Idiopathic Sudden Sensorineural Hearing Loss

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A healthy 58-year-old woman answers the telephone and realizes that her hearing is diminished on the left side. She notices aural fullness and loud tinnitus in the affected ear. Later that day she has several hours of mild vertigo that clears by the following morning. Over the next few days, repeated self-administered ear cleaning with the use of an over-the-counter kit does not relieve the symptoms. How should she be evaluated and treated?

The Clinical Problem

Idiopathic sudden sensorineural hearing loss (i.e., unexplained unilateral sensorineural hearing loss with onset over a period of less than 72 hours) has an estimated incidence between 5 and 20 per 100,000 persons per year.1 This is likely to be an underestimate, since many who recover quickly never seek medical attention.2 Several large case series, including a total of approximately 7500 cases in the United States, Europe, and Japan,1,3-11 indicate that sudden sensorineural hearing loss typically occurs between 43 years and 53 years of age, with equal sex distribution. Vestibular symptoms are present in 28 to 57% of patients. The likelihood of recovery of hearing has been reported to vary with the severity of hearing loss at presentation: patients with mild losses usually achieve full recovery, those with moderate losses often show some spontaneous recovery but seldom have a full recovery unless treated, and those with severe-to-profound hearing losses rarely show spontaneous improvement or make a full recovery.1,4,12 The prognosis for recovery of hearing also seems to be worse in older patients and those with associated vestibular symptoms.1,3-5

Approximately 1% of cases of sudden sensorineural hearing loss are due to “retrocochlear” disorders that may be related to vestibular schwannoma, demyelinating disease, or stroke.3 Another 10 to 15% are due to another identifiable cause, such as Meniere’s disease, trauma, autoimmune disease, syphilis, Lyme disease, or perilymphatic fistula.4,5,13,14 The remainder are idiopathic and almost exclusively unilateral. Rare cases of bilateral sudden deafness most often reflect a psychiatric (“functional”) cause or a neurologic process (e.g., neoplastic dural infiltration of the posterior cranial fossa, paraneoplastic syndrome, or encephalitis); transient bilateral sudden sensorineural hearing loss may also result from a sudden drop in intracranial pressure during a spinal tap or after intracranial surgery.

A common problem in sudden sensorineural hearing loss is delay in diagnosis. Ear fullness, a common presenting symptom, is often attributed by patients and clinicians to impaction of cerumen or congestion from upper respiratory illness or allergies. Insofar as evidence suggests that permanent hearing loss is more likely when treatment is delayed, it is important that a diagnosis of sudden sensorineural hearing loss is made as early as possible.
Screening for hearing loss can be performed by telephone (e.g., by a clinic nurse). The patient should be explicitly asked whether the hearing is reduced. The patient can move the phone from ear to ear or crinkle a few hairs next to the ear on each side to check for hearing asymmetry. To assess whether an apparent hearing asymmetry is likely to indicate sensorineural hearing loss, the patient should be instructed to hum and to report whether the sound is louder in one ear than in the other. In conductive hearing loss, sound lateralizes to the blocked ear, whereas in sensorineural loss, sound lateralizes to the good ear.

Office evaluation of hearing can be done by softly whispering simple words or numbers in each ear and asking the patient to repeat them aloud. Inspection of the ear canals and tympanic membranes with the use of a pneumatic bulb to assess drum mobility (to rule out middle-ear effusion) is essential. If cerumen cannot be adequately removed to visualize the tympanic membrane, then otolaryngology consultation is indicated. The Weber and Rinne tests should be performed using a 512-Hz tuning fork (Fig. 2). A focused neurologic examination should assess for evidence of central or vestibular system dysfunction. Especially relevant are assessments of ocular motility and sinusoidal gaze tracking by having the patient keep the gaze fixed on a target as the head is passively rotated side to side and up and down (second, third, fourth, and sixth cranial nerves, brain stem, and cerebellum); facial sensation for light touch and pinprick (fifth cranial nerve); voluntary and mimetic facial expression (seventh cranial nerve); presence of spontaneous, gaze-evoked, or positional nystagmus (eighth cranial nerve, cerebellum, and brain stem); extremity coordination and rapid alternating movements (cerebellum); and postural and gait stability during the Romberg test and tandem gait.

**STRATEGIES AND EVIDENCE**

**DIAGNOSIS**

Sudden sensorineural hearing loss is often accompanied by intense aural fullness or pressure, as well as tinnitus. If the patient has partial hearing in the affected ear, the sound may seem harsh and distorted (as if from a “blown” loudspeaker). Because aural fullness is nonspecific and often has nontologic causes (e.g., temporomandibular joint dysfunction or upper-airway congestion), the first step in diagnosis is to determine whether symptoms are caused by hearing loss (Fig. 1).

![Management of Sudden Sensorineural Hearing Loss (SNHL)](image-url)

be considered and prompt referral made to physicians with expertise in otolaryngology.

**AUDIOMETRY**

A complete audiogram, including threshold measurements of bone- and air-conducted pure tones and speech audiometry, is needed for definitive diagnosis in patients in whom hearing loss or asymmetry is suspected. Hearing threshold and speech audiometry assess the loudness and the clarity of hearing, respectively (Fig. 3). In sensorineural hearing loss, the sensitivity to sounds delivered as bone-conducted stimuli and the sen-
Sensitivity to sounds delivered as air-conducted stimuli are equal in the affected ear, but both are reduced (i.e., the threshold is elevated). In conductive hearing loss, the bone conduction is normal, but the air-conducted thresholds are worse (elevated) in the affected ear.

Gadolinium-enhanced magnetic resonance imaging (MRI) of the temporal bone and brain is warranted in cases of acute sensorineural hearing loss to rule out a “retrocochlear” abnormality (e.g., neoplasm, stroke, or demyelination). In patients who cannot have brain MRI, alternatives include computed tomographic scanning, auditory brainstem–response audiometry, or both, although these are less sensitive than MRI for the detection of retrocochlear abnormalities.

**Figure 2. Weber and Rinne Tests.**

The Weber test is conducted by ringing a 512-Hz tuning fork, placing it firmly on the middle of the forehead, and asking the patient where the tones are heard best: left, right, or center. If the hearing is symmetric, the tone is centered. The Rinne test is performed by comparing air conduction to bone conduction on each side.

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**Medical Treatment**

**Oral Corticosteroids**

The current standard treatment for sudden sensorineural hearing loss is a tapered course of oral corticosteroids (prednisone or methylprednisolone). However, data to support this recommendation are limited. An initial randomized, placebo-controlled trial involving 67 subjects with sudden sensorineural hearing loss showed significantly higher rates of improvement among patients randomly assigned to either oral methylprednisolone or dexamethasone (tapered over a period of 10 to 12 days) than among patients assigned to placebo (61% vs. 32%, P<0.05). However, the rate of improvement in the corticosteroid-treated cohort was
similar to the rate of recovery without treatment (65%) that was reported elsewhere. More recently, another randomized trial failed to show improvement in hearing with the use of corticosteroids as compared with the use of carbogen (an inhalational vasodilator composed of 5% carbon dioxide and 95% oxygen) or placebo. This study was seriously underpowered: the 41 subjects were randomly assigned to one of four groups (oral corticosteroid, oral placebo, inhalational carbogen, or inhalational placebo), yielding only 9 to 11 subjects per group for the 5 days of treatment.

A recent Cochrane Review based on these two trials, as well as another systematic review, both concluded that the efficacy of corticosteroid therapy for sudden sensorineural hearing loss remains unproven. However, because a short course of oral corticosteroids is generally associated with acceptable adverse-event rates and may confer benefit, routine practice at present is to administer a course of oral corticosteroids tapered over a period of 10 to 14 days (e.g., 60 mg of prednisone daily for 4 days, followed by a taper by 10 mg every 2 days). Data comparing different doses or durations of corticosteroid therapy are limited. A double-blind, randomized, controlled trial comparing a 7-day prednisone taper with a 3-day pulse of 300 mg of dexamethasone per day (followed by 4 days of placebo) showed no significant difference in the rates of hearing recovery. Side effects
of corticosteroid treatment include elevated blood sugar levels or blood pressure, mood changes, weight gain, gastritis, and sleep disturbances; these typically resolve as treatment is tapered or discontinued.

A natural-history study has shown that spontaneous recovery occurs almost exclusively within the first 2 weeks after the onset of sudden sensorineural hearing loss. Studies assessing the relationship between the duration of sudden sensorineural hearing loss before treatment and outcomes have reported the greatest recovery of hearing when corticosteroids are initiated within the first 1 to 2 weeks after symptom onset, and little if any benefit when initiated 4 weeks or longer after the onset of symptoms. Some patients have a very rapid and substantial recovery of hearing within the first 48 to 72 hours after the initiation of corticosteroids, some improve steadily after the onset of treatment and continue to improve after treatment is completed, and others have no improvement; the proportion of patients in each of these groups is uncertain. The sooner a response is seen, the better the overall prognosis appears to be. Patients who show no improvement at the completion of treatment with corticosteroids have a poor prognosis. Symptoms of aural fullness and tinnitus tend to abate gradually, regardless of hearing outcome.

Since there may be only a 2-to-4-week window for effective treatment of sudden sensorineural hearing loss, it is prudent to start therapy as soon as possible. Ideally, an audiogram should be performed before or within 24 to 48 hours after the initiation of treatment to document the sensorineural hearing loss and its magnitude. If imaging studies cannot be obtained promptly, treatment should be initiated pending this evaluation. Improvement in hearing after treatment with corticosteroids does not eliminate the need for imaging. Demyelinating lesions can have a fluctuating course or a transient response to corticosteroids, and occasionally, acute enlargement of an acoustic neuroma (e.g., from hemorrhage) can result in sudden sensorineural hearing loss that improves over days or weeks.

**INTRANATMYMPANIC STEROID INJECTIONS**

In lieu of oral corticosteroids, some otolaryngologists recommend local corticosteroid therapy for sudden sensorineural hearing loss, administered either by intratympanic injections (methylprednisolone or dexamethasone) or as eardrops by means of a ventilating tube or a “wick” running from a ventilating tube to the round window membrane in the medial wall of the middle ear. The rationale for intratympanic corticosteroid treatment is that it delivers a high concentration to the target tissue with minimal systemic exposure. Data to support the use of intratympanic corticosteroids for primary therapy are currently limited to case series reporting success rates similar to those reported with oral therapy. Among these are anecdotal cases in which hearing improved despite a 6-week or greater delay in treatment. Several reports of intratympanic corticosteroid therapy as “salvage therapy” in patients who did not improve with the use of oral treatment have suggested that intratympanic therapy may yield better hearing outcome than placebo or no treatment, but these studies have been small or retrospective. Intratympanic therapy is much more costly than oral therapy, with charges in excess of $2,000 for a course of treatment at some centers.

**OTHER THERAPIES**

Randomized trials comparing corticosteroids alone to corticosteroids plus antiviral agents for sudden sensorineural hearing loss have failed to show an added benefit for antiviral therapy; none of these studies included a placebo group. Other treatments, including volume expanders, anticoagulants, inhalational vasodilators, herbal remedies, and hyperbaric oxygen, have been suggested, but adequately powered randomized trials are lacking to support clinical benefit with their use. A retrospective observational study of 112 patients with sudden sensorineural hearing loss who were treated with tapered corticosteroids after a high-dose intravenous bolus of either 600 mg or 1200 mg of hydrocortisone showed a significantly greater incidence of complete recoveries in the higher-dose group, but randomized trials of treatment with high doses of intravenous corticosteroids are lacking.

Certain recommendations should be made to all patients with new monaural hearing loss to minimize the risk of hearing loss in the good ear. First, scuba diving should be prohibited because of the associated risk of ear injuries; these include rupture of the tympanic membrane (reported in 5.9% of 709 experienced scuba divers) as well as permanent disabilities such as hearing loss, tinnitus, and balance problems (in 2.3% of 709 experienced scuba divers). Even patients who have regained full hearing after an episode of sudden...
sensorineural hearing loss should be wary of scuba diving, since it is uncertain whether a history of such hearing loss increases vulnerability in the affected ear. Second, noise protection should be used when indicated. Acoustic trauma can result from exposure to loud music and the noise of power tools and yard equipment, for example. Earplugs or earmuffs designed for noise protection are inexpensive, widely available, and very effective when used properly. Finally, an immediate otolaryngologic evaluation (i.e., within 24 hours after the onset of symptoms) is recommended to assess any symptoms in the good ear.

PROGNOSIS
Although long-term data are lacking, there is concern that persons who have had an episode of sudden sensorineural hearing loss may be at higher risk of accelerated age-related hearing loss in the future. There is no evidence that sudden sensorineural hearing loss is more likely to occur in the contralateral ear of an affected person than it is to occur in the general population. A recent prospective cohort study suggested a slightly greater risk of stroke (adjusted hazard ratio over a 5-year period of follow-up, 1.64; 95% confidence interval, 1.31 to 2.07) among patients with sudden sensorineural hearing loss than among a comparison group of patients who had undergone appendectomy, although this warrants confirmation.

Patients who do not recover symmetric hearing permanently lose their ability to localize where sounds are coming from. These patients are at a disadvantage in adverse listening situations (e.g., situations with high ambient noise, poor acoustics, multiple talkers, or speakers who have foreign accents). Though both wearable and implantable devices are available to receive sound on the bad side and route it to the better ear, conventional hearing aids are of limited use if the contralateral ear is normal; however, their use on one or both ears may be beneficial if the contralateral ear is not normal.

It is commonly advised that patients with sudden sensorineural hearing loss have audiometric monitoring repeated over the course of a year (e.g., at 2 months, 6 months, and 12 months after the onset of the hearing loss) to document recovery, guide aural rehabilitation (especially the fitting of hearing aids), and monitor for signs of relapse in the affected ear or development of hearing loss in the contralateral ear, which would warrant consideration of other diseases (e.g., Meniere's disease or autoimmune disease) that may have been misdiagnosed as sudden sensorineural hearing loss. Particularly in patients with low-frequency hearing loss, the sudden loss may be an early manifestation of Meniere's disease. If so, further fluctuations in hearing and attacks of vertigo are likely to occur within a 3-year period. Meniere's disease has also been reported to occur as a late outcome in 4 to 8% of cases of sudden sensorineural hearing loss (i.e., years after onset)\(^3\)\(^6\)

AREAS OF UNCERTAINTY
The causes of sudden sensorineural hearing loss remain uncertain, as does the specific site of inner ear damage. Oral corticosteroids are routinely used for primary treatment of sudden sensorineural hearing loss, although data are limited to support their use; there are also limited data to support the use of intratympanic corticosteroids for primary therapy or for treatment of those whose hearing did not improve with initial therapy. A clinical trial sponsored by the National Institutes of Health that compares oral with intratympanic corticosteroid treatment for primary therapy is underway (ClinicalTrials.gov number, NCT00097448). Randomized trials are also needed to assess various other corticosteroid regimens and to evaluate treatments other than corticosteroids. Longitudinal studies of long-term outcome are lacking for any treatments of sudden sensorineural hearing loss.

GUIDELINES
There are no published guidelines for the evaluation or management of sudden sensorineural hearing loss.

CONCLUSIONS AND RECOMMENDATIONS
Patients such as the woman described in the vignette, who present with unilateral ear blockage or fullness, should promptly be evaluated for possible sudden sensorineural hearing loss. Sudden sensorineural hearing loss is considered by otologists to be a true otologic emergency, given the observation that there is less recovery of hearing when treatment is delayed. Hearing loss can be assessed over the telephone (e.g., by asking the patient to move the phone from ear to ear for comparison).
comparing). If hearing is reduced, the patient should be asked to hum and to report on which side the sound is louder; although this test is imperfect, sound usually localizes toward a conductive loss and away from a sensorineural loss. An office evaluation and complete audiogram is indicated if there is any suspicion of sensorineural hearing loss. If an audiogram reveals unilateral sensorineural hearing loss, then an MRI with gadolinium is needed to rule out a retrocochlear abnormality, such as demyelinating disease, neoplasm, or stroke. In the absence of these findings, sudden sensorineural hearing loss is the presumed diagnosis. Treatment should not be delayed, even if imaging cannot be obtained promptly.

Although supporting data are limited, corticosteroid therapy (usually a 2-week burst and taper of oral prednisone, starting at 60 mg per day, or equivalent doses of methylprednisolone) is the current standard of care, according to a randomized trial suggesting that it may improve or restore hearing and because of the absence of other known effective therapies. Intratympanic injections of corticosteroids may be an alternative, particularly for patients who have or are at high risk of complications from oral therapy, although evidence to support this strategy is even more limited.

After sudden sensorineural hearing loss has led to damage in one ear, it is imperative to protect the better-hearing ear from excessive pressure (e.g., scuba diving should be avoided) or exposure to noise. Development of hearing loss, tinnitus, pain, or discharge in the good ear warrants an immediate otolaryngologic evaluation.

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REFERENCES


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