Selected complex auditory disorders

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Abstract—This article provides an overview of four auditory disorders relevant generally to adults and especially to veterans. The disorders are noise-induced hearing loss, idiopathic sudden sensorineural hearing loss, otosclerosis, and Ménière’s disease. Sensorineural hearing loss characterizes each, but additional aspects vary with each of the four conditions. This article describes the conditions, discusses their diagnoses and treatments, and outlines current and suggested rehabilitation. The emphasis is on recent advances, some of which await confirmation prior to possible acceptance as standard practice.

Key words: audioligic rehabilitation, diagnosis, hearing loss, idiopathic, Ménière’s disease, noise-induced, otosclerosis, sensorineural, serial audiometry, spontaneous recovery, treatment.

INTRODUCTION

Audiologists face a number of disorders that demand their full expertise to diagnose and manage. Four are considered here: noise-induced hearing loss (NIHL), idiopathic sudden sensorineural hearing loss (ISSNHL), otosclerosis, and Ménière’s disease. All involve sensorineural hearing loss in combination with other significant symptoms. The common threads among the disorders in this article are recent advances in audioligic rehabilitation. New diagnostic and treatment techniques are the focus.

In each of the four sections that follow, we briefly describe the disorder, discuss its diagnosis, and outline its management. In each category, we highlight emerging ideas, some of which may yet be in the process of confirmation and/or adoption.
Research has shown that high-intensity noise destroys hair cells in the cochlea and disrupts connections to higher auditory centers, resulting in temporary and often irreversible hearing loss. Hair cells can recover within 48 hours (the usual duration of temporary hearing loss) [5]. Permanent hearing loss occurs when noise levels overwhelm this self-repairing capability and with constant or cumulative exposure to noise levels exceeding 75 to 80 dBA over long periods. The current standard with which employers in the United States must comply can be found in the Occupational Safety and Health Administration Hearing Conservation Amendment [6].

**Diagnosis**

Factors determining damage-risk criteria for NIHL are sound-pressure level, spectral characteristics, duration of exposure, and individual susceptibility. Additionally, exposure to solvents and heavy metals may act synergistically with noise to damage hearing and may cause hearing loss independent of noise exposure [7–8].

Distinguishing acoustic trauma from the far more prevalent NIHL that occurs over time is essential. Acoustic trauma following exposure to a single exposure to a noise of 120 dB or more is dramatic. Although hearing sensitivity sometimes returns after a period of hours or days, small amounts of permanent damage often occur following acoustic trauma and may increase susceptibility to future exposures. Organic changes resulting from chronic exposure to moderate noise levels are less definitive; there are signs of metabolic exhaustion—hair cell deterioration and reduced energy carried by the cochlear fluids. As the degree of the stimulation increases, the outer hair cells become distorted and degenerate [9].

Otoacoustic emissions (OAEs)—an electroacoustic technique for evaluating cochlear function and, indirectly, the status of the middle ear—cannot predict when a temporary threshold shift becomes permanent, but this audiological test may have value in determining who is susceptible to NIHL. A patient’s OAE following noise exposure might show early changes in outer hair cells, suggesting greater risk for NIHL.

**Management**

Most NIHL can be prevented with reasonable precautions. Limiting noise exposure can reduce the number of persons affected, and a vigorous hearing-conservation program can help reduce NIHL [1]. Although federal agencies have set standards for industry to follow, some situations cannot be managed in that way. Military personnel in war zones, and occasionally in training exercises, may experience sudden, extreme noise levels.

When excessive noise cannot be avoided, personal hearing-protective devices can attenuate or eliminate its damage. Its effects can be reduced by the protection afforded by earplugs, muff, etc., as part of a comprehensive program of occupational hearing conservation [1–2,6]. Drugs taken prior to noise exposure also may reduce NIHL. When a person is exposed to high-intensity noise, circulation to the cochlea drops. Antioxidants that detoxify free radicals or agents that increase blood flow to the cochlea may protect or rescue hair cells. Some drugs approaching clinical trials (e.g., D-Met and acetyl-L-carnitine) appear effective in animal studies when administered prior to noise exposure [10]. Although otoprotective agents are under active investigation to prevent NIHL, some may have their own toxicities [11]. While several hold promise, no drugs are currently approved by the Food and Drug Administration (FDA). Until the FDA approves a drug for prevention or recovery of hair cells, persons at risk should discuss drugs and dietary supplements with their health professionals, follow a healthy lifestyle, and adopt a well-balanced diet [12].

Ultimately, a preferred solution to NIHL rests with engineering methods to eliminate or reduce noise at its source. A program of public awareness, dubbed WISE EARS® directs attention to the wide variety of noises and the cumulative impact they can have [13].

**Summary**

Despite being preventable, NIHL remains highly prevalent. Avoiding noise exposure, consistently wearing personal hearing-protective devices and, in the future, using medications and/or dietary supplements may reduce the widespread prevalence of this disorder.

**IDIOPATHIC SUDDEN SENSORINEURAL HEARING LOSS**

Among the difficult clinical conundrums facing audiological and otologic rehabilitation, ISSNHL ranks high. “Idiopathic” refers to its unknown etiology, a feature of about one in three instances of sudden onset hearing loss. Since partial or complete spontaneous recovery occurs in approximately half or more of the cases, obviating the need for treatment, estimates of the incidence of this
condition probably underestimate its extent [14]. While imprecise, the estimated incidence of ISSNHL in the United States lies between 5 and 25 per 100,000 [15].

Although ISSNHL largely affects adults, instances of it in children have been noted [16–17]. The average age at onset is between 40 and 50 years [18]. Typically, unilateral instances are about 50 times greater than bilateral [19].

Diagnosis

Since “idiopathic” in its name denies identifying a cause, the term “diagnosis” may seem inappropriate. However, while initially indeterminate, an etiology has been established in about two-thirds of reported cases, following a careful, detailed case history and appropriate diagnostic procedures [20].

Spontaneous recovery from ISSNHL usually occurs within the first 30 days after onset. About half the cases regain all or almost all their hearing within that time [21]. The proportion of recovery declines through the next five months, after which little possibility remains for spontaneous recovery.

Recovery may be related to the degree of initial hearing loss. A rising or mid-frequency audiometric curve predicts spontaneous recovery more frequently than a sloping or flat configuration [18]. The possibility for complete remission is best when the loss is less than 70 dB hearing level (HL) [22].

Serial audiometry, with pure-tone and speech-recognition testing, is recommended to detect changes in hearing. Without appearing overly optimistic, audiologists should advise patients that some recovery not experienced early might occur over time, although slowly.

Management

The longer patients wait to seek treatment, the poorer the prognosis. Prompt attention to ISSNHL, then, is essential to obtain maximum benefit from a management program.

Early treatment should be directed to relieve two symptoms that usually accompany ISSNHL: tinnitus in about 70 percent and vertigo in about 50 percent of cases [23]. Some patients regard these symptoms as equal to, if not worse than, the hearing loss. Vertigo frequently occurs concurrently with the ISSNHL, but in some cases may follow hearing loss. Its presence indicates that spontaneous recovery is less likely, although many exceptions occur.

The most common therapeutic measure is steroid therapy. When short-term oral steroids were administered to 266 ISSNHL patients and compared with 52 patients who either declined treatment or had medical contraindications for steroid treatment, 40 percent of the steroid group did not recover any hearing or it worsened over the treatment period and losses became permanent. Of the remaining 60 percent of steroid-treated patients, hearing improved an average of 29 dB HL compared with 11 dB HL for the untreated patients. Both steroid and nonsteroid groups had improved performance in their word-recognition scores following prompt treatment, although a significantly larger portion improved among those receiving steroids [24]. A double-blind, randomized, placebo-controlled prospective study of 28 patients found that adding oral magnesium to the steroid treatment enhanced improvement [25].

Vasodilators have been prescribed with the rationale based on a theory of reduced cochlear blood flow. Following a review of the charts of 41 patients who had been treated with steroids, histamines, or Carbogen, researchers concluded [26], “There was no correlation of preexisting signs, symptoms, or findings with hearing recovery.” Similar conclusions appear throughout the ISSNHL literature [22,27–28].

The high rate of spontaneous recovery complicates research on this disorder. To be judged successful, a treatment must succeed more often than the spontaneous-recovery rate. Delaying rehabilitation intervention 60 to 90 days is justified by the odds favoring the return of normal to near-normal hearing. To wait longer, however, is inadvisable, in view of the handicap imposed by the hearing loss.

Once an otologist can reasonably concede that full recovery will not occur, patients should be offered audiological rehabilitation. A first step is counseling to avoid excessive noise, seek prompt treatment of middle-ear infections, obtain suitable amplification, and optimize conditions that improve sensory functions. The emotional consequences of permanent damage also should be addressed [29].

The decision to recommend hearing aids may depend on patients’ resistance and economic factors. Patients may resist suggestions to try a hearing aid, because the suggestion may indicate to them that the hearing loss is permanent. They will understand if the audiologist explains that use of a hearing aid is reversible and that it can be adjusted or discarded if and when hearing improves. Uppermost should be the possibilities for delayed spontaneous recovery.
Should the affected or unaffected ear receive the aid? If aidable residual hearing is detected in the affected ear, the audiologist may fit a hearing aid for that ear. If the affected ear is unaidable, a contralateral routing of signals (CROS) or bone-anchored cochlear stimulator (BAHA) may be selected. Recent studies report greater satisfaction and improved communication with BAHA over CROS [30]. Preferable to either the CROS or BAHA may be use of a frequency-modulated (FM) system broadcasting to the unaffected ear, since it can deliver an intense, relatively undistorted signal with an improved signal-to-noise ratio (SNR) [31]. The BAHA also can be connected to an FM system for an improved SNR.

Summary

ISSNHL is frequently a transitory condition. When spontaneous recovery does not occur and when available treatment options are unsatisfactory, audiologic rehabilitation that includes counseling and amplification should be offered the patient. Among amplification choices, either BAHA or FM or both appear to be most suitable for unilateral cases.

OTOSCLEROSIS

Otosclerosis is a focal disease of the otic capsule, characterized by excessive resorption of bone. New bone formation is soft (otospongiosis) and hypervascular, gradually changing into a dense sclerotic mass [32]. Initially affecting the ossicular chain, this genetic condition can invade the inner ear, progressing from a conductive to a mixed to a primarily sensorineural loss of hearing. In addition to the deterioration of hearing, tinnitus usually accompanies the hearing loss and, when the inner ear is involved, infrequent dizziness and imbalance also may occur.

Diagnosis

Otosclerosis is typically inherited (autosomal dominant transmission, with variable penetrance). Onset of the hearing loss and related symptoms usually arise between 15 and 45 years in 90 percent of cases, although earlier and somewhat later occurrences have been noted. It is bilateral in 90 percent of females and 80 percent of males [33].

Otosclerosis is a progressive disease with an unpredictable course. Hearing loss may show worsening during periods of hormonal-endocrine changes, e.g., during pregnancy and menopause. Although it is sometimes first noticed following pregnancy, the linkage between the disease and pregnancy may be casual, not causal; i.e., some women become pregnant during the same period as the disease’s onset. Otosclerosis commonly affects Caucasians and Asians and less frequently persons of African descent. While the differential incidence has been established, the reason for it remains unexplained [34].

Because the hearing loss develops slowly, patients are often unaware of its initial onset. They may experience tinnitus long before they recognize the lessened hearing ability. Tinnitus is present in a majority of otosclerotic patients, the reported occurrence of which varies between 56 and 79 percent [35–36]. Once patients are diagnosed, annual audiological examinations are recommended to check the disease’s course and to provide benchmarks for treatment evaluation.

Management

The conductive form has been successfully treated surgically, usually by removing involved portions of the stapes (stapedectomy) and replacing them with titanium or other suitable material. This procedure, originally introduced over four decades ago, has several versions; most have proved successful in restoring hearing when the disease is confined to the stapedial footplate and cochlear function is normal [37–38].

Fluoride to increase hardening of bones has a varied history of success in slowing otosclerosis [39]. This treatment has fairly innocuous side effects, so it can be prescribed with impunity. It should be understood, however, that otosclerosis presently has no cure.

Once there is significant inner-ear involvement, providing amplification is the most common approach to rehabilitation of otosclerosis. Amplification may vary in a patient’s lifetime from completely in-the-canal hearing aids to powerful behind-the-ear instrumentation. The latter’s electroacoustic characteristics equal or exceed the far-less cosmetically acceptable body-worn aids and have essentially replaced them. Some patients have or continue to use conventional bone-conduction aids. Some with far-advanced forms of the disease—in which the joint between the stapedial footplate and the oval window is obliterated—become candidates for aggressive drill-out stapes procedures that allow them to use powerful amplification. The procedure uses microdrills to thin out the footplate and then create an opening for a prosthesis. Others become candidates for cochlear implants when their hearing losses become profound or total.
Patients with solely middle-ear pathology are relatively easy to fit, since they are free of sensorineural distortions and only require sufficient gain and output to overcome their obstructive lesion. Hearing aids for these patients do not have undesirable complications and provide improved auditory function that is immediate and dramatic.

The case for amplification versus surgery becomes more complex if the patient presents with a mixed hearing loss. When it is of the vertical type—conductive below 1,000 Hz and sensorineural above—surgery that reduces or eliminates the low-frequency component will not provide serviceable hearing and will leave the patient a poorer candidate for amplification. Postoperatively, such patients will experience a drop in ability to discriminate speech in noise, a narrowed dynamic range, and the absence of a “cushioning” or dampening effect provided by the conductive component. This component is audiologically manifested by an air-bone gap contributing to better word recognition for the same air-conduction thresholds. A mixed hearing loss tends to improve suprathreshold tolerance, and the conductive component tends to “flatten” the audiometric configuration, both of which contribute to easier adjustment to amplification than when the loss is purely sensorineural.

In cases with mixed horizontal hearing loss—bone conduction depressed by 30 to 35 dB across the frequency range but significantly better than air conduction—surgical intervention has less deleterious effect on speech recognition, but the patient still lacks serviceable hearing in many listening situations and requires amplification.

For patients with severe mixed hearing loss in the range of 70 to 90 HL with large conductive components, surgery can reduce the conductive component to allow the use of more cosmetically acceptable in-the-ear aids. Patients should be advised that even when surgery is successful, they would still need amplification.

Summary

Otosclerosis is an inherited disease that may progress from mild to total hearing loss. Diagnosis seldom presents unusual problems, but a thorough audiological assessment is essential to selection of treatment at various stages of the disease. Surgery has a good prognosis in the early stages, but amplification is often needed when both the middle and inner ears are involved. Selection of the type of instrumentation will vary in accordance with the progression of the disease and patient preferences.

MÉNIÈRE’S DISEASE

Ménière’s disease consists of a combination of hearing loss, vertigo, and tinnitus and may also be accompanied by a sensation of fullness in the affected ear. First described by Prosper Ménière in 1861, it is now considered a disease of the membranous inner ear and attributable to excessive endolymphatic fluid causing Reissner’s membrane to become distended or to an injury of the fluid-absorption system [14]. It has been postulated that Reissner’s membrane may perforate during the acute attack and reattach itself between attacks. Because of the excess production or poor absorption of fluid, it is often called endolymphatic hydrops. The cause of the disease remains unknown, but emotional and/or physical stresses may trigger individual attacks [40–41].

The hearing loss may vary from mild to profound, although as it progresses, it usually becomes severe, ranging from 70 to 90 dB. The tinnitus is of a roaring character with pronounced low-frequency components. These symptoms occur paroxysmally, with their duration varying from 20 minutes to several hours or days. Patients usually have warnings of the impending vertigo enabling them to cease dangerous activities (such as driving) before its onset.

In about 70 to 80 percent of cases, the hearing loss occurs unilaterally. However, involvement of the unaffected ear often occurs as the disease progresses, reaching about 40 percent after 15 years [40]. Similarly, the hearing loss may become severe as the disease process continues.

Ménière’s disease has been estimated to be the third most common inner-ear disorder, after presbycusis and NIHL. Its occurrence per 100,000 has been variously estimated to be between 50 to 150 [41] and 218 [40].

Diagnosis

Although it has been attributed to many etiologies—food allergies, endocrine insufficiencies, vascular disease, syphilis, viral infection, and genetic factors—it is usually regarded as an idiopathic disorder whose precise etiology is difficult to determine [14]. Since Ménière’s disease presents as a unilateral, primarily low-frequency hearing loss in the early stages, lesions of the auditory nerve must be ruled out. Auditory brainstem and magnetic resonance imaging with contrast are necessary to rule out space-occupying lesions.
The characteristic audiometric configuration is a rising curve; i.e., as the frequency increases, the hearing loss decreases. This contour has also been called a “reverse slope,” since high-frequency losses are by far the most frequent. A much lower-than-expected word-recognition score often accompanies the pure-tone loss. Although high-frequency sensitivity tends to be normal in the early stages, it becomes involved as the disease progresses, leaving the patient with a “flat” audiometric configuration.

A complete audiological evaluation of the patient suspected of having Ménière’s should include electrocochleography and electronystagmography. A glycerol test is believed to aid diagnosis; it involves the patient ingesting a solution containing glucose before audiometric testing and comparing the results to the same tests 3 hours after. Improvements of at least 15 dB in one or more frequencies and/or 12 percent or more in word-recognition scores indicate presence of the disease [42]. The electrical responses of the cochlea and the assessment of vestibular function, with both positional and caloric stimulation, also provide important information for diagnosis and monitoring of treatment [43].

Management

The management of Ménière’s remains problematic. This condition does not occupy the same place in the panorama of auditory disorders as those described previously. Its paroxysmal character creates a conundrum for research and treatment. During acute attacks, patients’ markedly reduced ability to discriminate speech and the accompanying vertigo challenge audiologists’ efforts to prescribe amplification and direct audiologic rehabilitation. Between attacks in the early stages, auditory function may be normal or near normal, but the fear of another attack often leaves the patient psychologically compromised. Further complicating management, the hearing loss is accompanied by loudness recruitment, narrowed range of comfortable loudness, and severe acoustic distortion.

Digital amplification should be considered when the hearing loss stabilizes. The flexibility that these circuits allow is an important feature as significant fluctuation of the hearing loss continues.

Initial treatment is conservative, and a variety of medications are prescribed to provide symptomatic relief, e.g., vestibular suppressants and tranquilizers for the vertigo, and diuretics, vasodilators, intravenous histamine, nicotinic acid, Pro-Banthine, Benadryl, and lipoflavonoids for the presumed hydrops. A low-salt diet is often recommended [44].

Surgical treatments may be undertaken for patients whose vertiginous attacks become disabling; these include the endolymphatic shunt [43,45]. A controversial study by Danish surgeons who compared this procedure with a sham operation concluded that [46] “the impact of the various endolymphatic sac shunts upon the symptoms . . . is highly unspecific, and that the 70 percent improvement in both our groups [treated vs. controls] was most likely caused by a placebo effect.” In extreme cases, severing the vestibular portion of the eighth cranial nerve might be an option.

Chemical ablation of hair cells in the vestibular labyrinth is another approach that has been tried. Ninety Ménière’s patients were administered intratympanic injections of gentamicin. The investigators concluded that their results [47] “demonstrate the effectiveness of this treatment modality with very low side effects, and, although our experience is still limited, it allows for expanding the indication on early cases of Ménière’s disease before permanent hearing loss occurs.”

The Meniett device, a portable low-intensity alternating pressure generator, has been tested to alleviate the symptoms of Ménière’s disease. A standard, unsealed tympanostomy tube is inserted into the affected ear, and intermittent overpressures from the device are self-administered three times daily. In a study of 67 patients with unilateral Ménière’s disease assigned randomly to treatment with this device or to a control group, treatment was effective for at least 4 months in controlling severity and number of vertigo attacks [48]. In view of the natural course of this disorder with its spontaneous remissions, further studies should be undertaken.

It has long been assumed that endolymphatic hydrops is the histopathological hallmark of Ménière’s disease [49]. However, Danish scientists made an intriguing discovery, as yet unconfirmed, that the endolymphatic sac produces saccia, a hormone that plays a role in regulating the sodium level in the bloodstream. This finding has led to the possibility of blocking the hormone’s release with medication that would conquer Ménière’s disease [50–51].

Summary

While the cause(s) of Ménière’s disease remain in dispute, its severity usually prompts an immediate response. However, no one medical or surgical treatment has gained wide acceptance among sufferers of this disease.
CONCLUSION

The four conditions reviewed here are all characterized by sensorineural hearing loss. Their management, however, is complicated by additional symptoms or attributes.

NIHL may be transitory or permanent but, most importantly, it is preventable. Improved diagnosis and treatment were discussed; however, engineering to reduce or eliminate offensive noises, along with personal care to avoid noise exposure, offers the best hope of reducing the occurrence of this condition.

Because ISSNHL’s onset is both sudden and unexplained, treatment decisions may be delayed awaiting spontaneous recovery, which occurs in approximately half of reported cases. When hearing is not soon restored, however, instances of ISSHNL deserve aggressiveaudiologic rehabilitation, including counseling and fitting of hearing aids appropriate to the individual.

Otosclerosis is typically inherited and its course is unpredictable, exhibiting significant audiologic symptoms between 15 and 45 years of age. Treatment options include middle-ear surgery, amplification, and medication, the latter prescribed to slow progress of the disease. Tinnitus, which usually accompanies the hearing loss, requires attention, since it may be experienced as a disabling aspect of the disorder. The nature of the condition requires long-term rehabilitation planning and follow-up.

Ménière’s disease combines intermittent hearing loss and vertigo, making it unusually difficult to manage. Its paroxysmal nature challenges the prescription of amplification until the hearing loss stabilizes. Preventing vertiginous attacks occupies the management focus with a variety of surgical procedures, medications, and diets with varying degrees of success. A recent study arouses the possibility that blocking the hormone saccia into the bloodstream may control this disease.

Although these four conditions have long histories and have profited to some degree from research, they deserve more attention not only to further understand their underlying etiologies, but also to promote their optimal management. But presently available audiologic rehabilitation should not be withheld today while awaiting tomorrow’s research.

REFERENCES


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