoxicity (Schweitzer et al, 1986a).

Nitrogen mustard

Mechlorethamine (nitrogen mustard) is an alkylating agent used in cancer chemotherapy (Segal and Duckert, 1986). Treatment with massive doses of nitrogen mustard (0.6 to 1.5 mg/kg) is associated with the development of a moderately severe to profound bilateral SNHL in most patients (Segal and Duckert, 1986). In general, the SNHL is permanent, although one case report describes a patient whose SHNL after low-dose nitrogen mustard was reversible (Segal and Duckert, 1986).

Histopathologic examination in the human has shown shrinkage of the organ of Corti without hair cell loss, while experimental studies in the cat have documented outer and inner hair cell loss in the basal and middle cochlear coils (Schuknecht, 1974).

Deferoxamine

Deferoxamine is an iron-chelating agent that is used to avert generalized iron loading by promoting urinary iron excretion in patients with beta-thalassemia major and steroid-unresponsive Diamond-Blackfan anemia (Olivieri et al, 1986). High-dose deferoxamine therapy has been implicated in an acute onset SNHL, often associated with acute onset visual loss (Olivieri et al, 1986). The SNHL varies from 30 to 100 dB, mostly involves the high frequencies, and usually is permanent (Gallant et al, 1987). No histopathologic correlates have been reported.

Recommendations include monitoring auditory thresholds with routine audiometry, which appears to be more sensitive than ABR testing, and modifying or ceasing deferoxamine therapy when signs of ototoxicity develop (Gallant et al, 1987).

Neurologic Disorders

Multiple sclerosis

Only 4% of patients with multiple sclerosis (MS) experience SNHL in the course of their disease (Grenman, 1985); and in at least one study (Grenman, 1985) only 20% to 40% of those patients with MS who were found to have abnormal audiograms had complained of a hearing loss (Grenman, 1985). Despite the seeming infrequency of the occurrence of SNHL, several audiometric patterns are seen; the audiometric profile may be unilateral, bilaterally asymmetric, or bilaterally symmetric (Schweitzer and Shepard, 1989; Schweitzer et al, 1986b). Reversible and progressive SNHLs, variably associated with tinnitus and vertigo, have been reported (Daugherty et al, 1983; Schweitzer and Shepard, 1989). The most common pattern is a high-frequency loss (Musiek et al, 1989). Speech discrimination may be normal or disproportionally reduced in relation to pure-tone thresholds. The site of lesion may lie within the cochlear nerve (Hopf and Maurer, 1983) or in the brain stem (Daugherty et al, 1983).

Special tests of auditory function have been scrutinized for their ability to detect clinically inapparent auditory dysfunction in MS. Acoustic reflex dynamics (onset, latency, rise time, and amplitude) have been studied in asymptomatic (Keith et al, 1987; Wiegand and Pock, 1988) and symptomatic (Musiek et al, 1989) patients with conflicting results regarding the latency, rise time, and amplitude reduction (Jerger et al, 1986).

ABR testing similarly has shown varying results. Wave I latency has been reported to be prolonged in normal-hearing MS patients (Verma and Lynn, 1985) and in MS patients with SNHL (Hopf and Maurer, 1983). Other studies have found normal wave I latency and emphasized abnormal latencies for higher waves (Daugherty et al, 1983; Keith et al, 1987). Similarly, absence of waveform, poor waveform morphology, and poor repeatability of the waveform have been linked inconsistently to MS (Keith et al, 1987). ABR waveform morphology and amplitude may deteriorate when the rate of click presentation is increased from the usual 10 per second to 40 per second (Schweitzer and Shepard, 1989).

MR imaging has been able to document foci of demyelination, in the inferior colliculus, for example (Armington et al, 1988), documenting a brainstem site of lesion in some cases.

The hearing loss, along with the audiologic abnormalities, tends to recover over the course of months (Daugherty et al, 1983; Hopf and Maurer, 1983), although permanent SNHL may occur.

Benign intracranial hypertension

Benign intracranial hypertension (BIH), also known as pseudotumor cerebri, is manifested by increased intracranial pressure without focal neurologic signs and is typically encountered in young, obese females. The initial symptoms of BIH are headaches and/or visual blurring (Sismanis et al, 1985). SNHL, tinnitus, (pulsatile), and vertigo may also occur, and occasionally these symptoms prompt the patient to seek medical consultation. There are many causes of both tinnitus (Sismanis et al, 1990a) and pseudotumor cerebri (Sismanis et al, 1985). The diagnosis is suggested by fundoscopic evidence of papilledema, and it is

confirmed by careful lumbar puncture documentation of increased cerebrospinal fluid pressure in excess of 200 mm water (Sismanis et al, 1990a).

The SNHL is a variably severe, low-frequency, sometimes fluctuant loss, which can be present in one or both ears (Sismanis et al, 1985; 1990a). The pulsatile tinnitus is objective, and it is uniformly diminished or abolished by ipsilateral internal jugular vein compression or by cerebrospinal fluid drainage (Sismanis et al, 1990a); interestingly, the latter measure uniformly normalizes or improves the factitious SNHL from a venous hum of internal jugular vein origin (Rothstein et al, 1985). The low frequency SNHL in such cases may actually result from the masking effect of the pulsatile tinnitus (Sismanis et al, 1990b). Aural fullness and vertigo (Sismanis et al, 1990a) may also be present.

Both ABR and ECoG abnormalities have been documented (Sismanis et al, 1990b), most commonly involving increased interpeak latencies, but increased SP/AP ratios were also found in two of four patients tested. Stretching, compression, or edema of the cochlear nerve and brain stem from the increased intracranial pressure, or a combination of these effects, have been proposed as causes for the ABR abnormalities (Sismanis et al, 1990b).

Medical management includes weight reduction, acetazolamide, and furosemide therapy; progressive visual loss, headache, and disabling tinnitus may require lumbar-peritoneal shunt placement (Sismanis et al, 1990b).

Vascular Disorders

Migraine

Migraine headaches are very common in the US population, afflicting an estimated 15% of men and 20% to 25% of women (Olsson, 1991); historically, the nosology of migraine headaches has been confusing, and the currently recommended terminology substitutes *migraine-with-aura* for the term *classic migraine* (Olsson, 1991). Various auditory symptoms have been related to *migraine-with-aura,* particularly, but not exclusively, with the basilar migraine subtype (Harker and Rassekh, 1988; Kayan and Hood, 1984; Olsson, 1991), including SNHL, tinnitus, pitch distortion, speech distortion, poor localization of sound, and loudness intolerance (phonophobia) (Kayan and Hood, 1984).

In their series of 200 unselected migraine patients, Kayan and Hood (1984) found that 4.5% acknowledged some type of cochlear symptoms, while 15.5% had both cochlear and vestibular complaints, which could occur as part of the migraine prodrome, during the headache phase, or as a "migraine equivalent". Phonophobia (81% of the cases) was the most commonly identified symptom; SNHL occurred in 13 patients (7%), was restricted primarily to the high frequencies, and was either transitory or permanent. No ABR testing was performed, and other possible etiologies for the high-frequency SNHL were not specifically considered.

Olsson (1991) in a recent study of 50 patients with basilar migraine documented a bilateral, low-frequency SNHL in 46% of his patients, while a unilateral, low-frequency SNHL was seen in an additional 34%; fluctuation of hearing was noted in most patients (Olsson, 1991). In many cases, the low-frequency SNHL was mild, but a few patients had

severe SNHL. ABR testing revealed abnormalities of morphology or latency with moderate to severe SNHL (Olsson, 1991).

The pathophysiology of the SNHL in migraine has been postulated to result from vasoconstriction with ischemia and even infarction; indirect mechanical pressure from dilated arteries; transudation of edematous fluid; and the action of vasoactive transmitters (Kayan and Hood, 1984). Treatment of migraine consists of beta and calcium channel blockers, with or without antiserotonin agents (Olsson, 1991); no rigorously performed study regarding the use of these agents in migraine-associated SNHL could be found.

Vertebrobasilar dolichoectasia

Dolichoectasia of the vertebrobasilar arterial system is a rare cause of unilateral, progressive SNHL (Campbell et al, 1986); it comprises widening and serpentine tortuosity of the involved arteries, and it is generally seen in middle-aged men with hypertension and atherosclerosis (Gulya et al, 1986). The SNHL may be associated with vertigo or facial paresis (Gulya et al, 1986). Both pressure effects exerted by the dilated arteries and mural thrombi with distal embolism are thought to underlie the SNHL, vertigo, and facial paresis, and they may also have the potential to cause hemiparesis and dysfunction of the lower cranial nerves, brain stem, and cerebellum (Gussen, 1977; Moseley and Holland, 1979; Musiek et al, 1987).

Contrast-enhanced CT scanning may suggest the diagnosis, which has generally been confirmed by angiography; MR angiography may prove a less invasive and equally effective diagnostic modality.

In general, treatment options include attempts to reduce the risk of developing mural thrombi with aspirin and/or dipyridamole (Persantine) therapy.

Hematological Disorders

Sickle cell anemia

Sickle cell disease (SCD) is a common hereditary hematologic disorder associated with the homozygous state for the recessive sickle cell gene. hen abnormal hemoglobin of affected individuals is exposed to low oxygen tension, it becomes a semisolid gel that causes distortion (sickling) of the red blood cells, which then cannot pass through small arteries and capillaries causing a "sickle cell crisis" (Frenkel, 1982). The red blood cell clots essentially cause thrombosis and infarction. When this occurs in the end vessels of the cochlea, it seems likely that it could result in the SNHL observed in patients with sickle cell anemia, although the mechanism of the SNHL is still debated (Elwany and Kamel, 1988).

SNHL is seen in up to 22% of cases of SCD (Elwany and Kamel, 1988) and can be unilateral, sudden, reversible (Urban, 1973), and worst for the high frequencies (Elwany and Kamel, 1988). With repetitive crises, the potential for aggravation of SNHL exists. Some evidence indicates an increased incidence of SNHL as the crises increase in frequency and severity (Elwany and Kamel, 1988).

Temporal bone examination of one patient revealed strial degeneration and an absence of hair cells (Morgenstein and Manace, 1969). Central auditory structures can also be affected during crises, and ABR abnormalities have been documented both with and without other neurologic manifestations of sickle cell crisis (Elwany and Kamel, 1988).

As yet there is no information relating hyperbaric oxygen therapy for sickle cell crises to improvement or restoration of hearing loss.

Blood viscosity disorders

Blood viscosity alterations potentially disruptive to cochlear oxygenation have been suggested to explain otherwise idiopathic, bilaterally progressive SNHL (Gatehouse et al, 1989; Hildesheimer et al, 1990; Keith et al, 1987; Rubinstein et al, 1988). The viscosity of whole blood at high shear rates, as standardized for hematocrit, varies with plasma viscosity and red blood cell (RBC) deformability, since plasma viscosity and whole blood viscosity can be measured, RBC deformability can be derived (Gatehouse et al, 1989).

Published studies, variably incorporating appropriate control groups, suggest that SNHL, particularly at 2000 Hz and 4000 Hz, can be related to increased whole blood viscosity (Browning et al, 1986; Gatehouse et al, 1989; Hildesheimer et al, 1990); plasma viscosity has either shown no significant relationship to SNHL (Gatehouse et al, 1989) or, if increased, it seems to be associated with better hearing thresholds (Browning et al, 1986).

Much work remains in clarifying whether there is a relationship between rheologic characteristics of whole blood and red blood cells, such as RBC deformability or filterability, and SNHL.

Infections

Syphilis

Luetic or syphilitic otitis, arising from either congenital or acquired infections, has been linked to SNHL that can be sudden, fluctuating, or progressive; moderate or profound; flat or upward sloping; unilateral, bilaterally asymmetric, or bilaterally symmetric; and of childhood or adult onset (Darmstadt and Harris, 1989). Tinnitus, reduced discrimination, and vertigo are variably encountered.

There are two principal histopathologic correlates of syphilitic SNHL. The first is a mening-neuro-labyrinthitis, and it may be seen in early congenital syphilis or in acquired syphilis complicated by meningitis (Darmstadt and Harris, 1989; Schuknecht, 1974). The second is temporal bone osteitis with obliterative endarteritis, round cell infiltration, gumma formation, or fistulization of the labyrinth (Linthicum and El-Rahman, 1987; Schuknecht, 1974). Endolymphatic hydrops may develop (Linthicum and El-Rahman, 1987; Schuknecht, 1974).

With currently standard diagnostic techniques, a presumptive diagnosis is made by detecting a positive serum MHA-TP (or FTA-ABS), usually in the absence of positive tests on cerebrospinal fluid. Positive MHA-TP and FTA-ABS indicate previous exposure to the

treponema, but current activity of the disease is relegated to a clinical decision, since laboratory tests are not conclusive. False-positive treponemal and nontreponemal tests, especially with autoimmune disease, complicate clinical decision making. Recently, however, definitive diagnosis of active syphilitic infection has been made possible by the use of electrophoretic transfer blotting (Western blot technique) to identify serum and CSF IgG and IgM antibodies specific for multiple antigens extracted from *Treponema pallidum.* Identification of only IgG antibodies is consistent with inactive infection, while the finding of IgM antibodies to multiple antigens is indicative of active infection (Birdsall et al, 1990). With corroboration of these findings and more widespread availability of testing, the clinical diagnosis and managament of luetic SNHL is likely to be much more straightforward.

ECoG may show an enhanced SP/AP, suggestive of hydrops (Ramsden et al, 1977). As reported by Zoller et al (1979), therapy consists of at least 3 months of high-dose steroids (80 mg prednisone every other morning, orally) and penicillin (2.4 MU benzathine penicillin IM); many alternatives exist (Darmstadt and Harris, 1989).

Reported rates of hearing improvement vary from 15% to 80% (Darmstadt and Harris, 1989) as measured by pure-tone thresholds and/or speech discrimination testing (Zoller et al, 1979).

Chronic otitis media

Progressive SNHL, generally unilateral or asymmetric, may complicate chronic suppurative otitis media (COM). Such an association is well accepted for tympanogenic suppurative labyrinthitis and labyrinthine fistulization by chronic inflammation or cholesteatoma (Kobayashi et al, 1989; Schuknecht, 1974). The hypothesis that bacterial toxins can penetrate the round window membrane and cause SNHL in otherwise uncomplicated COM has logical appeal, but there is relatively scanty supportive evidence (Paparella et al, 1948).

Clinically, elevated bone conduction thresholds, particularly for the high frequencies, have been documented in prospective and retrospective studies of patients with COM (Levine et al, 1989; Paparella et al, 1984; Schuknecht, 1974; Walby et al, 1983); confounding factors such as presbycusis, noise exposure, window occlusion by inflamed middle ear mucosa, ototoxic drug exposure, and hereditary factors variably have been taken into consideration (Walby et al, 1983).

Other investigators have found no evidence for increased bone conduction thresholds in COM without cholesteatoma, once the Carhart effect was taken into account (Browning and Gatehouse, 1989).

Investigators have been unable to delineate the histopathologic correlates of the observed SNHL. Walby and associates (1983), in a light microscopic examination of temporal bones from patients with COM complicated only by SNHL, were unable to find any cochlear alterations that could explain the observed SNHL; they hypothesized that either abnormalities beyind the resolution of the light microscope, such as damaged cilia, or perhaps changes in the mechanics of sound transmission, could be considered.

Acute otitis media

Acute purulent otitis media (AOM) has been thought to cause temporary and permanent SNHL in the same way as chronic otitis media (Paparella et al, 1984). Experimental AOM in a chinchilla model (Morizono et al, 1985) has been associated with SNHL, as measured by compound action potentials, but no histopathologic correlate could be found by light microscopic examination. Studies of round window membrane permeability in the guinea pig with experimental *Pseudomonas* otitis media found that, although there was an initial increase in permeability to horseradish peroxidase 1 week after inoculation, by 4 weeks there was a decrease in permeability, which corresponded to the light microscopic findings of round window membrane thickening and the accumulation of purulent debris and granulation tissue at the round window membrane or niche (Kim et al, 1990). The relevance of these studies to the human remain undetermined.

Rocky Mountain spotted fever

Rocky Mountain spotted fever (RMSF) is an infection caused by *Rickettsia rickettsii,* and like Lyme disease, it is transmitted by a tick vector; fever, headache, and (petechial) rash are thought of as the "classic symptoms" at onset (Dumler, 1991). The systemic vasculitis that is associated with the infection is responsible for serious complications such as brainstem or multifocal encephalitis, nephritis, and hepatic abnormalities (Steinfeld et al, 1988). The disease has a mortality rate of 4% (Dumler, 1991).

Rapidly progressive SNHL has been reported in association with RMSF, although the site of the lesion has not been clearly established; one report suggests that vasculitis of the brain stem might explain the observed SNHL and dizziness (Steinfeld et al, 1988).

The diagnosis is based on clinical suspicions confirmed by rickettsial titers, and therapy consists of the use of broad-spectrum antibiotics (Steinfeld et al, 1988).

Lyme disease

Lyme disease is caused by the tick-borne spirochete *Borrelia burgdorferi;* both feral and domestic animals may harbor infected ticks. The protean manifestations of the full-blown disease include dermatologic, neurologic, musculo-skeletal, and cardiac abnormalities (Caruso, 1985).

Although facial palsy or paralysis has been the focus of otolaryngologic concern with Lyme disease (Glasscock et al, 1985), recent interest has focused on the possibility of SNHL related to Lyme disease (Hanner et al, 1989; Lesser et al, 1990). Of 98 patients with sudden, progressive, or fluctuating unilateral SNHL, 17 showed evidence of borreliosis by indirect immunofluorescence. Although low-frequency SNHL predominated, mid-frequency, flat, and high-frequency SNHLs were also seen (Hanner et al, 1989); acute facial palsy was variably associated. Of the 17 serologically positive patients who were treated with intravenous benzyl penicillin (30 mg/kg, three times a day, 10 days), five reported improvement in hearing (Hanner et al, 1989). Although more rigorous clinical study must be done, Lyme disease seems to be an important diagnostic consideration in otherwise unexplained SNHL, especially if the loss is unilateral and associated with vertigo and facial palsy.

Diseases of Bone

Otosclerosis

The relationship of progressive SNHL to cochlear involvement with otosclerosis, in the absence of footplate fixation, remains a matter of debate. Endosteal involvement by otosclerotic foci has been theorized to lead to SNHL by means of peptide (Sziklai et al, 1985), lysozymal enzyme, or toxic substance release; vascular alterations; or relaxation of the basilar membrane secondary to distortion of the cochlear capsule (Balle and Linthicum, 1984; Elonka and Applebaum, 1981). However, with rare exception (Balle and Linthicum, 1984), histologic studies have failed to associate SNHL with endosteal involvement, save for the most severely affected cochleas (Elonka and Applebaum, 1981; Hinojosa and Marion, 1987; Kwok and Nadol, 1989). The utility of medical therapy (fluoride, calcium, vitamin D) in arresting the progressive SNHL is also contested (Balle and Linthicum, 1984; Snow, 1985).

Paget's disease

Paget's disease is a disorder of bony resorption and deposition of indeterminant etiology, which may involve the skull and temporal bone; it is particularly common in the older population, affecting 11% of those over 80 years of age (Lando et al, 1988). Approximately 50% of patients with pagetic involvement of the temporal bone have hearing losses greater than those expected for age (Khetarpal and Schuknecht, 1990). In descending order of frequency, the progressive hearing loss seen in Paget's disease may be mixed, sensorineural, or rarely conductive (Khetarpal and Schuknecht, 1990); the hearing loss may simulate presbycusis by being bilaterally symmetric (Khetarpal and Schuknecht, 1990), or it may be unilateral or bilaterally asymmetric (Lando et al, 1988).

Even recent investigators have been frustrated in attempts to determine the histopathologic correlate(s) of the SNHL of Paget's disease (Khetarpal and Schuknecht, 1990). Four phases of pagetic change have been identified in the human temporal bone - the osteolytic, the mixed (bone formation and resorption), the osteoblastic ("burnt out"), and the lamellar remodeled. Periosteal bone is first involved, with enchondral and endosteal extension varying with the agressiveness of the disease (Khetarpal and Schuknecht, 1990). No correlate for the SNHL associated with Paget's disease could be found on light microscopic examination of 26 pagetic temporal bones; there was no evidence of nerve trunk compression in the internal auditory canal nor any pathologic changes in the cochleas that could be directly attributed to Paget's disease, even when pagetic lesions involved the endosteal layer of the otic capsule (Khetarpal and Schuknecht, 1990). Khetarpal and Schuknecht (1990) hypothesized that "changes in the density, mass, and form" of the middle and inner ears served to dampen their "finely tuned motion mechanics" and thus caused the observed hearing losses.

The treatment of Paget's disease consists of calcitonin and etidronate disodium (Lando et al, 1988); some case reports and retrospective reviews suggest that such therapy may be effective in halting, or even reversing, the SNHL of Paget's disease (El Sammaa et al, 1986; Lando et al, 1988).

Autoimmune Disorders

Autoimmune disorders can be associated with a variety of patterns and presentations of SNHL (Bowman et al, 1986; Harris and Sharp, 1990; Hughes et al, 1988; McCabe, 1979). The autoimmune entities that have been associated with SNHL include Cogan's disease (McDonald et al, 1985; Rarey, Bicknell, and Davis, 1986), systemic lupus (Bowman et al, 1986; MacFadyen et al, 1987), polyarteritis nodosa (Vathenen et al, 1988; Wolf et al, 1987), relapsing polychondritis (Schuknecht, 1974), Wegener's granulomatosis (McCaffrey et al, 1980; Murty and Birchall, 1989), Takayasu's disease (Siglock and Brookler, 1987), temporal arteritis (Kramer et al, 1988), and scleroderma (Abou-Taleb and Linthicum, 1987). Vertigo and facial nerve paralysis may also occur (Hughes et al, 1988; Vathenen et al, 1988).

Only a few reports describe the associated temporal bone histopathologic findings, which vary and include endolymphatic hydrops and degeneration of the organ of Corti and cochlear neurons with new bone formation in the basal scala tympani and posterior semicircular canal in Cogan's disease (Rarey et al, 1986; Schuknecht, 1974); granulation tissue invasion of the cochlea in Wegener's granulomatosis (Schuknecht, 1974); perisaccular fibrosis and loss of outer hair cells and cochlear neurons in scleroderma (Abou-Taleb and Linthicum, 1987); and immune vasculitis in polyarteritis nodosa (Gussen, 1977; Jenkins et al, 1981).

The SNHL variably improves with steroids alone (Kramer et al, 1988; McCabe, 1989; Siglock and Brookler, 1987) or in combination with cytotoxic therapy (About-Taleb and Linthicum, 1987; Murty and Birchall, 1989; Vathenen et al, 1988).

Presbycusis

Presbycusis is the term used to describe the bilaterally symmetric deterioration of auditory function that appears to have no basis other than the general aging process of the cellular elements of the cochlea. The extent to which this SNHL reflects a genetically programmed deterioration as opposed to a manifestation of the accumulated insults of a lifetime of noise exposure, disease, and/or toxic exposure continues to be debated (Gates et al, 1989).

Schuknecht (1974) has devised a classification of presbycusis that correlates histopathologic observation with audiometric findings; in the simplest form, the classification distinguishes progressive hair cell loss (sensory presbycusis (Schuknecht, 1974), cochlear neuronal degeneration out of proportion to hair cell loss (neural presbycusis) (Pauler et al, 1986; Schuknecht, 1974), and atrophy of the stria vascularis (metabolic presbycusis) (Pauler et al, 1988), and acknowledges a presbycusis for which no cochlear abnormality can be found (cochlear conductive) (Ramadan and Schuknecht, 1989; Schuknecht, 1974). A disturbance of cochlear motion mechanics, particularly with respect to the basilar membrane, is hypothesized to account for the SNHL (Ramadan and Schuknecht, 1989; Schuknecht, 1974). Although pure forms of each type of degeneration can be found, combinations of the types can also be seen.

Presbycusis manifests during middle age and beyond, but the causative degenerative processes may begin as early as the second or third decades of life (Ramadan and Schuknecht, 1989). Genetic factors may well play a role in cochlear neuronal degeneration (Schuknecht, 1974) and strial atrophy (Pauler et al, 1988).

In sensory and metabolic presbycusis, speech discrimination scores are generally serviceable; with neural presbycusis, the discrimination may be severely affected (Schuknecht, 1974). In cochlear conductive presbycusis, the speech discrimination varies inversely with the downward slope of the pure tone thresholds (Schuknecht, 1974). The utility of amplification for rehabilitation is dictated by the speech discrimination scores in particular.

Chronic Renal Failure

Chronic renal failure, particularly in association with treatment by hemodialysis, has been associated with progressive, fluctuating, and sudden SNHL, which can affect either one or both ears (Bergstrom et al, 1973; Johnson and Mathog, 1976; Rizvi and Holmes, 1980). Osmotic shifts, hypotension, embolism, and metabolic imbalance with resulting electrolyte abnormalities, and circulating uremic toxins have been thought to underlie the SNHL. Pathologic studies have reported variable findings without a consistent histopathologic correlate (Schuknecht, 1973); confounding variables include ototoxic drug treatment, intercurrent disease, noise exposure, aging, and hereditary factors (Schuknecht, 1973; Wigand, 1976). Thus, although suggestions exist that there is an excess SNHL in patients with chronic renal failure, it has been difficult to establish that the hearing loss relates solely to the renal dysfunction (Johnson and Mathog, 1976).

Endocrine Disorders

Diabetes mellitus

The SNHL typically thought to be associated with diabetes mellitus is bilaterally progressive, of gradual onset, and particularly affects the high frequencies and older patients (Kurien et al, 1989; Taylor and Irwin, 1978); thus, it is similar to presbycusis but "greater than expected for age" (Taylor and Irwin, 1978). Lack of appropriate control groups and small numbers have weakened the data from studies purporting to show a relationship between diabetes mellitus and SNHL (Axelsson et al, 1978; Kurien et al, 1989).

Diabetic microangiopathy has been theorized to be causally related to the SNHL. On histopathologic examination, all aspects of the auditory pathways from peripheral to central auditory structures have been found to be altered (Axelsson et al, 1978).

Disagreement does exist concerning the existence of an SNHL related to diabetes mellitus alone. Studies have failed to demonstrate a significant excess SNHL in patients with diabetes mellitus (Axelsson et al, 1978; Miller et al, 1983); for example, Miller and associates (1983) failed to find any significant difference in SNHL with standard audiometric testing in patients with diabetic retinopathy as contrasted to a stratified control group.

Hypothyroidism

Hypothyroidism has been associated with SNHL in approximately 25% of adults and in up to 50% of children with cretinism (Vanasse et al, 1989). The site of the lesion has been thought to be in the cochlea. Other studies have found no significant SNHL in adult hypothyroidism (Parving et al, 1983). Similarly, disparate findings regarding improvement in hearing and ABR results following thyroid hormone replacement therapy have been reported

(Anand et al, 1989; Vanasse et al, 1989).

Hypoparathyroidism

Investigators have studied the possible relationship between adult hypoparathyroidism and SNHL (Ikeda et al, 1987b). Interest has been stimulated by reports of the importance of calcium in acoustic transduction (Ikeda et al, 1989) and synaptic transmission in the auditory nerve (Ikeda et al, 1987a), as well as by reports of SNHL in man related to alterations in vitamin D and calcium metabolism (Brookes, 1983).

Efforts to establish a clinical correlation of calcium and vitamin D metabolism aberrations to human SNHL have been stymied by small numbers of patients and lack of appropriate control groups (Ikeda et al, 1987b). The situation is further complicated because, although the cochlea appears to be the site of the hearing loss, the audiometric threshold patterns are quite variable (Ikeda et al, 1987b).

Metabolic Disorders

Hyperlipoproteinemia

Hyperlipoproteinemia has been associated with bilaterally progressive SNHL that may be reversible if dietary intake is altered (Rosen et al, 1970). Rosen and associates (1970) investigated this association in a crossover dietary manipulation study performed in similar patient population groups in two Finnish mental hospitals; after 5 years, the patient group that was fed the normal Finnish high-saturated fat diet showed significantly poorer hearing thresholds than did the population fed a diet high in polyunsaturated fats. At the end of the 5-year crossover limb, the previously poorer-hearing group showed improved hearing, while the initially better-hearing group manifested a deterioration in thresholds.

More recent studies in the human have been plagued by lack of control populations and inability to document improvement in hearing after dietary alteration. However, in the rat model, Pillsbury (1986) found a relationship between hyperlipoproteinemia and SNHL, but only when influenced by concomitant noise exposure and hypertension.

Hereditary Disorders

This section will highlight SNHLs that have been recognized to progress throughout life, especially those disorders in which the tendency for progression has been relatively recently described.

Usher's syndrome (USH) is characterized by congenital bilateral SNHL and retinitis pigmentosa, occasionally associated with vertigo; it affects 3 of 100.000 in the general population (Karjalainen et al, 1989), and it is believed to be inherited as an autosomal recessive trait. There are two clinically distinguishable types of Usher's syndrome, USHI and USHII. Patients with USHI have congenital bilateral profound hearing loss and absent vestibular function, while individuals with USHII have moderate hearing loss and normal vestibular function. Patients with USHII can occasionally exhibit a bilaterally progressive SNHL.

Waardenburg's syndrome is transmitted in an autosomal dominant mode of inheritance and is typified by a white forelock, dystopia canthorum, heterochromia iridis, and distinctive facial features; in addition, in approximately 20% of cases, there is evidence of cochlear involvement in the form of a bilaterally-symmetric SNHL (Hildesheimer et al, 1989; Konigsmark and Gorlin, 1976). Waardenburg's syndrome has been classified into types I and II. Type I represents the more "classic" form of the disorder, which includes dystopia canthorum. Type II, which is rare, does not include dystopia canthorum and manifests a greater incidence (51%) of cochlear involvement in the form of a progressive, bilateral, moderately severe SNHL (Hildesheimer et al, 1989).

Histopathologically, Waardenburg's syndrome has been shown to be associated with atrophy of the organ of Corti, cochlear neurons, and stria vascularis (Schuknecht, 1974).

Osteopetrosis (Albers-Schönberg disease, marble bone disease) is a metabolic bone disease that characteristically shows decreased bone resorption caused by osteoclast dysfunction (Schuknecht, 1974) resulting in a diffuse, symmetric osteosclerosis. Both a malignant, recessively inherited form and a benign, dominantly-transmitted form occur (Schuknecht, 1974). Progressive osteosclerosis causes narrowing of the foramina of the skull (Bollerslev et al, 1988). Two subtypes of the autosomal dominant form have been distinguished by radiologic examination; type I is characterized by marked sclerosis, especially of the cranial vault, while type II is typified by lesser sclerosis, which predominantly affects the skull base (Bollerslev et al, 1988). The acoustic nerve is affected in both types I and II, but more frequently in type I; a significant narrowing of the internal auditory canal has been noted in only type I (Bollerslev et al, 1988).

Multiple forms of autosomal-dominant SNHLs have been described based on the frequencies most severely or initially affected (Konigsmark and Gorlin, 1976). WIthin dominant high-frequency SNHLs several subtypes can be distinguished by the onset of the hearing loss (Konigsmark and Gorlin, 1976) or the audiometric pattern, especially the steepness of the high-frequency slope (Higashi, 1988). The SNHLs are bilaterally symmetric and manifest the most severely reduced thresholds for the high frequencies, in some cases mimicking presbycusis.

Disorders of Unknown Etiology

Ménière's disease

Ménière's disease is of unknown etiology and is characterized by the onset of fluctuating SNHL, aural fullness, tinnitus, and episodic vertigo in a previously healthy ear (Schuknecht and Gulya, 1983); endolymphatic hydrops is the main histopathologic correlate (Hallpike and Cairns, 1938; Schuknecht, 1974). Initially, the disorder affects cochlear function unilaterally, but in up to 50% of the cases (Balkany et al, 1980) the disease may eventually manifest bilaterally. Although salt restriction therapy and diuretics have been advocated in the management of Ménière's disease, there is no definite treatment or cure. Surgical therapies are targeted exclusively toward the elimination of vertigo, with or without attempts to maintain auditory thresholds.

Sarcoidosis

Sarcoidosis is a granulomatous disease of unknown etiology, typified by noncaseating granulomata. Neurologic manifestations develop in 5% of patients (Delaney, 1977) and include hearing loss, which is predominantly sensorineural, and vertigo (Babin et al, 1984). The SNHL may be fluctuant, and both central and peripheral sites of lesion have been suggested (Babin et al, 1984). The primary therapy of sarcoidosis involves corticosteroid administration.

Pathologic findings in central nervous system sarcoidosis involve an obliterative granulomatous leptomeningitis. Histopathologic examination of the temporal bone has shown extensive degeneration of the organ of Corti and stria vascularis, round cell infiltration in the modiolus, and intraneural perivascular infiltratin of the nerve trunks in the internal auditory canal (Babin et al, 1984).