

Corridor Consult

Early and Accurate Diagnosis of Sudden Sensorineural Hearing Loss

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Case 1

An obese man, age 57 years, with a medical history of depression presented to his primary care physician with three days of sudden left-sided hearing loss. He did not report pain, tinnitus, vertigo, or external auditory canal discharge. He said that he had not experienced any recent trauma, upper respiratory infection, or loud noise exposure or taken any ototoxic medications.

His physical examination revealed normal-appearing external auditory canals and tympanic membranes. The membranes exhibited good mobility with insufflation. Findings on the Rinne test with a 512-Hz tuning fork were consistent with air conduction greater than bone conduction on the right side; the left side could not be tested because of profound hearing loss. Findings on the Weber test with a 512-Hz tuning fork were lateralized to the right side. On the basis of the patient's medical history and physical examination findings, the primary care physician suspected sudden sensorineural hearing loss, prescribed oral prednisone, and referred the patient for urgent audiologic and otolaryngologic examinations. At the four-week follow-up examination, an audiogram showed return of hearing to near baseline on the left side.

Case 2

A woman, age 54 years, with hypertension, presented to the urgent care clinic with two days of right-sided hearing loss and right ear fullness. She reported that she had recently had an upper respiratory infection. Physical examination revealed a normal tympanic membrane appearance and mobility, so a tuning-fork examination was not done. The physician diagnosed eustachian tube dysfunction (ETD) and gave the woman oral decongestants.

She was referred to the Head and Neck Surgery

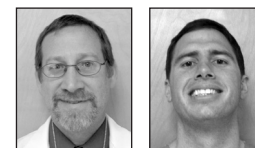
Department when there was no change in her symptoms two weeks after onset. After a thorough medical history was obtained and a physical examination, tuning fork tests were performed showing lateralization to the left ear on Weber test. An audiogram was obtained that showed severe right-sided sensorineural hearing loss, and a diagnosis of sudden sensorineural hearing loss was made. The patient was given oral prednisone and acyclovir. Later, magnetic resonance imaging (MRI) with gadolinium enhancement of the internal auditory canals revealed no retrocochlear lesion. A follow-up audiogram obtained two months after initial presentation showed no improvement in the patient's hearing.

Discussion

These case presentations illustrate the difficulty of accurate diagnosis of idiopathic sudden sensorineural hearing loss (SSNHL), treatment of patients who present with it in the primary care setting, and the variable prognosis for the disorder. Briefly, SSNHL is defined as a 30-dB hearing loss in three consecutive frequencies whose onset is less than three days. Its incidence in the US has been reported at 5 to 20 cases per 100,000 persons annually.¹ Although numerous etiologies and treatments have been considered, most researchers agree that SSNHL is likely to be of vascular, immunologic, or viral origin.

SSNHL is regarded as an otologic emergency, and the time between symptom onset and treatment initiation is one of the most important prognostic factors. Most studies report the greatest recovery of hearing when corticosteroids are initiated within the first one to two weeks after symptom onset and little if any benefit when initiated four weeks or more after the onset of symptoms.² Ideally, corticosteroid therapy should be started as soon as possible, with an audio-

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gram performed within 24 to 48 hours to document the hearing loss. The severity of the hearing loss at presentation is directly proportional to the likelihood of recovery. Those with mild losses usually obtain full recovery, and those with profound loss rarely do so.^{1,2} Other poor prognostic indicators are the presence of vertigo and age <15 years or >60 years.

Primary care physicians, in the Emergency Department, Urgent Care Clinic, or Outpatient Clinic, are often the first clinicians consulted for sudden hearing loss, making accurate diagnosis and treatment critical because delay can have serious consequences for prognosis. A recent study that we conducted revealed that 33 of 53 (63%) patients whose hearing loss was eventually diagnosed as SSNHL were initially given an incorrect diagnosis by the primary care physician. Most often, that diagnosis was ETD. This led to an average delay in referral for audiologic and otolaryngologic examination of 20.8 days.³

As seen in the illustrative cases, the hearing loss is incorrectly attributed to ETD and other diagnoses such as otitis media with effusion, because many patients with SSNHL have had a recent upper respiratory infection, and this acts as a red herring. Ear fullness is also

a common presenting symptom and often attributed by patients and clinicians to impaction of cerumen or congestion from allergies.² Furthermore, monocular otoscopy is the only option for otologic examination by the primary care physician. Although it remains a very important diagnostic tool, it decreases the ability to accurately assess a normal tympanic membrane, adequately clear the external auditory canal of cerumen, and insufflate to assess mobility as compared with binocular otoscopy. Simple Weber and Rinne tuning-fork tests are available yet are not routinely performed by most primary care physicians.

Although there are a multitude of potential causes of SSNHL, most cases are idiopathic; therefore, focused history, physical, and ancillary testing are essential. The first goal of evaluation is to rule out conductive loss by a thorough examination of the tympanic membrane and assessment of its mobility. The most critical step may be the use of a tuning fork to conduct the Weber and Rinne examinations. In the Weber test, a 512- or 1024-cycle tuning fork is placed on the forehead, on the premaxilla, or between the incisor teeth. If the sound is localized by the patient to the contralateral side of the involved ear, then the hearing loss is sensorineural. In unilateral conductive loss, the sound is localized to the involved side. The Rinne test, which compares loudness of sound when a tuning fork is placed on the skull versus near the external auditory canal, may supplement the diagnostic information obtained from the Weber test.

Although they are not good independent tools for screening for hearing loss, the Weber and Rinne tests should be used primarily to differentiate sensorineural from conductive hearing loss to accurately distinguish SSNHL from other causes of sudden hearing loss such as ETD and otitis media with effusion, which will not cause the sound to be localized in the contralateral ear. In our study, there was evidence for the efficacy of Weber and Rinne tests for patients with suspected SSNHL: Nine of the ten patients who underwent a documented tuning-fork examination performed by a primary care physician were initially given an accurate diagnosis of SSNHL, but of the 43 for whom no documentation of a tuning-fork examination was found in the chart, only 11 were given accurate diagnoses.

The goal of the initial evaluation of patients with suspected SSNHL is urgent referral to an audiologist and head and neck surgeon so that further workup can be conducted as necessary (see Figure 1). In patients who do not have contraindications to systemic

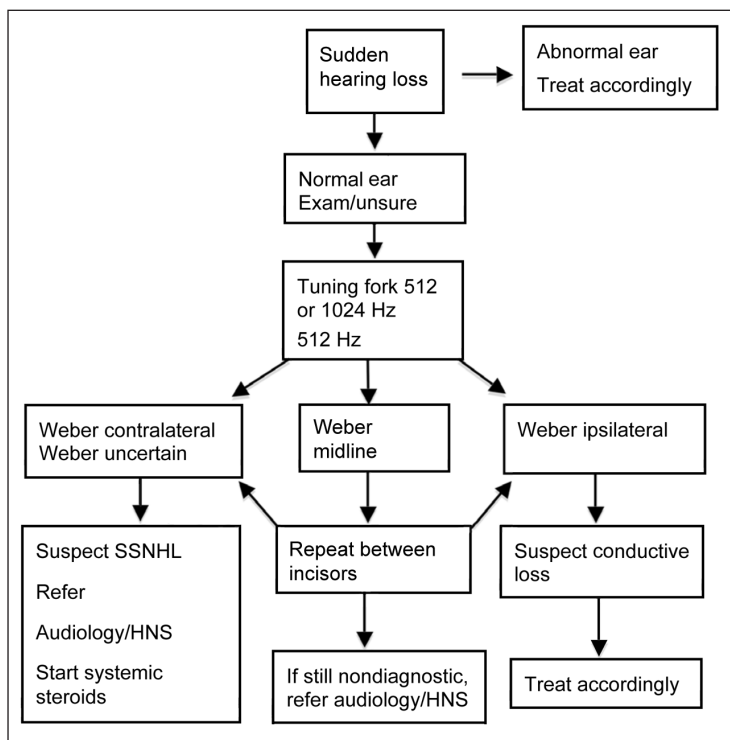


Figure 1. Clinical algorithm for evaluation of patients with suspected sudden sensorineural hearing loss (SSNHL).

HNS = Head and Neck Surgery

steroid treatment, medical treatment with oral steroids for 10 to 14 days should be carried out. There is no agreed-on dose or duration of treatment; many clinicians recommend starting with 60 mg of prednisone for five days, followed by a taper by 10 mg every two days. Medical therapy remains controversial because spontaneous recovery of hearing has been reported in 45% to 65% of patients with SSNHL and because good clinical studies are lacking. Wilson and colleagues⁴ performed a double-blind clinical study and found that steroids were effective in achieving at least partial recovery of hearing in 61% of patients, compared with only 32% who achieved recovery with the use of placebos. Intratympanic application of steroids to the middle ear has proven to be effective in some patients in whom SSNHL had been refractive to oral steroids. New research is examining the efficacy of intratympanic dexamethasone as first-line therapy. In a recent study, Battaglia and colleagues⁵ showed that intratympanic dexamethasone and high-dose prednisone taper (HDPT) used in combination resulted in higher rates of hearing recovery and better quality of hearing recovery than HDPT alone. In the future, initial treatment with intratympanic steroids may be indicated. At the conclusion of their study, Battaglia and colleagues recommended that treatment be initiated as quickly as possible, ideally within ten or fewer days of onset of hearing loss and that referral be made to an otolaryngologist who is comfortable with transtympanic injection. This reinforces the algorithm of accurately identifying SSNHL by using a tuning fork, starting oral steroids when not contraindicated, and providing urgent referrals to audiology and Head and Neck Surgery Departments for further treatment.

There is no evidence that antiviral medications make any difference in hearing outcomes. Two prospective, randomized, double-blind, placebo-controlled, multicenter clinical trials showed that antiviral medication was no better than corticosteroid alone in the treatment of SSNHL.⁵

A thorough discussion of all possible etiologies and diagnostic tests for sudden sensorineural hearing loss is beyond the scope of this article, but there are some basics to be aware of: Gadolinium-enhanced MRI examinations are routinely obtained in all patients with asymmetric sudden hearing loss from which the patient does not recover after four weeks, because vestibular schwannoma rarely presents as sudden hearing loss. Obtaining a thorough medical history investigating possible causes, including autoimmune

diseases, inner ear trauma, ototoxic medications, vasculopathies, and viral exposures, can eliminate the need for exhaustive diagnostic testing. Some advocate administering the fluorescent treponemal antibody absorption test or the microhemagglutination test for *Treponema pallidum* because syphilis is a potentially treatable cause of SSNHL. Furthermore, Ménière disease can initially present with sudden hearing loss, but the natural course usually defines these distinct entities. In rare cases, sudden hearing loss can be the initial presentation of multiple sclerosis, and MRI is frequently diagnostic.

Conclusion

Because most times SSNHL is idiopathic despite extensive diagnostic testing, the focus of initial evaluation of patients with sudden hearing loss is to distinguish a sensorineural from a conductive loss. If this cannot be reliably done with otoscopic examination, the use of the Weber and Rinne tuning-fork tests can help differentiate between the two. Once a sensorineural hearing loss is confirmed or suspected, urgent referral for audiologic and head and neck surgery evaluation should be made. In patients who can tolerate systemic steroids and are aware of the potential side effects, medical treatment with oral steroids should be initiated. Accurate diagnosis and early treatment has been shown to lead to better hearing outcomes in SSNHL. ❖

Disclosure Statement

The author(s) have no conflicts of interest to disclose.

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