

# Can you hear me?

## *Sudden sensorineural hearing loss in the emergency department*

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### Case description

A 62-year-old man presents to the emergency department complaining of loss of hearing in his left ear. When he awoke that morning, he felt a sensation of fullness in the left ear and later realized that he could barely hear his wife's voice. He also complains of ringing in the left ear and slight nausea. His hearing is unaffected in the right ear. His past medical history is unremarkable. He reports no recent illness or infection, and has no history of trauma. On physical examination, he has normal vital signs. There is no obstructing cerumen, and the tympanic membrane is clear and intact. Weber test results reveal lateralization to the unaffected ear, suggesting a sensorineural type of hearing loss. What is your next step?

Sudden sensorineural hearing loss (SSNHL) is a potentially devastating health problem leading to serious morbidity and negatively affecting a patient's quality of life. It is considered an otolaryngologic emergency and is defined as the acute loss of hearing of 30 dB or greater over 3 frequencies within a 72-hour period.<sup>1-3</sup> Population studies of SSNHL show a wide age distribution with an average age of 50 to 60 years, and that it affects men and women equally.<sup>4,5</sup> The incidence is about 5 to 20 cases per 100 000 people.<sup>1,5</sup> There are many differential diagnoses for SSNHL, but in about 70% to 90% of cases, the cause is idiopathic.<sup>1,3</sup> Many patients who experience sudden hearing loss might fail to seek medical attention immediately, as they might not take their symptoms seriously, resulting in a delay to initiation of therapy.<sup>1</sup>

### Pathophysiology

A 2010 review of the literature revealed that idiopathic causes remained the most predominant at 71%.<sup>6</sup> Additional suspected causes include infection (12.8%), otologic disease (4.7%), trauma (4.2%), vascular or hematologic causes (2.8%), neoplasms (2.3%), and other causes (2.2%).<sup>6</sup> There is very little hard evidence to support the

most common theories behind the pathophysiology of idiopathic SSNHL, including impairment of the blood supply to the labyrinth, tympanic membrane ruptures, viral infection of the labyrinth, and autoimmune-mediated cochlear malfunction.<sup>7</sup> Although the evidence is still unclear, it is thought that infection is the second most common cause of SSNHL, and the viral causes include influenza B, enterovirus, mumps virus, varicella-zoster virus, and herpes simplex virus type 1.<sup>6,8</sup>

### Clinical manifestations

Patients with idiopathic SSNHL present predominantly with unilateral hearing loss.<sup>1,4</sup> The hearing loss occurs over a period of seconds to days, most commonly within a 72-hour time frame.<sup>6</sup> Bilateral hearing loss occurs less than 2% of the time.<sup>4</sup> Patients might first note the hearing loss upon waking.<sup>2,6</sup> Associated symptoms commonly include tinnitus, aural fullness, and vertigo.<sup>1,3,4</sup> The hearing loss can occur across some or all frequencies and can range from mild to severe.<sup>3,4,6,8</sup>

### Diagnosis and workup

The history should include a detailed exploration of the patient's symptoms, including tinnitus, pain, aural fullness, previous hearing loss, ear discharge, fever, upper respiratory tract symptoms, headache, and focal neurologic symptoms. Recent trauma, flying or scuba diving, and ototoxic medication use should be determined. The physical examination must differentiate between conductive and sensorineural hearing loss<sup>3</sup> and attempt to exclude more serious causes, such as stroke or neoplasm. The physical examination should always include otoscopic evaluation and the removal of cerumen, if present, to assess the tympanic membranes.<sup>1</sup> Basic screening tests for hearing loss should include a whispered voice test as well as tuning fork tests such as the Rinne and Weber tests.<sup>9-11</sup> In sensorineural hearing loss, the Weber test should demonstrate sound lateralizing to the unaffected ear. If a tuning fork is unavailable, a simple hum test can be performed: the patient is asked to hum and if there is lateralization of the sound toward the unaffected ear, this suggests sensorineural hearing loss.<sup>1</sup> In conductive hearing loss, the sound would lateralize to the affected ear with humming.<sup>1</sup> The neurologic examination should focus on cranial nerves and ocular function, as well as cerebellar testing to determine whether there is central or peripheral vestibular dysfunction, or any posterior circulation involvement.<sup>1,8</sup>



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Routine bloodwork is not indicated in the workup of SSNHL unless the clinical history suggests a specific cause that could be elucidated by laboratory investigations.<sup>3</sup>

The most recent evidence suggests that patients presenting with SSNHL should undergo magnetic resonance imaging at some point during their assessment to detect retrocochlear pathology, or structural lesions of the vestibulocochlear nerve, brainstem, or brain.<sup>3</sup> Unfortunately, the retrocochlear space is poorly viewed on computed tomography of the head.<sup>3</sup>

A definitive diagnosis of SSNHL should be made with audiometric testing. Audiometry distinguishes between conductive and sensorineural hearing loss, and helps to determine frequency-specific hearing thresholds.<sup>3</sup>

### Treatment

Given the potentially devastating effects associated with hearing loss, early recognition and treatment are essential. Treatment should be directed toward the specific underlying pathogenesis. If the cause is idiopathic, there are few proven therapies and the current standard is corticosteroid therapy.<sup>4,12</sup> Primary care and emergency physicians can initiate this therapy and refer the patient to an otolaryngologist urgently for audiometry and ongoing management. Antiviral medications are not currently indicated, despite the implication of certain viruses.<sup>13</sup>

**Oral corticosteroids.** Corticosteroids are thought to improve idiopathic SSNHL by reducing inflammation and edema in the inner ear.<sup>4,12,14</sup> Current clinical practice guidelines recommend a course of oral corticosteroids involving 1 mg/kg body weight of prednisone per day (maximum 60 mg per day) for 7 to 14 days, gradually tapered over a similar period thereafter.<sup>3</sup> As an alternative, 10 mg of dexamethasone per day or 48 mg of prednisolone per day can be used.<sup>3</sup> Comparison between different corticosteroids has shown nonsuperiority.<sup>12</sup>

If using corticosteroid therapy, it should be initiated within the first 1 to 2 weeks following symptom onset.<sup>1,3,4</sup> This therapy appears less efficacious at improving hearing recovery if started beyond 4 weeks.<sup>1,3</sup> Rates of recovery with corticosteroids appear to be variable, with some patients experiencing improvement within days of initiating treatment and others requiring a longer duration for possible recovery.<sup>1</sup> Improvement with corticosteroids does not necessarily mean that head imaging can be omitted.<sup>3</sup> Treatment can be initiated while awaiting this further workup.

**Intratympanic steroids.** An alternative to oral corticosteroids is intratympanic (IT) steroids, which are given as methylprednisolone or dexamethasone formulations. Intratympanic steroids have been shown to be noninferior to oral steroids in several trials. Current treatment consensus still favours oral steroids over IT

steroids as first-line therapy, but this might change as further evidence emerges.<sup>12,15</sup> Presently, IT steroids are used for patients with relative contraindications to oral steroids, as well as in those patients who have not improved with oral steroids.<sup>12,15</sup>

### Prognosis and follow-up

Hearing recovery appears to be related to the degree of initial hearing loss.<sup>1</sup> Of interest, some studies have shown that even without treatment, 32% to 65% of patients with idiopathic SSNHL recover their hearing, typically within 2 weeks of onset, which suggests a relatively high spontaneous resolution rate in the natural history of this condition.<sup>4,16</sup> Factors that appear to negatively affect recovery of hearing include older age, presence of associated vertigo, and a flat pattern of hearing loss across frequencies (as opposed to only low and mid frequencies).<sup>4</sup> Follow-up with an otolaryngologist for all cases of SSNHL is recommended to ensure symptom resolution and appropriate diagnosis.

### Case resolution

After a more thorough history and physical examination, there are no obvious underlying causes for the sudden hearing loss. After a complete evaluation, you determine that the patient likely has idiopathic SSNHL. You prescribe the patient 50 mg of prednisone once daily for a total of 7 days and arrange urgent follow-up with your hospital's otolaryngologist. Magnetic resonance imaging is arranged on an outpatient basis within the next week through the otolaryngologist. Six months later, the patient reports almost complete resolution of the hearing loss. 🌿

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#### Competing interests

None declared

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## BOTTOM LINE

- Sudden sensorineural hearing loss (SSNHL) is considered an otolaryngologic emergency and has potentially challenging health consequences if not identified and treated promptly.
- Emergency department evaluation of a patient with SSNHL should include otoscopy, bedside hearing tests, and a careful neurologic examination.
- Current treatment recommendations for idiopathic SSNHL include an early and short course of oral steroids.
- Otolaryngology follow-up, including audiometry and possible brain magnetic resonance imaging, are usually arranged on an outpatient basis.

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