

# Sudden sensorineural hearing loss

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**A** 36-year-old woman presents to the clinic and reports left-sided hearing loss. She first noticed it five days prior when she could not use the telephone on her left side. She notes a buzzing sound and “plugged” left ear. She says she does not have vertigo or neurologic symptoms. Examination by otoscopy is normal. There is lateralization to the right for the Weber test, and air conduction using a tuning fork (512 Hz) is greater than bone conduction bilaterally.

## What is the likely diagnosis?

Given the patient’s symptoms and findings on examination, the most likely diagnosis is sudden sensorineural hearing loss. Sudden sensorineural hearing loss is subjective hearing loss that occurs acutely over 72 hours and is usually unilateral.<sup>1</sup> The disturbance is thought to originate in the cochlea or acoustic nerve. The pathophysiology of sudden sensorineural hearing loss is unknown; viral and vascular causes have been proposed.

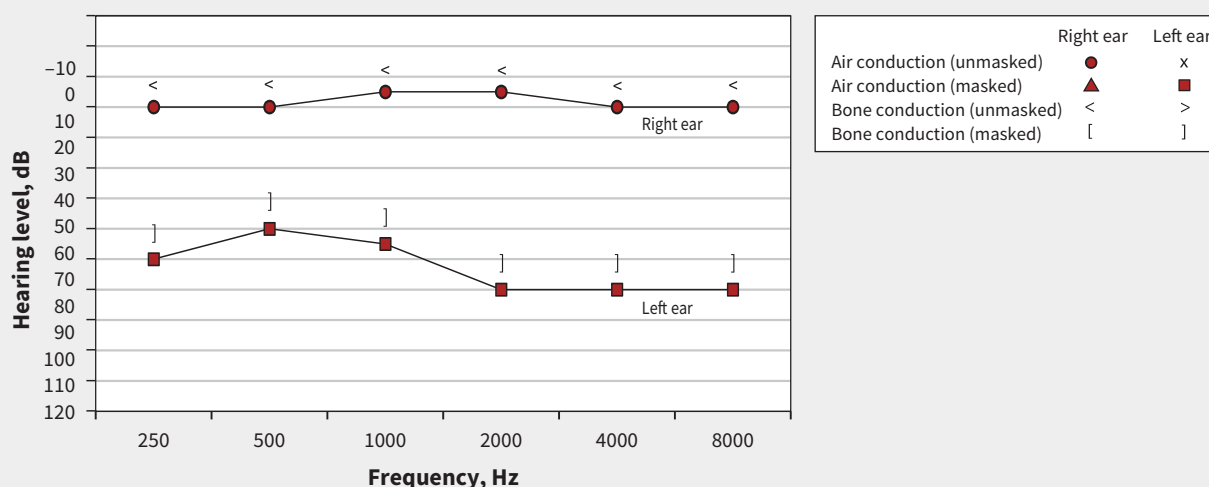
The incidence of sudden sensorineural hearing loss is estimated between 5 and 20 cases per 100 000. Men and women are equally affected, and all age groups can be affected; however, patients in their 40s and 50s make up the most affected age range.<sup>1-3</sup>

Sudden sensorineural hearing loss must be distinguished from the more common presentation of conductive hearing loss caused by middle ear effusion and alternate sensorineural causes. A Weber test is the critical first step, because lateralization to the opposite side confirms a sensorineural hearing loss. If the Weber test shows lateralization to the affected ear, a conductive cause should be suspected.

In addition to helping make the diagnosis of sudden sensorineural hearing loss, history and physical examination are important for excluding more severe pathologies, such as vestibular schwannoma or stroke.<sup>1</sup>

Clinicians should inquire about the following: otologic symptoms (vertigo, tinnitus, aural fullness) that may suggest sudden sensorineural hearing loss, Ménière disease or vestibular schwannoma; neurologic symptoms that may suggest stroke or multiple sclerosis; constitutional symptoms that may suggest malignant disease or infection (e.g., Lyme disease); and family history of stroke or autoimmune disease that may suggest one of these causes.<sup>1</sup>

Sudden sensorineural hearing loss typically presents with an otherwise unremarkable history and normal otoscopic examination.<sup>2</sup> The timeline of symptoms is important, particularly in



**Figure 1:** Audiogram of a 36-year-old woman with left-sided hearing loss, confirming its sensorineural nature. The normal hearing level is 25 dB and above: in the right ear, the pure tones are at 15 and 20 dB, whereas in the left ear, the pure tones are heard between 60 and 80 dB (confirming a substantial hearing loss). The symbols in the legend represent typical markings on audiograms and are not all represented in this audiogram.

distinguishing sudden sensorineural hearing loss from conditions with overlapping symptoms, such as Ménière disease, which is more commonly associated with fluctuating hearing loss.

### What investigations and referrals are indicated for the initial workup?

Sudden sensorineural hearing loss is an urgent otologic problem, because early treatment is thought to improve prognosis, although the evidence is weak.<sup>1,4,5</sup> Patients should be referred immediately to an otolaryngologist and should be seen ideally within one week. In the meantime, an audiogram should be obtained to confirm the nature and degree of the hearing loss and to provide a baseline for comparison following treatment (Figure 1). Many audiologists will perform this on an emergent basis; however, delay in obtaining an audiogram should not delay referral to otolaryngology. In areas where expeditious audiogram and otolaryngology referrals are not feasible, the family physician should begin treatment.<sup>1,2</sup>

Although sudden sensorineural hearing loss is not usually caused by a slow-growing vestibular schwannoma, magnetic resonance imaging (MRI) of the internal auditory canals should be ordered to rule this out; however, the referral can be nonurgent.<sup>1</sup> Laboratory investigations are not routinely recommended in sudden sensorineural hearing loss.<sup>1</sup>

### What treatment options should be considered?

Literature findings report varying rates of recovery without treatment; spontaneous improvement in hearing occurs in 31%–65% of cases, and this typically occurs within the first two weeks of treatment.<sup>2,3,6</sup> Corticosteroids are first-line therapy, despite limitations in the evidence for their efficacy, and should be started unless a major contraindication is present. Multiple protocols for this therapy are available and based primarily on expert opinion.<sup>7</sup> It is reasonable to prescribe prednisone (1 mg/kg/d, to a maximum of 60 mg) for 10 to 14 days.<sup>1,7</sup> A 2013 Cochrane systematic review did not show conclusive efficacy for

corticosteroids taken orally; only three trials met the inclusion criteria, and all three were at high risk of bias. Of the three trials included, only one trial showed a benefit from use of steroids.<sup>6</sup> The recommendation to start treatment early with corticosteroids is supported by two relatively small retrospective studies that showed poorer prognosis when corticosteroids are started more than seven days<sup>4</sup> and one month<sup>3</sup> following onset of sudden sensorineural hearing loss.

Otolaryngologists may prescribe intratympanic steroids as an alternative to treatment taken orally, or as adjuvant or salvage therapy.<sup>1</sup>

### Case revisited

Given the high suspicion for sudden sensorineural hearing loss, the patient was prescribed prednisone taken orally (60 mg daily) for 10 days. She was sent for an urgent audiogram, which confirmed the sensorineural nature of her hearing loss (Figure 1). She was also referred urgently to an otolaryngologist who performed intratympanic steroid injections, because there was only partial recovery of her hearing. The injections provided minimal benefit, and she was left with a moderate to severe hearing loss. An MRI showed no acoustic neuroma.

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