

Chapter 177: Sudden Sensorineural Hearing Loss

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A useful definition of sudden sensorineural hearing loss (SSHL) is a loss that is greater than 30 dB in three contiguous frequencies and that occurs in less than 3 days. Most sudden hearing loss occur within minutes to several hours. Approximately one third of these patients awaken in the morning with a hearing loss; about one half note an impaired sense of balance or vertigo. The intensity of the vertigo correlates in general with the degree of hearing loss.

The potential causes of SSHL are varied (see box). Some are immediately apparent; some are revealed by the history and testing. However, the majority of cases elude precise etiologic determination and are termed *idiopathic sudden sensorineural hearing loss* (ISHL). Consequently, idiopathic sudden sensorineural hearing loss remains a diagnosis of exclusion, and an appropriately focused diagnostic evaluation (see box, p 3104) should be undertaken to detect all known potential causes. This chapter will first discuss the evaluation and then concentrate on those causes of sudden sensorineural hearing loss where the etiology remains unknown despite a complete evaluation, that is, ISHL.

Diagnostic Evaluation

The evaluation of the patient with SSHL should include a history and review of symptoms, otologic and neurotologic examination, audiologic testing, and laboratory studies. It is impractical to test specifically for every potential etiology, and therefore screening tests should be used. A suggested list of tests (see box on p 3105) provides for initial screening. A decision regarding the method of treatment should be made promptly when the patient is first examined and after the immediately available laboratory work is reviewed, that is, at most within 1 day. The presumed etiology dictates the appropriate therapy.

The primary development in the diagnostic evaluation of patients with SSHL is the early use of magnetic resonance imaging (MRI) with gadolinium enhancement. These studies have helped to document multiple sclerosis (Barratt et al, 1988) and small cerebellar strokes (Huang and Yu, 1985; Rubenstein et al, 1980) as the bases for sudden hearing loss. In addition, it appears that MRI may be sufficiently sensitive to demonstrate presumed viral inflammation of the cochlear or vestibular membranous labyrinth in patients with ISHL on T-1 weighted images (Seltzer and Mark, 1991) as well as seventh cranial nerve inflammation in Bell's palsy (Millen et al, 1989). When available, these studies are very useful.

Among new blood tests, the Western blot test for identification of IgG and IgM antibodies specific for *T. pallidum* antigens should aid in the diagnosis of syphilis. For the first time, concrete evidence of active infection can be found in patients with congenital or latent disease (Birdsall et al, 1990).

Box: Potential causes of sudden sensorineural hearing loss

Viral infection

1. Viral cochlear labyrinthitis
2. Viral vestibular labyrinthitis
3. Cochleovestibular labyrinthitis
4. Viral neuritis, auditory nerve
5. Viral polyneuropathy (including Ramsay-Hunt syndrome)
6. Viral-induced meningoencephalitis
7. Specific viruses: mumps, CMV, rubella, rubeola, varicella zoster, HSV I, HSV II, parainfluenza A, B, and C, Lassa fever (arena virus)
8. EB virus, HIV.

Vascular occlusion

1. Partial
 - a. High-viscosity syndromes: macroglobulinemia, polycythemia vera, decreased blood filterability
 - b. Small vessel obstruction: sickle cell anemia, microemboli, bubble (caisson disease)
 - c. Small vessel narrowing: diabetes mellitus, atherosclerosis, Buerger's disease (thrombangitis obliterans)
 - d. Hypercoagulability states
 - e. Vasospasm
2. Complete: thrombus or embolus of labyrinthine or cochlear artery; microemboli secondary to routine or pump bypass surgery
3. Inner ear hemorrhage: leukemia, anticoagulated states.

Cochlear membrane breaks

1. Intracochlear breaks: Reissner's membrane tear, with and without hydrops (theoretic)
2. Oval window and round window membrane tears
 - a. Secondary to head injury
 - b. Compression or decompression of ear
 - c. Poststapedectomy
 - d. Secondary to congenital malformation

Bacterial infections

1. Meningitis, labyrinthitis secondary to chronic ear infection or surgery
2. Syphilis, primary through tertiary stages.

Autoimmune disorders

1. Inner ear autoimmune disease
2. Ulcerative colitis
3. Relapsing polychondritis
4. Lupus erythematosus
5. Polyarteritis nodosa (Kussmaul's disease)
6. Cogan's syndrome (oculovestibuloauditory syndrome)

Neurologic disorders

1. Multiple sclerosis
2. Migraine.

Neoplasms

1. Vestibular schwannoma (acoustic neuroma)
2. Metastatic cancer
3. Paraneoplastic disorders.

Ototoxic drugs

1. Bilateral
2. Unilateral (otic drops).

Psychogenic causes

1. Malingering
2. Converse reaction.

As more is learned about the specificity of whole blood filterability (Ciuffetti et al, 1991) vis-à-vis ISHL, these tests may become of more than research interest.

Box: Initial assessment of patients with sudden sensorineural hearing loss

History and neurootologic evaluation

Audiologic evaluation

Air and bone conduction (pure tone)
Speech audiometry, rollover screening (PIPB)
Auditory brain stem response (ABR)
Upright and recumbent audiograms (for suspected fistulas only)

Laboratory studies

CBC/ESR
Glucose Hb Alc
Cholesterol/triglycerides
T3, T4, TSH
PT, PTT, serum filterability tests (if available)
VDRL, FTA-ABS (MHA-TP)
HIV antibody
Lyme disease antibody titer (*Borrelia burgdorferi* antigen)
Acute and convalescent viral antibody titers (onset, 1 month, and 3 months)

MRI with gadolinium

Electronystagmography with calorics and positional testing.

Clinical Observations in ISHL

Viral causes

Evidence available to date indicates that the most common cause of ISHL is viral cochleitis. Vouri et al (1962) established a relationship to clinical mumps in a study demonstrating that 4.4% of soldiers with mumps (parotitis) suffered a sensorineural hearing loss. Mumps virus has been grown from the perilymph of a patient with SNHL (Westmore et al, 1979); however, serologic studies have implicated many other viral species that appear to have an indirect or direct relationship to ISHL. Maasab (1973) found fourfold titer changes to parainfluenza virus, adenovirus, and herpes hominis virus, as well as to *Mycoplasma pneumoniae*. Studies by Wilson et al (1983) uncovered a significantly increased incidence of positive viral seroconversion in ISHL patients, as compared with control subjects, for influenza B, mumps, rubeola, cytomegalovirus, and varicella-zoster virus ($p < 0.05$). In addition, patients also had positive titers for rubella and herpes simplex I. There was no difference in the incidence of seroconversion on the basis of age, an indication that viral infections are dispersed throughout age groups.

New evidence implicating viral infection as the source of sudden hearing losses has come from a study by Cummins et al (1990), who noted sensorineural hearing losses among patients hospitalized with Lassa fever in Sierra Leone, West Africa. Fourteen of 49 patients with antibody titer -confirmed Lassa fever suffered hearing loss, whereas none of 20 febrile controls had hearing loss. The incidence of permanent sensorineural hearing loss among seropositive individuals (81% of the local population) is 18%. It appears that viral hearing loss may be responsible for widespread hearing impairment in West Africa (Rybak, 1990).

Cytomegalovirus (CMV) is known to be a leading cause of human congenital viral infection, which may lead to hearing loss (Woolf, 1990). In addition, by means of viral antibody titer and temporal bone studies (Veltri et al, 1981), CMV infection has been implicated as a cause of sudden hearing loss. Fukuda et al (1988) studied the pathogenesis of guinea pig (GP) CMV and related sensorineural hearing loss (SNHL) and found that the virus gained access to the inner ear by means of the modiolar blood vessels and subsequently spread to the spiral ganglion cells. The question of whether the hearing loss is due to direct viral cytopathic effects or to a secondary inflammatory response was answered in part by demonstrating that immunosuppressed guinea pigs suffered less CMV-induced hearing loss than normal controls. There was a direct correlation of hearing loss with the degree of cellular infiltration of the scala tympani (Harris et al, 1990). The predominant role of the inflammatory response in GPCMV-induced hearing loss was again demonstrated (Darmstadt et al, 1990). These animal studies provide a rationale for the observed efficacy of steroid therapy in the early treatment of ISHL in humans since steroids would theoretically reduce the inflammation associated with viral infection.

Further circumstantial evidence for the theory of viral injury as the major cause of ISHL comes from temporal bone histopathologic studies. Schuknecht and Donovan (1986) studied 12 ears with sudden hearing loss and reviewed the literature. They concluded that (1) the histopathologic findings are similar to those occurring with known viral disorders; (2) there is no evidence for recent or remote membrane breaks; and (3) the changes associated with vascular obstruction are not present. Yoon et al (1990) in a report of 11 temporal bones from 8 patients with sudden hearing loss found that atrophy of the organ of Corti and loss of cochlear neurons were the most common changes noted, again suggesting viral infection. They also point out that autoimmune disease of the inner ear associated with hearing loss results in fibrosis and ossification of the labyrinth and cochlea, similar to the changes seen in ears subjected to vascular injury.

Acute viral infection of the ear can take several forms (see the box on p 3104), and testing methods are often not sufficiently specific to differentiate direct involvement of the end organ or organs, for example cochlear or vestibular labyrinths, as opposed to viral eighth cranial nerve neuritis. Temporal bone studies suggest ISHL is often caused by an acute cochleitis or cochleovestibular labyrinthitis, the latter characterized by hearing loss accompanied by vestibular upset. Schuknecht (1985) believes that viral cochlear neuritis is rare, with the exception of Ramsay-Hunt syndrome, which is a varicella-zoster polyneuropathy involving cranial nerves VII and VIII and others as well (Adour, 1976).

The neurotropic herpes virus family may have a special relationship to ISHL, in addition to the Ramsay-Hunt syndrome; Nakajima et al (1976) have detected the herpes virus in the cerebrospinal fluid (CSF) of two of three patients studied with sudden hearing loss. Elevations of antibody titers to the herpes virus usually occur in association with two or more other viral titer elevations (Wilson, 1986), suggesting a possible reactivation of this virus. Recently an uncontrolled study of acyclovir treatment for varicella-zoster-induced facial paralysis and associated hearing loss suggests this may be effective treatment (Dickens et al, 1988). Sudden hearing loss in association with infectious mononucleosis has been reported by Beg (1981).

AIDS

At the time of this writing, there are several reports of sudden sensorineural hearing loss occurring in acquired immunodeficiency syndrome (AIDS) patients (Rarey, 1990; Real et al, 1987). The relationship between AIDS and sudden sensorineural hearing loss is unclear because of the protean manifestations of this disease and the many opportunities for associated otologic and CNS infections, CNS neoplasia, and vascular injury (Levy, 1990). In addition, sudden conductive hearing loss is a frequent occurrence in this disease because of the high incidence of serous and acute otitis media. No figures as to the incidence of sudden sensorineural hearing loss in this group of patients have been published.

Since the recognition of AIDS in 1981 (Gottlieb et al, 1981) and the report of the discovery of the initiating virus in 1983 (Barré-Sinoussi et al, 1983) much has been learned about the disease. The HIV family consists of retroviruses that were first found to be lymphotropic, that is, attaching to and multiplying in the CD4+ lymphocyte (T4 helper lymphocyte); however, it is clear the virus is neurotropic as well, commonly infecting brain cells, as well as cells of the gastrointestinal tract, kidney, lung, and heart. Cultures of inner ear tissues have demonstrated the presence of HIV antigens; however, correlations to sudden sensorineural hearing loss and the HIV virus have not been made.

Perhaps more important in the understanding of the possible mechanisms for the putative AIDS-induced or associated sudden sensorineural hearing loss is the marked predisposition for opportunistic infection and neoplasia in this disease (Kwartler et al, 1991; Nelson et al, 1990). The initial presentation of AIDS as an opportunistic infection other than *Pneumocystis carinii* was noted in only 27% of 231 Danish AIDS victims (Pedersen et al, 1990). It is among this small subset of patients that sudden hearing loss might be the first indication of HIV infection. The potential opportunistic or reactivated infections and neoplasms that might induce an abrupt hearing loss are listed in Table 177-1. Syphilis may take a more aggressive course in these patients (see discussion of syphilis).

Table 177-1. AIDS opportunistic infections with possible relationship to sudden sensorineural hearing loss

Pathogen	Mechanism
	Viral Infections
CMV	Encephalitis
EB virus	CNS lymphoma
HSV-I or HSV-II	Encephalitis, reactivated latent infection
Varicella-zoster virus	Encephalitis, Ramsay-Hunt syndrome
	Fungal infections
Cryptococcus	Meningitis
<i>Candida albicans</i>	Meningitis
<i>Toxoplasma gondii</i>	CNS
	Bacterial infections
Syphilis	CNS and otologic involvement
<i>Mycobacterium tuberculosis</i>	CNS.

In addition, an occasional patient may suffer vascular thrombosis and cerebral hemorrhage possibly in relationship to bacterial or mycotic aneurysms, which could conceivably occur in the auditory system, either in the brainstem or peripherally.

Finally, ototoxic drugs used in the treatment of AIDS or associated infections and/or neoplasia may be responsible for abrupt bilateral sensorineural hearing loss in these unfortunate patients.

Vascular occlusion

Sudden hearing loss can occur following partial or complete occlusion of the cochlear vasculature. Examples of hearing loss from partial blockage include high-viscosity syndromes such as Waldenström's macroglobulinemia in which there are excessive monoclonal IgM antibodies. Treatment consists of reduction in viscosity by plasmapheresis, which can rapidly reverse sensorineural hearing loss (SNHL) and other neurologic symptoms. Polycythemia vera and thrombocythemia are myeloproliferative disorders that have been implicated in SNHL secondary to small vessel obstruction from increased numbers of red blood cells or platelets. Hearing losses are often reversible by phlebotomy or, in the case of thrombocythemia, phlebotomy with removal of platelets (Grisell and Mills, 1986).

Jaffee and Penner (1968) found evidence of hypercoagulability in a group of five of six patients with ISHL; they measured an increased prothrombin consumption, which was suggestive of platelet liability. Such platelet liability can occur in association with acute viral infection as well as other disease states.

In a recent study of hemorrheologic profiles of 16 patients with ISHL and 32 matched controls, Ciuffetti et al (1991) found whole blood filterability and red blood cell filterability to be significantly impaired; however, plasma viscosity and white blood cell filterability were normal. This study suggests diminished microcirculation may have direct relationship to ISHL, but the nature of this relationship will require further study.

The partial blockage of small vessels that occurs in sickle cell disease is associated with SSHL that may or may not return (Orchik and Dunn, 1977). Plasse et al (1981) found that the incidence of SSHL immediately following cardio-pulmonary bypass surgery was 1:1000 patients. These losses presumably are caused by microembolic obstruction of the cochlear artery or its branches, since none of the patients studied had vertigo. Approximately half the patients showed some improvement, but none recovered fully. Millen et al (1982) reported similar findings.

Disorders such as diabetes mellitus that cause small vessel narrowing would seem logically to be related to SSHL, however, to date no series of patients has been large enough to provide statistically significant proof of a relationship (Miller et al, 1983; Wilson et al, 1982). In fact, conflicting evidence exists as to whether diabetic patients as a group have an increased incidence of sensorineural hearing loss (Axelsson et al, 1978; Schuknecht, 1974; Taylor and Irwin, 1978). Hyperlipoproteinemia has been linked to fluctuating hearing loss in children (Strome et al, 1988) and adults (Spencer, 1973) and appears to warrant further study.

Studies of the obstruction of the internal auditory artery in the guinea pig (Perlman et al, 1959) demonstrate a loss of the cochlear microphonic (CM) within 85 seconds. A complete recovery of cochlear responses was observed after occlusions of 8 minutes or less; however, after 30 minutes occlusion, neither CM nor N1 returned to within 50% of the reference amplitude.

Studies of both animal and human temporal bones indicate that arterial obstruction of the labyrinth eventually results in fibrosis and new bone formation (Kimura and Perlman, 1958). Temporal bone series of patients with SSHL (Schuknecht and Donovan, 1986; Yoon et al, 1990) have failed to document fibrosis or calcification and thus attest to the relative infrequency of vascular obstruction as the etiology of ISHL, compared to findings consistent with viral injury.

Vascular spasm is an hypothesized but unproven cause of ISHL. To date no controlled clinical trial of vasodilators has shown these medications to be efficacious.

Buerger's disease

Kirikae et al (1962) reported bilateral sequential SSHL in a patient occurring over the period of 1 month. The patient was diagnosed by arterial biopsy as having thrombangiitis obliterans (Buerger's disease). Angiographic studies demonstrated involvement of the basilar artery system.

Small cerebellar strokes

The precise central nervous system imaging provided by computerized tomographic (CT) and magnetic resonance (MR) scans has facilitated clinical evaluation so as to enable recognition of new clinical disorders, such as small cerebellar infarctions resulting from occlusion of distal branches of either the anterior inferior cerebellar artery (AICA) or the posterior inferior cerebellar artery (PICA). Cerebellar infarctions can mimic acute labyrinthitis, because the primary signs and symptoms are vertigo, nystagmus, nausea and vomiting, usually in conjunction with imbalance, and ataxia of both gait and the extremities. A small portion of these patients suffers tinnitus and sudden hearing loss, and it is this group that is most likely to be referred for neurotologic examination (Huang and Yu, 1985). Audiograms, electronystagmography (ENG), and lumbar puncture are nondiagnostic; however, the majority of these patients manifest neurologic evidence of cerebellar injury such as positive Romberg and past-pointing tests. A CT or MR scan is diagnostic. No specific otologic treatment is necessary other than that required for the cerebellar infarction. The hearing loss frequently resolves over a period of weeks to months (Rubenstein et al, 1980).

The relationship of ISHL to atherosclerosis remains unclear although recent diagnosis of small cerebellar infarcts sheds some light.

Inner ear hemorrhage

Schuknecht (1974) reported finding an intracochlear hemorrhage as the cause of sudden hearing loss in an 11-year-old child who died 6 days later from acute leukemia. Schuknecht believes bleeding into the inner ear does not occur in otherwise healthy individuals. In the future, MRI studies will clarify this clinical impression.

Cochlear membrane breaks

Cochlear membrane breaks are potential causes of SSHL. Intracochlear membrane tears have been identified frequently in patients with cochlear hydrops. A theoretic mechanism for the sudden fluctuations of hearing in patients with hydrops is that these result from tears in Reissner's membrane, with subsequent potassium poisoning of the sensorineural structures of the cochlea.

Round and oval window fistulas may occur following abrupt compression or decompression injury, head trauma, or heavy lifting or straining (Goodhill, 1980; Goodhill et al, 1973). The history often suggests the diagnosis. Physical findings include fluctuating hearing loss and/or tinnitus, which may improve overnight and worsen during the day. The hearing loss may be associated with positional vertigo and nystagmus with the affected ear down. The fistula test is unreliable and should not be used as a guide for surgery. Patients who have had a stapedectomy are at greater risk for developing an oval window fistula (House, 1977).

Cochlear membrane breaks should be considered in pediatric SSHL, especially if temporal bone anomalies such as the Mondini defect are present (Supance and Bluestone, 1983).

Schuknecht and Donovan (1986) found little evidence for membrane rupture in their histopathologic study of 12 temporal bones of patients with SSNHL. However, Gussen (1981, 1983) has demonstrated healed ruptures in the temporal bones of patients with prior sudden hearing loss.

Bacterial causes

Meningitis

Unilateral and bilateral sudden hearing loss is a well-recognized sequela of bacterial meningitis (Munoz et al, 1983; Ravio and Koskiniemi, 1978). This is discussed more fully in Chapter 176.

Syphilis

The incidence of syphilis among patients with sudden hearing loss is not precisely known, but it is thought to be less than 2%. The mechanism for sudden hearing loss in early syphilis (that is, primary or secondary syphilis) is presumed to be syphilitic meningitis and/or otitis manifested by a sudden hearing loss, possibly accompanied by vertigo, nausea, vomiting, headache, meningismus, fever, and blurred vision (Balkany and Dans, 1978; Willcox and Goodwin, 1971). Recently it has been proposed that HIV infection facilitates syphilitic meningitis in early syphilis or the reactivation of latent otosyphilis, thereby precipitating the sudden hearing loss seen in this patient group (Smith and Canalis, 1989).

Treatment of sudden hearing loss secondary to early acquired syphilitic meningitis with intensive penicillin therapy appears to reverse the hearing loss in some reported cases (Balkany and Dans, 1978; Willcox and Goodwin, 1971); however, in patients with congenital or tertiary latent syphilis 12 weekly injections of 2.4 million units benzathine penicillin G suspension (BPG) combined with high-dose steroids appear to give only temporary improvement of the sudden hearing loss, and steroids may be required chronically to slow the advancement of the progressive chronic hearing loss (Zoller et al, 1979). The Centers for Disease Control recommend 2.4 million units BPG for the treatment of early syphilis, and two weekly doses of 2.4 million units BPG for neurosyphilis, but they suggest this treatment has an unacceptable failure rate and that treatment trials with drugs with better CNS penetration than BPG should be initiated (Darmstadt and Harris, 1989; Zenker and Rolfs, 1990).

Neurologic causes

Multiple sclerosis

Abrupt unilateral hearing impairment is an uncommon initial or subsequent symptom of multiple sclerosis (MS) (Shea and Brackmann, 1987). Fischer et al (1985) observed 705 patients with presumed MS for 5 years; hearing loss was noted in 12 patients (1.7%), and abrupt unilateral hearing loss developed in 4 (0.6%). Other symptoms such as tinnitus, vertigo, impaired gait, and ophthalmoplegia were variably present.

The acute hearing loss associated with MS has some unique characteristics in that it generally occurs over a few days and often resolves over the course of several months in association with remission of the disease. It has retrocochlear features evidenced by poor speech discrimination relative to the pure tone audiogram, positive tone decay, and rollover on PIPB. ABR may demonstrate poor wave-form morphology and poor repeatability of waveforms. Other ABR abnormalities include an absent wave I, suggestive of demyelination of the cochlear nerve, and a delayed wave V, consistent with central auditory disruption. Alterations in the ABR can occur without evidence of pure tone hearing loss; however, the ABR tends to normalize as subjective hearing improves.

Autopsy studies (Brock and Gagel, 1933) have shown demyelination of the root entry zone in association with hearing loss. Barratt et al (1988) substantiated these findings by demonstrating lesions located in the root entry zone of the eighth nerve on MRI scans.

Migraine

Sudden hearing loss can occur with migraine but is rare. Migraine and its neurootologic manifestations are discussed more fully in Chapter 182.

Tumors and tumorlike conditions

Temporal bone neoplasms

Metastatic tumors involving the temporal bone are unusual causes of sudden hearing loss. The most common primary lesions have been the breast, lung, kidney, stomach, pharynx, larynx, cervix, uterus, and thyroid gland (Nelson and Hinojosa, 1991). Bergstrom et al (1977) reported sudden deafness secondary to bronchogenic carcinoma. Schuknecht (1974) described a patient with sudden sensorineural hearing loss and facial paralysis secondary to metastatic adenocarcinoma of the breast, and Igarashi et al (1979) reported a patient with bilateral sudden hearing loss secondary to adenocarcinoma of the pancreas.

Acoustic neuroma

Acoustic neuromas may present as sudden deafness (Chow and Garcia, 1985; Higgs, 1973; Pensak et al, 1985) and may account for approximately 1% to 2% of SHL cases. The fact that the hearing loss improves or resolves may not rule out the presence of acoustic tumor (Berg et al, 1986). Therefore every ISHL patient should be evaluated for cerebellopontine angle tumor. In addition, one must be certain that there is no evidence of bilateral acoustic neuroma, since this occurs in approximately 3% of cases (Clemis et al, 1982).

Paraneoplastic syndromes

Paraneoplastic syndromes (PNS) constitute manifestations of malignancy that are not related to local tumor effects or to metastatic dissemination; the major mechanisms by which tumors generate PNS are believed to relate to either the elaboration of hormonelike substances (for example, melanocyte-stimulating hormone (MSH)-like activity in lung cancer) or by inciting an autoimmune response in which the target antigens displayed by the tumor and

responded to by the immune system are also shared by normal host tissue (Bunn and Minna, 1985).

Sudden hearing loss may be an unusual manifestation of PNS. McGill (1976) examined the temporal bones of a woman with sudden, unilateral hearing loss associated with carcinomatous encephalomyelitis and oat cell carcinoma of the lung; there was a near-total loss of cochlear neurons in the affected ear with good preservation of the remaining elements of the cochlea. These findings were thought to be consistent with an autoimmune etiology, that is, a PNS related to the oat cell carcinoma.

Ototoxic drugs

In addition to sudden hearing loss occurring following the administration (either parenterally or aurally) of known ototoxic drugs, sudden hearing loss has been noted after large doses of propoxyphene in two patients (Harell et al, 1978b), after ingestion of piroxicam (Vernick and Kelly, 1986), and after ingestion of naproxen (Chapman, 1982). Harell et al (1978a) also reported a sudden hearing loss following exposure to malathion (75%) and methoxychlor (15%) spray.

Sudden hearing loss has also occurred following chromic acid application to the rim of a tympanic membrane perforation (Taylor, 1975). Sudden sensorineural hearing loss has been noted following repeated streptomycin instillation into the middle ear (Nedzelski, 1991; Schuknecht, 1974) and application of gentamicin-soaked Gelfoam to the round window membrane (Wilson, unpublished data) in the experimental treatment of Ménière's disease. The topical use of neomycin-containing ear drops in an open middle ear has been suggested as a cause of chronic progressive SNHL (Paparella et al, 1970; Podoshin et al, 1989), but potentially ototoxic (gentamicin) ophthalmologic drops used as ear drops per se have not been reported to cause sudden hearing loss (Gyde, 1976). Sudden deafness is known to occur after the use of neomycin irrigations for osteomyelitis and draining wounds (Nilges and Northern, 1971).

Reports of high-frequency SSHL associated with cisplatin chemotherapy place the incidence between 46% and 86%. The hearing loss appears to be dose related, particularly in patients receiving bolus infusions greater than 100 mg/sq m (Guthrie and Gynther, 1985). Below is a short list of ototoxic drugs that may cause sudden hearingloss.

1. Streptomycin
2. Gentamicin
3. Tobramycin
4. Kanamycin
5. Erythromycin (IV)
6. Neomycin
7. Cisplatin
8. Nitrogen mustard
9. Furosemide
10. Ethacrynic acid
11. Deferoxamine.

Malingering

Psychosomatic sudden hearing loss is generally a response to stress and, in my experience (WRW), is more common in the military than in the civilian population. The diagnosis is readily confirmed by ABR, but the treatment should include a search for the underlying cause.

Management and Prognosis

Management of ISHL must be based on etiologic evidence and therapies known to be efficacious. The efficacy of many of the drugs commonly prescribed for ISHL has been studied. In general, the use of potentially harmful medications of unknown efficacy is discouraged except in controlled clinical studies, since deaths have been reported secondary to therapeutic complications (Zaytoun et al, 1983).

Because there is evidence that ISHL is caused primarily by viral infections, Wilson et al (1980) studied the effect of steroid therapy on recovery rate in a double-blind clinical trial involving 119 patients who were seen within a 10-day period following sudden hearing loss; they found that patients with midfrequency hearing losses tended to recover regardless of treatment and that in most instances no treatment was required. Patients with hearing losses greater than 90 dB in all frequencies (profound hearing losses) had no response to corticosteroids and had a very limited recovery rate that was not improved by steroid therapy. Between these two extremes, the study identified a zone of "moderate hearing loss" in which steroid therapy, administered according to a dosage schedule such as that shown in Table 177-2, is beneficial. Among patients with moderate hearing losses, 38% of those in the placebo group recovered, whereas 78% in the steroid group recovered.

Table 177-2. Prednisone (mg) treatment schedule for patients with moderate hearing loss

Day	7 am	1 pm	7 pm
1	40	20	20
2	40	20	20
3	40	20	20
4	40	20	20
5	40	10	10
6	40	10	10
7	20	10	10
8	20	10	10
9	20		10
10	10		10
11	10		
12	10		

Moskowitz et al (1984), in an additional double-blind study of the treatment of ISHL using a similar steroid dosage regimen, confirmed that steroids are beneficial if the hearing loss is not profound and especially if there is an absence of vertigo.

When attempting to predict the likelihood of recovery, the physician must consider factors other than audiogram type. These factors include (1) patient age of 40 years or older, (2) presence of vertigo, (3) existence of changes on the electronystagmogram, (4) time from onset of treatment, and (5) treatment method. Based on these variables, Laird and Wilson (1983) have analyzed the probability of recovering from ISHL. Patients who have an electronystagmogram done at their first visit demonstrate a high correlation between positive ENG findings and the symptom of vertigo ($p < 0.001$); it is evident that ENG is a more sensitive indicator of vestibular injury than vertigo. Of patients without vertigo who demonstrate mild or no vestibular injury on ENG, 70% recover some hearing. In patients without vertigo who have a reduced vestibular response in the affected ear, the recovery rate falls to 45%. Finally, vertigo combined with positive ENG findings indicates a low recovery rate of approximately 30%. In short, the presence or absence of vertigo gives useful predictive information; however, the ENG findings clarify this information.

Patients in the same study whose hearing was affected in high frequencies (4 and 8 kHz) and who had indications of vestibular injury on ENG recovered less well than patients with low-frequency hearing losses. This finding is not surprising, since the basal turn of the cochlea is closest to the vestibular sense organs and the combination of symptoms would indicate that the injury was large enough to include both the auditory and the vestibular sense organs.

It is of interest that in a double-blind, crossover study (Ariyasy et al, 1990) oral corticosteroid therapy was demonstrated to be efficacious in controlling the symptoms of acute (presumably viral) vestibular vertigo.

A second therapeutic approach to ISHL is to attempt to improve presumed disturbances in cochlear blood flow and perilymph oxygenation by use of oxygen and vasodilating agents. Suga and Snow (1969), Snow and Suga (1975), and Fisch et al (1976) found 100% oxygen alone decreased cochlear blood flow and perilymph oxygenation but when combined with CO₂, there was an increase in these same parameters. Additionally, in these experiments, papaverine was only mildly beneficial, and histamine and nicotinic acid were not effective. In studies designed to raise perilymphatic oxygen levels in cats and humans, 5% CO₂ and 95% O₂ (carbogen) were found to be safe and effective, but in a controlled, prospective study of the use of carbogen for the treatment of sudden hearing loss, Giger (1979) could not demonstrate significant immediate improvement in hearing, and Freeman et al (1985), using a CO₂ rebreathing apparatus to raise serum CO₂ levels and improve cochlear blood flow, also failed to influence the recovery rate of hearing.

Other studies have attempted to increase the rate of hearing recovery by altering the clotting and flow characteristics of blood, thereby improving microcirculation.

Donaldson (1979) in a study of 23 patients used historic controls and found no improvement with intensive heparin therapy. Prostaglandin E therapy, despite its effects of vasodilatation and platelet aggregation inhibition, was also found not to be beneficial (Nakashima et al, 1989). Defibrinogenation therapy with batroxobin, a fibrinolytic enzyme derived from venom and as yet not released from use in the USA, proved beneficial in a controlled study by Kubo et al (1988).

The efficacy of such treatments as stellate ganglion block therapy (Haug et al, 1976) remains in doubt (Simmons, 1976), and these treatments await more carefully controlled studies. Certainly a drawback to this therapy is that patients require two blocks per day for a period of 3 to 5 days. Goto et al (1979) combined stellate ganglion block with hyperbaric oxygen treatment; however, their criterion for recovery, namely, improvement over 10 dB, was too small to draw meaningful conclusions. Likewise, judgments about the efficacy of Urografin (Hirashima, 1978) or Hypaque (Shea et al, 1977) and microwave therapy (Kawamoto and Naito, 1976) await further clinical experimental work.

For ISHL patients without a clear history or physical findings suggestive of a cochlear membrane break, random surgical exploration of the ears yields a very low incidence of round window or oval window fistulas and therefore is not recommended. Many leaks will heal spontaneously, and in patients with a questionable history, a trial of bed rest and elevation is warranted to permit spontaneous healing of the ruptured membrane (Singleton et al, 1978).

Pullen et al (1979) explored the middle ears of 16 patients with ISHL associated with the marked barotrauma of scuba diving as soon as the diagnosis was made. Of 11 patients with flat hearing losses, 9 showed improvement to within 10 dB of the hearing in the normal ear. Six patients with high-frequency hearing losses, however, failed to improve with surgical exploration. Most authors agree that surgery is more useful in the control of vertigo than in the restoration of hearing.

Thus, for patients with flat sensorineural losses and clear histories of sudden hearing loss secondary to barotrauma, some evidence indicates that immediate exploration and repair are the best therapy, although patients with high-frequency losses fail to recover hearing as efficiently. In patients with a suggested but uncertain diagnosis of a fistula, a period of bed rest and head elevation may be indicated.

Follow-Up

The diagnosis of ISHL is a diagnosis of exclusion, leaving a potential for error. Therefore all patients, regardless of whether they recovered fully or not, should have a follow-up audiogram to check for progressive sensorineural hearing loss and other symptomatology at 6 months to 1 year after the loss.