Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 4: Inner Ear

Chapter 43: Sudden Deafness

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The phylogenetically newer portion of the inner ear, which is known as the pars inferior and includes the cochlea and saccule, is relatively more susceptible to various injurious agents than the pars superior. Present evidence indicates that only a limited number of etiologic factors are responsible for acute injury to the pars inferior that results in sudden deafness. Sudden deafness may be defined as a sensorineural hearing loss that develops over a period of hours or a few days. The severity of the hearing loss may vary from mild to total loss of perception of the most intense sound. The loss of hearing may be permanent, or the hearing may spontaneously return to normal or near normal. Although sudden deafness is usually unilateral, it may be bilateral.

Classification

Rubin (1968) has provided a classification based on the severity and audiometric configuration of the hearing loss. In type I, according to his scheme, the hearing loss is mainly in the lower frequencies, with some elevation of thresholds for 2 to 8 kHz. The speech reception threshold is decreased, and the discrimination is severely depressed. Type II includes more uniform and severe elevation of thresholds for pure tones with a 50 to 60 dB loss for the speech frequencies of 500 Hz, 1 kHz, and 2 kHz. There is a sharp loss above 3 kHz. The speech reception threshold and discrimination scores are consistent with what would be expected with similar pure tone thresholds in more gradually developing forms of hearing loss. Type III is a complete loss of hearing with no measurable discrimination.

Sheehy (1960) evaluated 223 patients and classified then into four groups: low tone, flat, high tone, and total hearing loss. A flat type of hearing loss was found in 41 per cent of his patients. The next most common type was the high tone loss, and this type accounted for 29 per cent. The low tone type made up 17 per cent of the group, and total loss of hearing occurred in 13 per cent.

The classifications of Rubin and of Sheehy are useful primarily because they are simple and easy to keep in mind. Of equal importance to the severity of the hearing loss and its audiometric configuration is the presence or absence of associated vestibular symptoms and signs. Likewise, the rapidity of onset of the hearing loss is important, but a classification including all of these factors becomes so elaborate that its usefulness is limited.

Etiology

The importance of predisposing factors is debatable. Nevertheless, frequently cited predisposing factors include the use of ethanol, the emotional state of the patient, fatigue, diabetes, arteriosclerosis, age of the patient, and pregnancy. The presence of any of these factors and the onset of sudden deafness are most likely coincidental.

Viral Agents

Viral infections are well-documented causes of sudden deafness. The sites of the viral-induced pathologic changes are the cochlea and the components of the eight cranial nerve. Cochlear involvement is best described by the term "viral endolymphatic labyrinthitis", and the neural involvement may be described as "viral neuronitis and ganglionitis". The viruses of mumps, measles, and influenza, as well as the adenoviruses, deserve particular attention in view of their clearly demonstrated etiologic role in sudden deafness of the viral endolymphatic labyrinthitis type. The virus of herpes zoster is the sole agent that has been demonstrated to produce viral neuronitis and ganglionitis. It appears likely that other viral agents will, in time, take on the dubious distinction of being documented causes of sudden deafness. Evidence for viral infections that affect the cochlea will be considered first.

The evidence for the causative role of the mumps virus comes from studies of the pathology of sudden deafness and from immunologic studies. It is generally well known that epidemic parotitis (clinical mumps) may be complicated by sensorineural hearing loss of sudden onset. Some patients have mumps without parotitis. A few patients who have mumps without parotid swelling develop sudden deafness. Lindsay and colleagues (1960) described the pathologic condition of the temporal bone in a patient with bilateral sudden deafness due to well-documented mumps. Degenerative changes consisting of atrophy of the stria vascularis, tectorial membrane, and organ of Corti were found; these progressed from mild changes in the apical turns to total loss of these structures in the lower and basal turns. The spiral ganglion cells tend to be preserved in the upper turns. Saunders and Lippy (1959) demonstrated in a well-controlled study using the reliable and consistent mumps complement fixation test a positive titer in six of nine patients with sudden deafness. None of these patients had parotid swelling, and all clearly recalled childhood mumps infection. In a control group of 370 patients without sudden deafness, only 2.5 per cent had a positive titer for mumps. The other three patients with sudden deafness had similar clinical histories and findings to the six with mumps. Therefore, Saunders and Lippy suggested that the other three patients with sudden deafness may have had some other viral infection for which immunologic tests were not performed. In view of evidence that other viruses produce sudden deafness, the absence of a positive titer in each patient does not detract from the possibility that the mumps virus is responsible for two-thirds of all cases of sudden deafness.

The role of the measles virus in sudden deafness is well known and was documented in 1954 by Lindsay and Hemenway. Likewise, the role of influenza in the etiology of sudden deafness has been established by Van Dishoeck and Bierman (1957). In 1967 Jaffe and Maassab proved the role of adenovirus type 3 in sudden deafness by isolation of the virus and by

demonstrating a diagnostic rise in antibody titer to the same virus. Gregg and Shaeffer (1964) implicated the virus of infectious mononucleosis as a cause of sudden deafness. Van Dishoeck (1963) reported a case of sudden deafness during a well-documented Eaton agent infection. In 1967 Jaffe reported on a patient who developed sudden deafness following a rabies injection.

Less specific as to the type of virus involved but important evidence for the viral etiology of sudden deafness is the close correlation between the pathologic condition of the temporal bone of most cases of sudden deafness and those pathologic changes found in well-documented viral endolymphatic labyrinthitis. Schuknecht et al (1962) described the findings in four cases of sudden sensorineural hearing loss in which the pathologic changes closely resembled what had been found in viral endolymphatic labyrinthitis of mumps, measles, and prenatal rubella. Beal and associates (1967) described the pathologic condition of the temporal bone in two cases of sudden deafness with similar findings.

The presence of active upper respiratory infections in 25 per cent of patients at the time of the onset of sudden deafness has been described in several reports. Considering the possible chance coincidence of upper respiratory infection and sudden deafness, this type of evidence for a viral etiology of sudden deafness is not as convincing as are rises in antibody titer and similarities in pathologic findings in cases of sudden deafness with known viral endolymphatic labyrinthitis.

Viral infections affecting cranial nerve VIII are less common. Bocca and Giordano (1956) reported pure cochlear syndromes, pure vestibular syndromes, and cochleovestibular syndromes of sudden onset on the basis of neuronitis. The cutaneous lesions associated with herpes zoster infections make this form of viral neuronitis and ganglionitis relatively easy to diagnose clinically and to relate etiologically to the virus. Other viruses have not been satisfactorily related etiologically to neuronitis and ganglionitis of cranial nerve VIII.

There are three routes of access of viral particles to the inner ear. Probably the most common is a viremia in the early phase of a viral illness in which virions are deposited within the membranous cochlea. In viral meningoencephalitis, for example, as a complication of measles, the virus may gain access to the inner ear through the perilymphatic space from the subarachnoid space via the cochlear aqueduct. A third possible route, proposed by Lindsay in 1959, is by direct extension from the middle ear to the inner ear in nonsuppurative otitis media during an upper respiratory infection.

Once the virus particles have gained access to the membranous cochlea, replication of the virus leads to rapid pathophysiologic changes that are frequently reversible but that may be extremely destructive and result in permanent loss of hearing. From what is known of the development of viral lesions in other tissues, we may suppose that similar changes occur in the inner ear. Initially the vascular endothelium is invaded, and inclusion bodies may be seen in the endothelial cells in some viral diseases. The capillary endothelium is often swollen and may proliferate. These changes narrow the lumen of the capillaries and may reduce blood flow. Nevertheless, there is generally an overall hyperemia at the site of inflammation, and total

effective blood flow may not be altered. Hemagglutination and sludging of blood may occur when virus particles attach to erythrocytes. Commonly in viral inflammation, extravasation of blood results in micropetechiae. Infiltration of lymphocytes, plasma cells, macrophages, and polymorphonuclear leukocytes occurs, and perivascular spaces fill with edema fluid. Necrosis of neuroepithelium may occur and is indicated by hyperchromatism, fragmentation, and lysis of nuclei.

Other Well-Established Causes

Well-established and usually rather obvious causes of sudden deafness, such as bacterial meningitis, labyrinthitis due to bacterial invasion of the inner ear, mycoplasma infections (Shannon et al, 1982), syphilis, trauma, and ototoxic drugs have been excluded from the discussion of documented, specific etiologic agents.

Acoustic Neuroma

Shaia and Sheehy (1976) have reported that 1 per cent of 1220 patients presenting with sudden sensorineural hearing loss were ultimately found to have acoustic neuromas.

Studies by Pensak and colleagues (1985) and Berg and co-workers (1986) have shown that approximately 15 per cent of patients with acoustic neuromas will present with sudden sensorineural hearing loss as their initial symptom. In Berg's report, four of 17 such patients actually recovered auditory function prior to surgery. This suggests that even patients with documented recovery of hearing should undergo a comprehensive evaluation for an acoustic neuroma or other cerebellopontine angle lesion.

Noise

In addition to noise-induced deafness, which can be divided into a slowly progressive deafness caused by prolonged intense noise exposure and acute acoustic trauma caused by a single exposure to very intense sound, Kawata and Suga (1967) described noise-induced deafness that occurs suddenly after a certain period of exposure to uniform, intense noise. Seventeen of their patients developed sudden unilateral hearing loss during exposure to noise to which they had long been accustomed. The audiometric configuration of these losses is often U-shaped or flat.

Pressure Changes

Simmons (1968) reported 15 cases of instantaneous sudden deafness which he theorized was caused by a break in Reissner's membrane. Eight of these patients experienced the loss of hearing while engaged in some activity that might have resulted in pressure changes in the inner ear fluids, such as getting out of bed, sneezing, coughing, bending, performing a Valsalva maneuver, or scuba diving.

Gussen (1981, 1983) subsequently provided histopathologic support for the concept of cochlear membrane breaks in temporal bones from three patients with documented sudden sensorineural hearing loss. In all three, the left temporal bone was involved and there was a rupture in Reissner's membrane near the ductus reuniens in the basal turn. The third patient had suffered a sudden loss in the right ear as well. Examination of the right temporal bone revealed healed Reissner's membrane breaks at two locations. Gussen hypothesized that the third patient's underlying cerebrovascular disease may have predisposed her to membrane breaks.

Perilymph Fistula

Fee (1968) first described one case of spontaneous oval window fistula without preceding injury or subjection to pressure change. Goodhill (1971, 1973) described oval and round window fistulas with and without preceding ambient pressure changes or activities that might result in pressure changes in the inner ear fluids. Simmons (1973) emphasized that it is important to recognize these few from other patients with sudden deafness. Seltzer and McCabe (1986) have reviewed an extensive experience with this problem.

There is little doubt that perilymph fistula - either spontaneous or following surgery, barotrauma, or acoustic trauma - can cause sudden deafness. Nevertheless, this is an area of significant controversy because of the lack of firm criteria for its diagnosis, including intraoperative observation. Even the trained observer exercising responsible judgment often has difficulty determining whether a fistula is present or if he or she is being misled by the accumulation of blood, local anesthetic, or other fluids in the niche of the round or oval window. At the present time we must recognize that there is no conclusive means of demonstrating the existence of all but the most obvious fistulae; conversely, there may indeed be some fistulae present that are not detectable at the time of exploration. Because of this uncertainty, some surgeons (Parell and Becker, 1986) recommend surgical "repair" of inapparent fistulae at the time of tympanotomy when the symptoms are suspicious enough to warrant exploration of the ear. Others feel that the long-term results are equivalent if the procedure is merely terminated. Most now agree that fibrous connective tissue is superior to fat for repair of labyrinthine fistulae (Seltzer and McCabe, 1986; Parell and Becker, 1986).

Vascular Causes

Turning now to those factors often purported to be etiologic in sudden deafness, but for which there is less documentation, we may first consider the vascular causes. Vasospasm, thrombosis, embolism, hemorrhage into the inner ear, hypercoagulation, and sludging of blood are frequently mentioned as common causes of sudden deafness, but the evidence for this point of view is lacking.

Vasospasm (ie, arterial vasoconstriction) is often attributed to stress, fatigue, and the emotional state of the patient. Allergic reactions of the antigen-antibody type have been implicated as the cause of arterial vasoconstriction, as has autonomic imbalance. The mechanism of venous vasoconstriction is less clearly understood.

Thrombosis and embolism are usually attributed to arteriosclerosis. As mechanisms for sudden deafness, they become more plausible in the older age group, but they do not provide a very satisfactory explanation in younger patients or in patients who have no other evidence of arteriosclerosis.

Jaffe (1967) reported hypercoagulation in two patients with sudden deafness on the basis of finding a rapid prothrombine consumption time. Further study of this problem is necessary.

Little information regarding sludging of blood as a cause of sudden deafness is available. Clinical observation in the form of microscopic studies of fingernail beds, clumping of red cells in retinal vessels, and clumping of red cells in vitro involves considerable subjective interpretation.

Kirikae and colleagues (1962) presented the clinical findings in a patient with thromboangiitis obliterans (Buerger's disease) and unilateral sudden deafness and discussed the possible relationship between the two.

Jenkins and associates (1986) presented the findings from the temporal bones of a patient with polyarteritis nodosa who suffered a sudden hearing loss 7 months prior to death. They showed severe arteritis in the small vessels of the cochlea with loss of the organ of Corti in the basal coil, absence of the tectorial membrane, and atrophy of the stria vascularis. In addition, there was a loss of spiral ganglion cells on the affected side.

Dubs (1956) reported three cases of sudden deafness due to fat embolism following crushing injuries of various parts of the body. This may also occur following breast or femur surgery. There is no serious reason to doubt fat embolism as a possible mechanism of sudden deafness. Fat droplets are elastic and can pass through lung capillaries 5 to 7 microns in diameter, or they can circumvent the pulmonary circulation through direct anastomosis between small pulmonary arteries and veins.

Sudden deafness has been reported to occur following nonotologic surgery. Of particular concern is the issue of sudden deafness after cardiac surgery. Millen and co-workers (1982) rejected the notion that sudden hearing loss complicating nonotologic surgery occurs by coincidence, particularly in the setting of cardiopulmonary bypass. They cited a male predominance of 12 out of 13 cases and a poor spontaneous recovery rate as evidence that this was related to the surgery rather than a chance occurrence of hearing loss in the perioperative period. Putative mechanisms include the formation of platelet emboli or inadequate perfusion of the cochlea due to loss of the pulse pressure and hypocapnia in patients with pre-existing cerebrovascular disease placed on cardiopulmonary bypass. They pointed out that, unlike idiopathic sensorineural hearing loss, there is strong reason to suggest a vascular insult in these cases, accounting for the severe degree of loss and the poor recovery rate.

Gussen (1976) has reported the temporal bone findings in a patient with hypertension, congestive hear failure, and renal failure who developed sudden deafness 2 months prior to death.

The pathologic changes were consistent with vascular embarrassment of the inner ear of 2 months' duration. The organ of Corti was absent throughout the cochlea, and the stria vascularis was atrophic throughout. The spiral ganglion cells were considerably decreased in the basal turn and somewhat decreased in the apical turn. Proliferating fibrous tissue and small spicules of new bone were present in the perilymphatic space of the posterior semicircular canal.

Despite these specific instances, it must be noted that the general popularity of the concept of vascular causes of sudden deafness is based not on evidence but rather on analogy with the rapid onset of other neurologic symptoms and signs in vascular accidents. The temporal bone changes in most of the cases of sudden deafness that have been examined pathologically are not similar to the findings that Perlman and colleagues (1959) and Kimura and Perlman (1956, 1958) demonstrated by experimental arterial and venous occlusion in animals. In contrast to the pathologic changes seen in most of the cases of sudden deafness, the changes in the inner ear of guinea pigs following temporary occlusion of the internal auditory artery involved more severe damage to the ganglion cells than to the hair cells, and there was little variation in the degree of damage from one turn of the cochlea to another. There tends to be a loss of spiral ligament cells, variable hair-cell loss, and little effect on the tectorial membrane in temporary arterial occlusion. Permanent and complete arterial occlusion produces generalized destruction of the inner ear structures and fibrous tissue invasion and, finally, complete ossification of the cochlea within 6 months.

Autoimmunity

In recent years, the issue of immune-mediated sensorineural hearing loss has received significant attention. McCabe (1979) described a clinical entity that he felt was immune-mediated, consisting of asymmetric sensorineural hearing loss of slow onset (several weeks) and accompanied by facial paralysis in five of 18 cases. The evidence suggesting an immune process was three-fold: (1) of six patients tested, all had positive lymphocyte inhibition tests to a crude mixture of inner ear antigens; (2) frank vasculitis was seen on biopsy of a granulomatous lesion from one patient's temporal bone; and (3) all patients with residual hearing improved on corticosteroid and cyclophosphamide therapy. He felt this entity was important to consider because it was one of the few causes of sensorineural hearing loss that will reliably respond to treatment.

Subsequently, Harris (1983) demonstrated experimentally that the inner ear is capable of mounting an immune response to antigenic challenge, and that the antibodies were indeed produced in perilymph rather than being transported from the middle ear or cerebrospinal fluid. Mogi and associates (1982) also demonstrated the presence of higher levels of IgG and IgA in perilymph than in cerebrospinal fluid. They argue that these are an ultrafiltrate of blood from the perilymphatic vessels. Schiff and Yoo (1985) have reviewed the spectrum of otologic disease felt to be immune-mediated and feel that the endolymphatic sac and perisaccular tissues are the site of inner ear immunoreactivity. Hughes and colleagues (1986) have further studied the role of the lymphocyte transformation test in patients felt to be at "high risk" for autoimmune inner ear disease. Only 13 of 58 patients (22 per cent) had positive results, but they concluded that the test

was fairly specific when positive, in that only one of 15 controls (7 per cent) was positive. They also pointed out that one-fourth of patients with autoimmune inner ear disease have an underlying systemic immune disorder such as rheumatoid arthritis. The role of autoimmunity in sudden sensorineural hearing loss has yet to be clarified.

Pathology

The pathology of sudden deafness presented by Schuknecht et al (1973) is similar to the findings in viral endolymphatic labyrinthitis and, of course, represents the final stage of these processes in patients in whom spontaneous recovery did not occur. Therefore, their findings can be considered the end result of the most virulent infections and do not represent the outcome in all cases.

The organ of Corti is often missing in the basal turns, and individual hair cells tend to be missing at higher turns. Ganglion cell populations are decreased at the basal turn but are more normal toward the apex. The stria vascularis tends to be atrophic. The tectorial membrane is often atrophic, rolled up, and ensheathed in a syncytium of cells on the limbus. Reissner's membrane may be collapsed and adherent to the basilar membrane. The saccule is often involved, but the utricle and semicircular canals usually escape severe damage.

The contrast of these changes with the findings in the experimental pathology of vascular occlusion and the one human case presented by Gussen (1976) has already been cited. Alford and co-workers (1965) and Suga and associates (1970) have studied experimental microembolization in animals and, as in other forms of vascular occlusion, degeneration of the spiral ganglion cells to the apex of the cochlea and fibroblastic invasion and new bone formation in the cochlea were prominent.

Clinical Manifestations

The incidence of sudden deafness has been estimated at one case per 5000 population per year (Van Dishoeck and Bierman, 1957). Probably many more cases of sudden deafness develop than present clinically. It is likely that many of these hearing losses resolve spontaneously. The relatively high percentage of physicians with sudden deafness in several series indicates that they may seek attention more promptly. The high recovery rate of 80 per cent in physicians may more accurately reflect the true recovery rate than series in which early and late cases are combined (Simmons, 1968).

Males and females appear equally likely to develop sudden deafness. Each ear is equally vulnerable. There does not appear to be any specific occupation predisposition to sudden deafness, although the relationship to workers subjected to atmospheric pressure changes and noise has been noted, as well as the apparent high incidence in physicians.

The loss of hearing is usually unilateral. The percentage of patients with bilateral involvement varies from 4 to 17 per cent (Sheehy, 1960; Van Dishoeck, 1957; Jaffe, 1967).

Deafness

A great deal of variation occurs in the evolution of the symptoms of sudden deafness. The deafness may begin instantaneously and, when the onset is so sudden, it may be accompanied by the sensation of a loud sound in the affected ear. More often the hearing loss develops over the course of an hour, a day, or several days. Frequently the hearing loss is first noticed by the patient on awakening in the morning. Some patients are awakened from sleep by the associated tinnitus. If the loss is bilateral, it will be noted promptly once interpersonal communication is hindered. On the other hand, unilateral losses may escape the patient's detection until some specific test of the affected ear, such as use of the telephone, occurs.

Dating of the onset is often difficult in children and in adults as well. Usually the hearing loss draws the patient's attention promptly, regardless of the presence or absence of tinnitus. The patient is often aware of the profound loss of discrimination that is initially present. Unusual sensitivity to intense sound and diplacusis are rare. As a rule, difficulty in localizing sound is experienced.

Tinnitus

Roughly 70 per cent of patients with sudden deafness experience tinnitus of varying degrees sometime during their illness. The tinnitus may precede the hearing loss by several hours; it usually subsides within 1 month but may persist and even outlast the deafness. It usually has a roaring quality.

Vertigo

There is considerable variation in the incidence of vertigo from one series of patients to another. In general, some 40 per cent of patients with sudden deafness have mild or transient vertigo, and 10 per cent have incapacitating vertigo that lasts for 4 to 7 days. Lesser degrees of vertigo may then persist for up to 7 weeks. Nausea and vomiting are usually associated with severe vertigo. A sensation of pressure in the affected ear is experienced by many of these patients.

Headache is occasionally encountered, and symptoms of viral upper respiratory tract infections occur in as many as 25 per cent of the patients in some series. Fever, usually of mild degree, may be present. Generally, however, the patient feels perfectly well except for the loss of hearing and tinnitus.

Usually the otoscopic examination is normal, but serous otitis media is occasionally observed and may add a conductive component to the loss of hearing.

Diagnostic Tests

Audiologic Studies

A comprehensive audiologic assessment should be performed initially, including pure-tone air and bone conduction thresholds, spondee thresholds, performance-intensity function for speech discrimination using phonetically balanced words, and acoustic immittance.

Pure-tone thresholds can be categorized according to the classification of Sheehy (1960) or Rubin (1968), which were mentioned earlier. These tests, as well as spondee thresholds and discrimination scores, should be obtained initially and at 2- to 3-day intervals until the hearing loss is stable. Stenger pure-tone and speech tests should be performed initially to rule out the possibility of a psychogenic hearing loss.

Auditory brain-stem response (ABR) testing can be useful as an electrophysiologic correlate to the pure-tone threshold determined by the audiogram. A latency-intensity function for wave V can be generated that would be expected to correspond to the pure-tone threshold in the middle- to high-frequency range. Perhaps more important, ABR can help to exclude a retro-cochlear lesion as the cause of the hearing loss. In 33 patients with recordable ABRs found to have acoustic tumors, the ABR was abnormal in every case (Pensak, 1985), usually showing a prolonged I-III interpeak latency, absence of all waves except wave I, or a prolonged absolute latency of wave V. A normal ABR is highly suggestive of a cochlear hearing loss. When the ABR is equivocal, site-of-lesion testing can be performed, such as tests for recruitment and tone decay, the small increment sensitivity index, and Békésy audiometry.

Initially recruitment usually is not present, but it may appear later if the loss of hearing becomes permanent. In the permanent losses, recruitment is present in less than half the patients (Van Dishoeck, 1957; Jerger et al, 1961). Jerger (1960) reported that the majority of patients with permanent hearing loss of sudden onset had type III Békésy audiograms. On subsequent study, Jerger and colleagues (1961) reported that two-thirds of patients with permanent hearing losses of sudden onset had patterns of response generally associated with a neural hearing loss (low SISI scores, type III Békésy audiograms, nonmeasurable discrimination scores, and minimal recruitment), while one-third had patterns associated with cochlear hearing loss (high SISI scores, type II Békésy audiograms, some recruitment, and measurable but low discrimination scores). Stephens and co-workers (1967) reported a patient in whom a type IV Békésy audiogram was found during the acute phase; the type IV Békésy audiogram then converted to type II within 5 months. Such variability of these special audiologic tests with time in a given patient has been observed by others.

Vestibular Studies

In general, vestibular studies using the Fitzgerald-Hallpike test with electronystagmographic monitoring are of interest but need not be carried out in the acute phase. Approximately 50 per cent of patients with sudden deafness will have some permanent

abnormality of response of the vestibular apparatus to caloric stimulation. Abnormalities vary from complete loss of response to mild variations from normal.

A fistula test can be performed by varying the pressure in the external auditory canal with an impedance bridge and monitoring the eye movement response with electronystagmographic tracings.

Tests of Eustachian Tube Function

Differences of opinion exist regarding the advisability of testing eustachian tube function. Simmons (1968) advised against it in order to avoid fluid pressure changes in the inner ear.

Imaging of the Temporal Bone and Cervical Spine

Computed tomography (CT) of the temporal bone with enhancement and radiographic examination of the cervical spine are indicated to avoid overlooking mastoiditis, primary cholesteatoma, acoustic neuroma, and spondylosis deformans. High-resolution CT permits a high-quality image of the temporal bones. It should be performed with intravenous contrast enhancement to reliably exclude acoustic neuroma. If an acoustic tumor or other cerebellopontine angle tumor is suspected and the CT scan is normal, a CO_2 contrast-enhanced CT or a magnetic resonance imaging study of the cerebello-pontine angle and internal auditory canal is indicated.

Lumbar Puncture

Judgment must be exercised in the individual patient with sudden deafness in determining whether a lumbar puncture should be done. Measurement of opening and closing pressures and examination of the cerebrospinal fluid for color, clarity, cells, protein content, electrophoresis, and serology may provide essential information that is available by no other means. One must bear in mind that sudden deafness does occasionally occur with cerebellopontine angle tumors, meningoencephalitis, multiple sclerosis, and trauma.

Physical Examination and Laboratory Tests

In the patient's general evaluation and physical examination, the blood pressure, eye grounds, and peripheral pulses may shed light on the problem. Blood studies of value indicate a complete blood count, fasting blood glucose, blood urea nitrogen, serologic test for syphilis (FTA-ABs or MHA-TP), total protein and albumin-globulin ration, cholesterol, lipid fraction, total lipid, and coagulation studies.

Differential Diagnosis

In evaluating a patient with sudden deafness, one must remember that the most likely cause, viral endolymphatic labyrinthitis, is essentially a diagnosis of exclusion. Consideration should be given to possibilities such as ototoxicity, Ménière's disease, trauma, bacterial

labyrinthitis, and perilymph fistula. Any of these entities can masquerade as sudden hearing loss but can usually be differentiated on the basis of history or physical examination. A history of a systemic immune disorder should be sought. Labyrinthine otosclerosis should be considered in patients with a family history of otosclerosis. Such patients may have abnormalities on acoustic immittance testing, despite the absence of a detectable conductive hearing loss.

The physical examination may reveal an unsuspected cholesteatoma or a positive fistula test, which will guide the evaluation and therapy differently than if the otoscopic findings are normal. Associated neurologic signs, although rare, should not be overlooked.

Auditory and vestibular evaluations as previously outlined should be performed. Laboratory testing is often unproductive but can detect important disease processes that might otherwise be overlooked. The serologic test for syphilis should never be neglected unless another diagnosis becomes obvious.

Radiographic evaluation should be performed to eliminate the possibility of acoustic neuroma or other cerebellopontine angle tumors, as well as primary lesions of the temporal bone such as petrous apex cholesteatoma, even when the ABR is normal.

Some patients will require a diagnostic exploratory tympanotomy if perilymph fistula cannot be reliably excluded, particularly when brief episodes of positional vertigo are present or when the hearing is actively fluctuating.

When those avenues of evaluation are unrevealing, it is safe to conclude that the patient has had an idiopathic sudden sensorineural hearing loss, recognizing that present knowledge suggests that most of these are viral in origin and only rarely due to vascular insults.

Natural History and Prognosis

The prognosis for the recovery of hearing in sudden deafness is not as bad as is generally supposed. Approximately one-third of patients have a return of normal hearing, one-third are left with a 40 to 80 dB speech reception threshold, and one-third have total loss of useful hearing. Spontaneous recovery of normal hearing is more likely to occur if the deafness is not associated with severe vertigo and if the deafness is not total initially. Nishida and associates (1976) have correlated a dominant negative summating potential and a high response action potential of the eight nerve in electrocochleography with substantial improvement in hearing within 1 month. The prognosis cannot be easily related to the rapidity of onset, presence of tinnitus, recruitment, or type of Békésy audiogram.

Once recovery of hearing begins, it is likely to take place very rapidly in a matter of a few days. The longer the delay between the onset of deafness and the onset of recovery, the worse the prognosis for complete recovery. Nevertheless, complete recovery of hearing does occur at times even after several weeks of profound loss. Even when severe initially, the vertigo tens to subside within 1 week and, as a general rule, all vestibular symptoms clear spontaneously

within 6 weeks.

Therapy

The frequent spontaneous recovery of hearing to normal or near-normal levels makes evaluation of any form of therapy for sudden deafness very difficult. No controlled studies have been performed with any of the forms of the therapy that have been advocated. Each form of therapy appears to be effective in a large number of patients, and no form of therapy has been found to be effective in all patients with sudden deafness. In fact, it is difficult to judge whether any form of therapy advocated for sudden deafness produces a higher recovery rate than would have occurred spontaneously.

The therapy currently advocated includes vasodilatation, anticoagulation, reduction of the viscosity of the blood, corticosteroids, vitamins, sedation, tranquilization, bed rest, and the surgical repair of oval or round window fistulae. Vasodilatation, anticoagulation, and reduction of the viscosity of the blood are based on the theory of vascular causes of sudden deafness. Corticosteroid therapy is based on the anti-inflammatory effects of these agents in viral infections. The rationale for the vitamin therapy is less clear. Sedation, tranquilization, and bed rest have been advocated without regard to cause. The exploration for and repair of oval and round window fistulae with tissue grafts at tympanotomy are indicated in sudden deafness that occurs while the patient is engaged in some activity that might result in pressure changes in the inner ear.

Vasodilatation has been advocated by Van Dishoeck and Bierman (1957), Shaia and Sheehy (1976), Jaffe (1967), and Rubin (1968). Van Dishoeck and Bierman mentioned spasmolytics but did not specify the agents. Shaia and Sheehy (1976) advocated intravenous histamine phosphate initially, to be followed by subcutaneous histamine phosphate. Subsequent oral therapy consisted of sublingual histamine phosphate and nicotinic regimen, 40 per cent of patients had significant hearing improvement. Jaffe also advocated intravenous histamine and oral nicotinic acid.

The manufacturer has excluded cochlear vasodilatation as an indication for the use of intravenous histamine. Adverse effects of intravenous histamine include tachycardia, cardiac irregularity, hypotension, hyperthermia, and bronchiolar constriction. It should not be used in patients with asthma. It can produce a hypertensive crisis in patients with pheochromocytomas due to the release of catecholamines. Phentolamine should be available to manage this situation. Likewise, antihistamines and epinephrine as well as standard resuscitation equipment should be on hand whenever histamine is administered intravenously.

Among other forms of vasodilatation, Rubin (1968) advocated hyoscine or atropine intramuscularly or intravenously in the acute phase and procaine hydrochloride intravenously after several days or weeks. He also recommended nylidrin, a noncatecholamine and beta-receptor stimulant. Rubin reported return to normal hearing in 50 per cent of his type I patients, but no appreciable effect in his type II and III patients. Stellate ganglion blocks have been advocated

by Haug and co-workers (1975) in a controversial study.

Shea and Bowers (1975) emphasized abstinence from the use of tobacco and the inhalation of 5 per cent carbon dioxide and 95 per cent oxygen in the treatment of fluctuant hearing loss.

In a study by Siegel (1975), the results of vasodilatation therapy appear to approximate spontaneous recovery rates.

Fisch (1983) measured changes in perilymphatic PO_2 in response to changes in arterial PO_2 and PCO_2. Hypoventilation or inhaled CO_2 induced a marked increase in perilymphatic PO_2. He suggested that cochlear blood flow is not subject to the same autoregulation found in the rest of the cerebral circulation and therefore can be increased by vasodilatation with Carbogen (5 per cent carbon dioxide and 95 per cent oxygen) inhalations.

Fisch extended these studies to human subjects and demonstrated a baseline reduction in perilymphatic PO_2 in patients with sudden hearing losses compared with patients with progressive hearing losses. He also demonstrated a transient rise to 175 per cent of baseline in response to Carbogen, peaking at 13 minutes of therapy. He noted a significant difference in outcome of patients treated with Carbogen over papaverine when studied at 1 year after their sudden hearing loss. However, no prospectively randomized untreated control group was studied, nor were the results compared with a historical control group. The use of Carbogen is somewhat impractical because of its poor patient tolerance (headache) and the need for frequent administration.

Study of the effect on cochlear blood flow in guinea pigs of adrenergic, adrenergic-blocking and antiadrenergic, cholinergic, anticholinesterase, and cholinolytic agents indicates that the adrenergic and cholinergic nervous systems have relatively weak control over cochlear vessels, which is also true of their effect on cerebral vessels (Suga and Snow, 1969).

In their study of vasodilating drugs and some related agents, Suga and Snow (1969) showed that nicotinic acid, even in massive doses, has no measurable effect on cochlear blood flow. Inhalation of 10 per cent carbon dioxide and 90 per cent oxygen for 15 minutes produces marked and progressive cochlear vasodilatation. Histamine phosphate and betahistine increase cochlear blood flow in dosages that produce bronchospasm in the guinea pig and may well produce vasodilatation on the basis of anoxia. Betahistine has been removed from the market by the US Food and Drug Administration because of the lack of demonstration of its clinical efficacy. Papaverine relaxes smooth muscle of large blood vessels, particularly during vasospasm. It reliably increases cochlear blood flow in guinea pigs.

The rationale of vasodilatation therapy for sudden deafness is questionable in view of the preponderance of evidence for the viral etiology of most cases of sudden deafness. Some now advocate vasodilatation in view of the vascular changes that occur in viral diseases. Granting these changes in individual capillaries, overall blood flow may not be decreased but may actually increase in the area of inflammation. Should cochlear vasodilation be achieved clinically by

intermittent inhalation of 5 per cent carbon dioxide and 95 per cent oxygen, or in a more sustained fashion with papaverine, there is serious question whether such an effect is desirable in the presence of micropetechiae in viral infections. Resolution of the question of the efficacy of this therapy must await well-controlled clinical studies.

Bolognesi (1960) advocated anticoagulant therapy after improvement in the hearing of three of five patients on heparin and coumadin therapy. Clemis (1975) suggested that the beneficial effect of heparin might be mediated through its ability to detoxify histamine locally as in an allergic reaction. Siegel (1975) was not able to demonstrate that treatment with heparin or coumadin improved the spontaneous recovery rate. Viral infections often produce temporary bleeding tendencies. In view of the probable viral etiology of most sudden deafness, such therapy may well be contraindicated.

Jaffe (1967) advocated low molecular weight dextran to reduce the viscosity of the blood, but a controlled study of this form of treatment is lacking. In addition, there have been at least two mortalities from the use of this therapy for sudden hearing loss.

Shea and Bowers (1975) advocated corticosteroid therapy in the form of dexamethasone for fluctuant hearing loss. It was advocated for sudden deafness as well on the basis of a rather carefully controlled study by Byl (1975). Van Dishoeck and Bierman (1957) mentioned its use without enthusiasm. It appears safe in view of the vast neurologic experience of treating herpes zoster meningoencephalitis with corticosteroids. We believe that this is the only currently acceptable form of medical therapy for idiopathic sudden hearing loss and advocate the use of prednisone in doses of 40 to 60 mg per day in a single morning dose for a full week, followed by a rapid tapering schedule.

Vitamins in nontoxic doses do no harm. Sedation and tranquilization may be appropriate in certain patients but may not be innocuous in this situation. Barbiturates may potentiate the ototoxic potential of ethacrynic acid.

Unfortunately, there is no specific therapy for viral infections at the present time. Until there is, care must be taken to avoid aggravating the delicate balance in a gravely injured inner ear that has at least a 33 per cent and perhaps an 80 per cent chance of complete recovery. The patient should be advised to withdraw from the stress and strains of daily work, avoid all strenuous activity, and abstain from the use of tobacco and ethanol. The patient should rest at home, and the rest should be continued until the hearing loss becomes stable.

Rehabilitation

Young patients who do not recover spontaneously from unilateral sudden deafness should have preferential seating in the classroom and be advised on maneuvering persons in conversation to the side of their better ear. Children and adults must be acquainted with their inability to localize the source of sound and must truly stop, look, and listen, rather than just listen, when crossing the street. The use of amplification with the contralateral routing of offside signals

(CROS) hearing aid may benefit some of these patients (Harford and Barry, 1965; Harford and Dodds, 1966).

Those patients who do not recover serviceable hearing from bilateral sudden deafness should have speech reading and auditory training to enable them to make the most of their residual hearing. Appropriate amplification with a hearing aid should be utilized when appropriate. If no benefit is obtained with amplification, these patients usually are suitable candidates for cochlear implantation.