Introduction

Sudden sensorineural hearing loss (SNHL) causes great concern not only in the patient, but for the physician as well. Most otolaryngologists define sudden SNHL as a loss that is greater than 30 dB in three contiguous frequencies and that occurs over a period of less than 3 days.

Sudden SNHL may be due to a number of known causes of hearing loss including bacterial labyrinthitis, meningitis, encephalitis, multiple sclerosis, syphilis, autoimmune diseases such as Cogan’s syndrome, acoustic neuroma, ototoxic drugs, otitic barotrauma, temporal bone fracture, concussion, acoustic trauma, fat embolism, leukemia, Meniere’s disease, and metastatic carcinoma. Therefore, a thorough search for potentially treatable causes of sudden SNHL must be undertaken with an exhaustive history, thorough physical examination, audiologic evaluation, and carefully selected laboratory and radiographic studies.

No cause can be found, however, for the majority of cases of sudden SNHL, and the diagnosis of idiopathic sudden SNHL is given. This chapter will focus on this diagnosis of exclusion.

Sudden SNHL is usually unilateral and frequently accompanied by tinnitus(70%) and vertigo(50%). The intensity of the vertigo roughly correlates with the degree of hearing loss. The degree of hearing loss may vary from mild to profound, and involve a part or all of the frequency range. The SNHL may be temporary or permanent. About one third of these patients awaken in the morning with a hearing loss.
Diagnostic Evaluation

Every patient suffering from sudden SNHL must be thoroughly evaluated in order to treat any underlying disorders that may be contributing to this disorder. This assessment begins with a comprehensive history which should outline the complaint (speed of onset, duration of the SNHL, and associated symptoms such as vertigo, tinnitus, aural fullness, headaches, vision changes). The history should also include questions regarding potential precipitating events preceding the SNHL such as recent air travel, strenuous exercise, weight lifting, diving, falls, other head trauma, viral illnesses, febrile states, previous otologic surgery, medications, and exposure to pesticides. Past medical history should include questions regarding diabetes mellitus, hyperlipidemia, arteriosclerosis, hypertension, and syphilis. Family history of hearing loss and other diseases should also be elicited from the patient.

The physical examination should be complete, with special attention directed toward the otolaryngologic and neurologic portions of the exam.

Audiometry should be performed in all patients presenting with sudden SNHL. Additional tests may be required to determine the site of lesion, such as auditory-evoked brainstem response (ABR) and electronystagmography, especially if vertigo is present. If retrocochlear pathology, such as an acoustic neuroma, is suspected, then magnetic resonance imaging with gadolinium (MRI), or less preferably computerized tomography (CT), is indicated to examine the internal auditory canals and posterior fossa. CT imaging should be obtained for hearing loss that may be trauma-induced.

Laboratory studies should include a complete blood cell count with differential (CBC), erythrocyte sedimentation rate (ESR), coagulation profile (including prothrombin time, partial thromboplastin time, and clotting time), electrolytes with fasting blood glucose, cholesterol and triglycerides, thyroid function tests, ACTH plasma cortisol stimulation test, serologic tests for syphilis (FTA-ABS/MHA-TP preferable to VDRL/RPR because the latter may be negative), and autoimmunologic tests such as antinuclear antibody (ANA) and rheumatoid factor (RF).

Incidence

The incidence of idiopathic sudden SNHL is estimated to be approximately 5 to 20 per 100,000 persons per year, although the incidence may be even higher because some patients who recover spontaneously from sudden SNHL may not seek medical attention. The frequency appears to increase with advancing age, ranging from 4.7 per 100,000 persons 20-30 years of age to 15.8 per 100,000 persons 50-60 years of age. The mean overall age for sudden SNHL ranges from 46-49 years of age. The incidence of bilateral involvement is quite variable (1-80%). There is no predilection to sex, and sudden SNHL does not occur in a seasonal pattern.

Etiology

Most cases of sudden SNHL are idiopathic and little is known about its pathophysiology. However, several theories have been proposed regarding the development of sudden SNHL. The
three most prevalent ones include the viral theory, vascular theory, and the membrane rupture theory.

Several studies support viruses as the etiology of sudden SNHL. Mumps, measles, herpes zoster, and infectious mononucleosis may involve the labyrinth as part of a recognized clinical disease. However, viruses that cause upper “respiratory tract infections” such as adenovirus have also been postulated to cause sudden SNHL. Unfortunately, only 20-60% of the patients suffering from sudden SNHL report recent URI symptoms, and almost 40% of the general population give history of such symptoms recently. Furthermore, unlike many upper respiratory infections, sudden SNHL is neither seasonal nor epidemic.

Other viruses have been incriminated as possible causes of sudden SNHL by increased viral titers in these patients as compared to controls, and include rubella, rubeola, influenza, cytomegalovirus, herpes simplex, parainfluenza types 1 and 3, herpes hominis, as well as the bacteria Mycoplasma pneumoniae. Recently, an etiologic relation between human spumaretrovirus and sudden SNHL was proposed. Four of 30 patients with sudden deafness had positive titers to HSRV, and only 2 of 1310 controls had positive titers to this virus. Also, 14 of 49 patients with antibody titer confirmed Lassa fever in Sierra Leone, Africa suffered acute SNHL, whereas 20 febrile controls with negative titers for Lassa fever did not experience sudden SNHL.

Additional evidence supporting the viral theory comes from temporal bone histopathologic studies. These studies have shown changes in the temporal bones of patients who had sudden SNHL consistent with, but not necessarily specific to, changes occurring with known viral disorders such as measles and mumps. These changes include atrophy of the organ of Corti with loss of hair cells and supporting elements, as well as tectorial membrane encapsulation.

The vascular theory holds that partial or complete occlusion of the cochlear vasculature may cause idiopathic sudden SNHL. There are several clinical disease states that have been shown to cause sudden hearing loss by disrupting the blood supply to the labyrinth. Sudden hearing loss can be caused by hyperviscosity syndromes such as Waldenstrom’s macroglobulinemia and polycythemia rubra vera, and be relieved by plasmapheresis and phlebotomy, respectively. Inner ear hemorrhage as a complication of leukemia or heparin and small-vessel destruction in polyarteritis nodosa have been shown to cause sudden SNHL. In addition, microemboli can cause postoperative unilateral hearing loss in about 0.1% of the patients who undergo coronary artery bypass grafting. Vascular spasm has also been implicated as a factor in sudden hearing loss, due to a reported association between migraine headaches and a temporary, reversible hearing loss.

In one study, 18 of 31 patients with sudden SNHL had evidence of hypercoagulability based on prothrombin tests. Twelve of those 18 had recovery, but only 2 of the 13 patients with normal coagulation recovered good hearing. This recovery in the patients with hypercoagulability was attributed to activation of the fibrinolytic system. For those with poor recovery, the hearing loss was thought to caused by something other than hypercoagulability. In another study, 5 of 6 patients with sudden SNHL had hypercoagulability as evidenced by increased prothrombin consumption, which is suggestive of platelet lability. Hyperlipoproteinemia has also been linked
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to sudden SNHL, due to its effects of lipid imbalance as well as its association with atherosclerosis.

Additionally, intraoperatively decreased oxygen tensions have been demonstrated in the perilymph of patients with sudden SNHL. In another study, 16 patients with sudden SNHL, compared to controls, had significant impairment in whole blood and red blood cell filterability. This suggests that diminished microcirculation (as a result of erythrocyte sludging) has a causal role in sudden hearing loss.

Although the vascular theory seems attractive in explaining sudden SNHL, there is plenty of data to cast doubt on it. The histopathologic findings of temporal bones from patients with sudden SNHL differ from those in animal models following obstruction of the labyrinthine artery. Vascular insults would result in fibrosis and new bone formation which have been infrequently seen in temporal bone studies. Furthermore, irreversible loss of hearing occurs after just 60 seconds of anoxia. This temporary vascular ischemia is not consistent with the reversible nature of idiopathic sudden SNHL in about 65% of the cases. Many who experience sudden hearing loss are young and free of any vascular disease, also casting doubt on the vascular theory. In addition, no studies to date have conclusively shown a relationship between diabetes mellitus (and its compromised vasculature) and sudden SNHL. Also, the finding of middle range or high frequency hearing loss with preservation of low frequency argues against a vascular etiology since the vasculature supplying the apical turn (lower frequency) of the cochlea lies distal to the same blood vessel supplying the basal turn (higher frequency).

The third theory is based on the rupture of the delicate inner ear membrane and fistulae of the round and/or oval window. Round and oval window ruptures are reported to occur when pressures from within (cerebrospinal pressure) or without (middle ear pressure) suddenly increase causing breaks in the cochlear membrane, resulting in sudden hearing loss. Often, there is a history of strenuous physical activity or straining, or sudden changes in barometric pressure, such as in flying or scuba diving. A perilymphatic fistula at the oval window is commonly associated with vertigo, whereas a fistula at the round window is not. There is often a fluctuating hearing loss with or without tinnitus, which may improve overnight and worsen during the day. The hearing loss may be accompanied by positional vertigo and nystagmus with the affected ear down. The fistula test is unreliable and should not be used to decide surgical candidacy.

Inner ear membrane rupture has been theorized to cause the fluctuating hearing loss with endolymphatic hydrops (Meniere’s disease), presumably due to tears in Reissner’s membrane, with resulting potassium poisoning of the cochlear sensorineural structures.

Recently, a study of 10 patients with history of sudden SNHL during life showed the presence of labyrinth capsule patencies in all instances either at the fissura ante fenestram or at the round window niche (forming connection to posterior canal ampulla). Fluorescein tracer studies have shown intra-operatively that perilymphatic fistulae can occur in the areas of the fissura ante fenestram and the fissure of the round window niche. However, the fact that patencies of the labyrinth capsule can be present without sudden SNHL argues against this data. Other histopathologic studies of temporal bones, though, have failed to reveal any evidence of inner ear membrane rupture.
Given that any one of the above theories have not been clearly prevalent suggests that sudden SNHL may simply be the end result of a multitude of possible pathophysiologic mechanisms. In addition, all three theories may actually play a role in the development of sudden SNHL. For example, a patency of the labyrinthine capsule may be a prerequisite for a viral infection that subsequently induces hyper-coagulability, resulting in hearing loss.

**Prognosis**

It is difficult to accurately predict the outcome of sudden SNHL given the low incidence and unpredictability of its natural course. Also, there are problems evaluating recovery statistics, especially for treatment protocols, because of the significant percentage of patients that recover spontaneously (65%). There are, however, some factors that have been linked with prognoses. Patients who seek medical treatment within 7-10 days after onset of the hearing loss do better than those waiting 30 or more days. In addition, the severity of the initial hearing loss has some prognostic significance, in that the severity of the hearing loss is inversely proportional to the rate of recovery. Patients with an upsloping audiogram or mid-frequency loss do better than those with high frequency or downsloping hearing loss. Both vestibular changes by ENG and the presence of vertigo herald poorer recovery rates in those patients with these findings. Elevated erythrocyte sedimentation rate (> 25) also signals a poorer prognosis. The hearing status of the opposite ear also plays a role in determining prognosis. Patients with a normal audiogram in the other ear do better than those with hearing loss in the opposite ear. Age does not appear to play a significant role in recovery from sudden SNHL except that those younger than 15 years or older than 60 have poorer recovery.

**Treatment**

Attempts to treat idiopathic sudden SNHL are difficult because the exact etiology of this disorder is unknown. As a result, any treatment is given empirically. Although various classes of treatment are available (including vasodilators, diuretics, anticoagulants, plasma expanders, corticosteroids, contrast dyes, vitamins, surgery, and combined therapy), none have been conclusively proven to be efficacious. Determining the efficacy of the treatment modality is also difficult given the relatively high rate of spontaneous recovery.

Nonspecific therapy includes bed rest, stool softeners, decreasing stressful situations, low salt diet, avoidance of alcohol, stimulants, and tobacco, avoidance of noise exposure, and elevation of the head of bed, and should be prescribed to all patients who present with sudden SNHL.

The use of vasodilators is based on the premise that it promotes blood flow and eliminates vasospasm (i.e., the cause is vascular). Drugs in this class include 5% CO2 (in the form of Carbogen), atropine, histamine, procaine hydrochloride, and papaverine hydrochloride. The facts that no study has provided conclusive evidence of their benefit and their potential side effects has limited their use.

Diuretics have been proposed for treatment of sudden SNHL because this disorder has been associated with endolymphatic hydrops, and some believe that it may be due to labyrinthine edema. No data clearly support their usage and some diuretics are ototoxic.
The use of anticoagulants, such as heparin and warfarin, is based on the idea of decreasing “blood sludge” in the vessels supplying the organs of hearing. Their use is limited by potential side effects. Plasma expanders such as low molecular weight dextran also work on the concept of decreasing “blood sludge,” by reducing blood viscosity and platelet aggregation. However, its use is potentially fatal.

Probably the most popular form of treatment for sudden SNHL is corticosteroids. Studies have shown a 78% response to steroids and a 38% response to placebo in a double blind study. They found that the steroids had the greatest benefit in patients with moderate, unilateral sudden hearing loss less than 40 years of age. Those with severe losses did not benefit from corticosteroids, and those with mild hearing losses tended to recover spontaneously. Most otolaryngologists prescribe steroids, unless contra- indicated.

Exploration of the middle ear with repair of an inner ear fistula is recommended in patients with a clear history of sudden hearing loss associated with diving, straining, altitude change, or recent otologic surgery. The role of surgery in patients who do not improve with non-surgical therapy remains controversial.

Other treatment methods include a “shotgun”(or combination) approach, vitamins, contrast dye(diatrizoate meglumine), and stellate ganglion blocks. None have proven to be conclusively effective.

**BIBLIOGRAPHY**


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