

# Early identification of idiopathic sensorineural

By Elizabeth Ann H. Jensen, DNP, FNP-BC, CNE;  
Elizabeth D. Harmon, DNP, APRN, FNP-BC; and  
Whitney Smith, MSN, APRN, ANP-BC, GNP-BC

**Abstract:** *This article discusses the physiology of normal hearing, the pathophysiology and differential diagnoses of sudden sensorineural hearing loss, and an approach for definitive diagnosis and treatment. A focus is placed on idiopathic sudden sensorineural hearing loss in the adult population in a primary care setting.*

**H**earing loss is a common adult patient complaint; however, for patients with sudden sensorineural hearing loss (SNHL), this symptom can cause fear and disability. Although there are many relatively benign causes of hearing loss, sudden SNHL is an important diagnosis to consider. If left untreated within a narrow window of opportunity, sudden SNHL can become permanent.<sup>1,2</sup> During the initial patient encounter, clinicians only discover the etiology of sudden SNHL in 10% to 15% of cases (although an etiology for sudden SNHL is eventually identified in up to 29% of cases).<sup>3,4</sup> Idiopathic sudden SNHL is the term used to describe the large majority of cases in which there is no attributable cause. This article is founded on the evidence-based guidelines from the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) and augmented with findings from a review of the literature.

**Keywords:** hearing loss, idiopathic sudden sensorineural hearing loss, primary care, psychosocial implications, sudden sensorineural hearing loss



# sudden hearing loss

## ■ Physiology and pathophysiology

The range of audible sound is approximately 20 hertz (Hz) to 20,000 Hz for the human ear.<sup>5</sup> In comparison, an upright bass can create a note at 40 Hz while a violin can play notes with frequencies well above 20,000 Hz.<sup>6</sup> The frequency of a sound wave determines the *pitch*; the lower the frequency, the lower the pitch. Another important component of hearing is the volume of the sound registered in decibels (dB). A whisper is approximately 30 dB; a thunderclap is 120 dB.<sup>7</sup> Normal conversations are between 500 to 3,000 Hz at 45 to 60 dB.<sup>8</sup> An audiogram measures both the pitch (Hz) and volume (dB) perception of an individual.

To understand hearing loss, it is important to understand the physiology of the ear. The pinna is responsible for channeling sound down the ear canal to the tympanic membrane. The middle ear comprises the area from the tympanic membrane to the inner wall of the cochlea. Within this space is the thin, air-filled Eustachian tube, which connects the nasopharynx to the inner ear. (See *Anatomy of the ear*.) It is lined with

a mucous membrane and has the ability to secrete mucus.<sup>9</sup> Also within the middle ear are three small bones: the malleus, incus, and stapes, which mechanically conduct sound vibrations from the outer ear to the inner ear by oscillation.<sup>10</sup>

The inner ear houses the cochlea, which is housed within the temporal bone. The cochlea is a membranous labyrinth bathed in a fluid called perilymph. This labyrinth is lined with hair cells that convert the vibration of sound waves into nerve impulses. These impulses are then transmitted to and from the brain via nerve fibers, resulting in the perception of hearing.<sup>10</sup> (See *Path taken by sound waves reaching the inner ear*.)

Knowledge of conductive hearing loss (CHL) compared with SNHL is essential to understand how to best approach the patient with a chief complaint of hearing loss. CHL occurs when there is a partial or complete impedance of sound to the middle ear or when there is not sufficient acoustic energy for the sound to transfer from the middle ear to the inner ear.<sup>11</sup>

SNHL is caused from either damage to the cochlea (which hinders the conversion of vibratory sound waves



into nerve impulses) or impedance of the neurologic electrical impulse from the cochlear apparatus to the brain.<sup>11</sup> CHL is a problem of physical impedance, whereas SNHL is a problem of neurologic impedance.

In the United States, the incidence of sudden SNHL is 5 to 20 cases per 100,000 individuals, with about 4,000 new cases diagnosed annually.<sup>4,12</sup> Sudden SNHL tends to occur in middle-aged adults and occurs equally in both genders. Probability of recovery appears to be inversely related to the degree of hearing loss.<sup>13</sup> Approximately 10% of all cases of

sudden SNHL have a definable etiology that is identified early in the evaluation process.<sup>4</sup>

Examples of known etiologies include acoustic neuroma, stroke, demyelinating disease, syphilis, Ménière disease, trauma, and perilymphatic fistula.<sup>4,8,12,14</sup> Roughly 90% of sudden SNHL cases have no obvious identifiable cause.<sup>4</sup> Proposed theories for the cause of idiopathic sudden SNHL include viral infection, vascular occlusion, immune mechanisms, and labyrinthine membrane ruptures.<sup>3,15</sup>

### History and physical exam

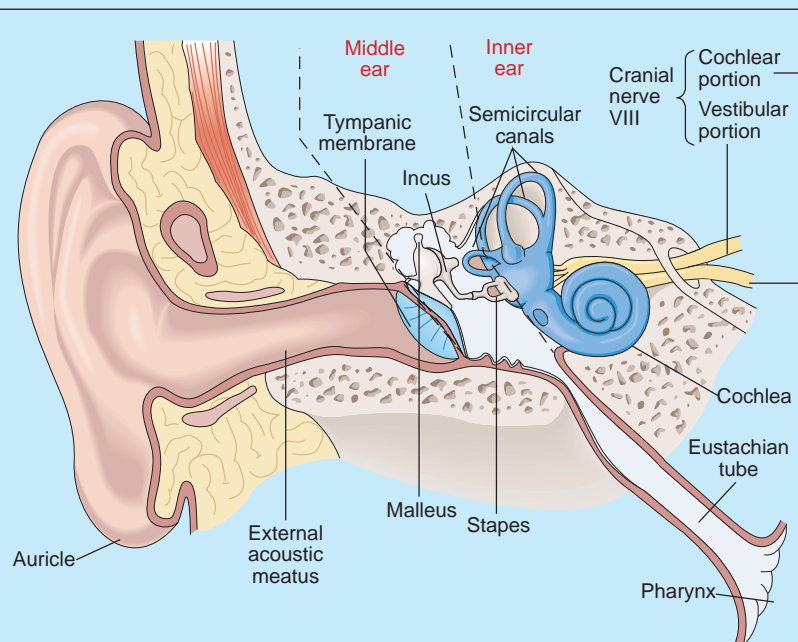
A delay in diagnosis of sudden SNHL is not uncommon. Clinicians must consider sudden SNHL as a differential diagnosis for conditions that are frequently observed in the clinic, including cerumen impaction, Eustachian tube dysfunction, trauma from foreign objects, otitis externa, otitis media, and serous otitis media. Understanding the presentation of a CHL versus a SNHL is paramount. This knowledge will lead to obtaining an appropriate history, performing a proper physical exam, interpreting the findings correctly, and instituting an effective treatment plan.

The history should contain questions to differentiate a CHL from an SNHL. Inquiry about the onset and duration of hearing loss is important because CHLs are usually associated with gradual, fluctuating hearing loss, whereas sudden SNHL occurs abruptly, is often noticed immediately after a patient awakens in the morning, and is typically more constant and rapidly progressing.<sup>4,16</sup>

Clinicians should ask whether the patient has experienced trauma, pain, otorrhea, previous hearing loss, vertigo, tinnitus, and systemic symptoms such as fever, chills, or myalgia to better distinguish CHL from sudden SNHL.<sup>4</sup> One of the most common symptoms of sudden SNHL is ear fullness, blocking of sound, and pressure instead of frank ear pain.<sup>4,17,18</sup> Tinnitus is almost always a clinical feature of sudden SNHL, and vertigo is present in 30% to 40% of cases.<sup>4</sup>

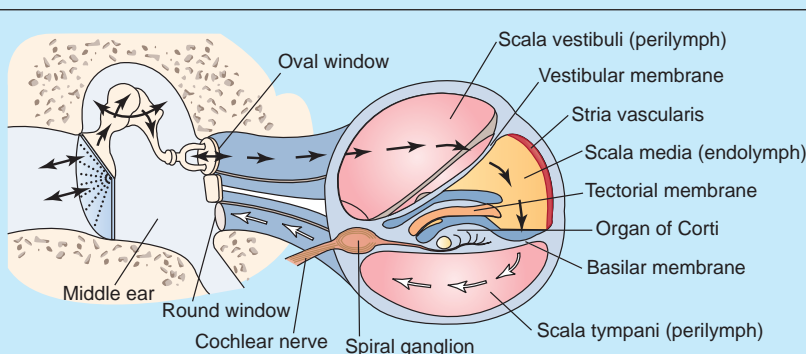
In addition to inquiring about a history of hearing loss, clinicians should ask about the location of hearing loss

### Anatomy of the ear



Source: Porth CM. *Essentials of Pathophysiology Concepts of Altered Health States*. 4th ed. Philadelphia, PA: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2015:975.

### Path taken by sound waves reaching the inner ear



Source: Porth CM. *Essentials of Pathophysiology Concepts of Altered Health States*. 4th ed. Philadelphia, PA: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2015:980.

(unilateral or bilateral) and the presence of other neurologic symptoms, such as sudden onset of bilateral hearing loss, diplopia, severe headaches, focal weakness, oscillopsia, gait disturbances, and balance disturbances.<sup>4</sup> Asking these questions can assist with early identification of an underlying disease process such as stroke, acoustic neuroma, or malignancy.<sup>4</sup> Genetic factors such as hereditary hearing loss, tobacco use, and medication histories can also help identify causes of hearing loss.<sup>18</sup> Medications frequently associated with sudden SNHL include loop diuretics, aminoglycosides, and cisplatin chemotherapy medications.<sup>18</sup>

The physical exam is instrumental to reach the correct diagnosis. Cerumen impaction, Eustachian tube dysfunction, trauma from foreign bodies, otitis externa, acute otitis media, and serous otitis media can be confirmed by examining the ear canal and tympanic membrane. If the patient has a cerumen impaction, it is important to remove the cerumen to adequately visualize the tympanic membrane for signs of additional abnormalities.<sup>4,17</sup>

The otoscopic exam is usually normal in a patient with sudden SNHL.<sup>19</sup> When sudden SNHL is suspected, a thorough neurologic exam must be conducted to identify possible serious conditions, such as stroke, malignant neoplasms, and acoustic neuroma.<sup>19</sup> The neurologic exam should include an assessment of the cranial nerves, cerebellum (rapid alternating and point-to-point movements), balance (Romberg and tandem gait), facial sensation (light touch and pinprick), nystagmus (positional, gaze, or spontaneous), and resistive muscle strengths.<sup>4,19</sup>

Another helpful resource during the physical exam is the tuning-fork test, also known as Weber and Rinne tests. Tuning-fork frequencies often used are 256 Hz, 512 Hz, and 1,024 Hz. Although tuning-fork tests are a useful first step in differentiating CHL from SNHL, patients may have hearing loss in a frequency other than the range of the tuning forks, and there may be up to a 20% false-negative response to the Weber test in patients with sudden SNHL.<sup>12,20</sup>

If the Weber test lateralizes to the unaffected ear and the Rinne test is normal, the clinician should be suspicious of sudden SNHL. A helpful video example of how to perform the Weber and Rinne tests can be found online ([www.youtube.com/watch?v=RVH4K4EcsiA](http://www.youtube.com/watch?v=RVH4K4EcsiA)).<sup>21</sup> (See *Interpreting Weber and Rinne tests*.)<sup>22</sup>

Part of the clinician's responsibility to patients with sudden SNHL is excluding retrocochlear pathology as the cause of the hearing loss. A common cause of retrocochlear pathology is an acoustic neuroma, also called a vestibular schwannoma. Contrast magnetic resonance imaging (MRI) is the modality of choice to eliminate the diagnosis of an acoustic neuroma due to its sensitivity in detecting retrocochlear pathology as well as other sudden SNHL-associated pathologies.<sup>4</sup> Computed tomography is a reasonable alternative if the pa-

### Interpreting Weber and Rinne tests

	Weber lateralizes	Rinne test
<b>Conductive loss</b>		
Good ear		AC > BC
Bad ear	To bad ear	BC > AC
<b>Sensorineural loss</b>		
Good ear	To good ear	AC > BC
Bad ear		AC > BC
AC > BC: Air conduction better than bone conduction (normal Rinne).		
BC > AC: Bone conduction better than air conduction (abnormal Rinne).		
Weber PC. Evaluation of hearing loss in adults. UpToDate. 2016. <a href="http://www.uptodate.com">www.uptodate.com</a> . Reproduced with permission from Weber PC.		

tient has a pacemaker, focal neurologic findings, claustrophobia, and financial constraints, or if an MRI is not accessible.<sup>4,18</sup>

### ■ Making the diagnosis

Idiopathic sudden SNHL can be presumed up to this point; however, more information is needed to make a definitive diagnosis. A diagnosis of idiopathic sudden SNHL can be made definitively when the hearing loss occurs over a 72-hour period, if no other cause of hearing loss is identified during the history and physical, and a hearing loss of 30 dB or greater has occurred in three consecutive frequencies with standard pure tone audiometry.<sup>4</sup> If there are no previous audiograms for comparison, the opposite ear may be used as a reference point (see *Sudden SNHL audiogram*).<sup>4,23</sup>

Routine lab testing is not advised unless there is a specific finding in the history and physical that warrants further investigation.<sup>4</sup> Cues that an underlying etiology of sudden SNHL exists include bilateral or fluctuating hearing loss; focal neurologic findings; nystagmus; and a history of trauma, vestibular symptoms, and ocular symptoms such as erythema, pain, and increased lacrimation.<sup>4,24</sup>

While some causes of sudden SNHL are rare, others are more common. Common causes of SNHL requiring further diagnostics include meningitis, syphilis, Lyme disease, Ménière disease, acoustic neuroma, malignancy, trauma, rheumatic fever, systemic lupus erythematosus, thyrotoxicosis, and multiple sclerosis.<sup>2-4,18,25</sup> Toxic exposures to medications or heavy metals such as mercury, lead, or arsenic can also cause bilateral hearing loss.<sup>4</sup> If a discoverable cause of sudden SNHL is suspected, lab testing should be tailored to the suspected disease state.

### ■ Treatment

Once sudden SNHL has been determined to be idiopathic in nature, the next step is to consider treatment options. Many

patients will experience a spontaneous recovery without treatment; however, idiopathic sudden SNHL is considered by some to be an otologic emergency that requires urgent intervention.<sup>4,17,26-28</sup> The advanced practice registered nurse (APRN) should initiate an urgent consultation to an otolaryngologist for future patient care. Improved outcomes occur when corticosteroids are initiated during the first 2 weeks of idiopathic sudden SNHL; there is little benefit if corticosteroids are initiated after 4 weeks of the onset of hearing loss.<sup>4</sup>

Evidence of the efficacy of treatment with either oral corticosteroids or intratympanic corticosteroid injections is mixed. For many patients, however, the benefit of possible hearing recovery outweighs the risks of treatment.<sup>4</sup> When making treatment decisions, it is crucial that the patient is regarded as an equal and active participant and that a discussion of benefits versus risks of treatment occurs. The efficacy, cost, and adverse reactions for various treatment options should also be considered and discussed because they may affect the final decision on the patient's treatment plan.

Furthermore, shared decision making may provide the patient, who may be anxious about additional hearing loss versus hearing recovery, with a sense of control and proactivity.<sup>4</sup> Although the literature is ambiguous in regards to the benefit of corticosteroids, early treatment of idiopathic

sudden SNHL has been associated with a more favorable prognosis.<sup>4,17-19,25,28</sup>

Patients with certain conditions will need to be treated cautiously. For example, for a patient with diabetes mellitus with idiopathic sudden SNHL, short-term adjustment of antidiabetic medications and more frequent self-monitoring of blood glucose levels should be considered. Other conditions for which the clinician would want to use caution include a prior psychiatric reaction to corticosteroid treatment, cataracts, glaucoma, thyroid disease, labile hypertension, heart failure, ulcerative gastrointestinal disease, tuberculosis, and osteoporosis.<sup>4,29</sup>

The importance of reviewing medication adverse reactions during the aforementioned shared decision-making discussion should not be underestimated. Common adverse reactions of systemic corticosteroids include hyperglycemia, vertigo, mood changes, weight gain, insomnia, increased gastric acid secretion, and increased sweating.<sup>4,26,29</sup> The AAO-HNS recommends prescribing prednisone for 10 to 14 days, then tapering it off over another 10 to 14 days.<sup>4</sup> Other corticosteroids considered for treatment include methylprednisolone and dexamethasone.

Another treatment includes intratympanic corticosteroid injections, which often requires multiple visits to the otolaryngologist and can result in increased cost to the patient. When used after initial treatment with systemic oral corticosteroids, intratympanic corticosteroid injection treatment is called *salvage therapy*. Intratympanic corticosteroid injections are generally safer and better tolerated than oral systemic corticosteroids.<sup>4,30</sup> Overall, there is no sufficient evidence to suggest the superiority of intratympanic corticosteroid injections over oral systemic corticosteroids.<sup>31</sup>

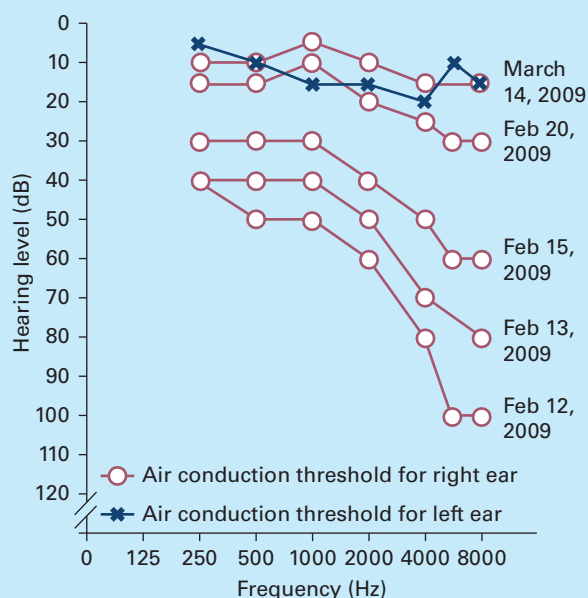
Although more evidence is needed, a growing body of literature supports a combination of intratympanic corticosteroid injections and systemic oral corticosteroid treatment.<sup>31</sup> Although not yet approved by the FDA, a promising treatment option for idiopathic sudden SNHL is hyperbaric oxygen therapy (HBOT). The best results are seen when HBOT is provided in the first 2 weeks; however, HBOT can be considered in the first 3 months after idiopathic sudden SNHL diagnosis.<sup>4</sup>

### Follow-up

Because the most improvement usually occurs during the first 2 weeks of treatment, patients should have a repeat audiogram and should be evaluated by an otolaryngologist to determine if salvage therapy is indicated.<sup>4,24</sup> If the patient has chosen a watchful waiting treatment approach, hearing outcomes should be assessed by follow-up audiometric evaluation within 6 months of the idiopathic sudden SNHL diagnosis.<sup>4</sup> Depending on the patient's hearing level, measured in dB and word recognition percentages, recovery is classified as either complete recovery, partial recovery, or no recovery.<sup>4</sup>

### Sudden SNHL audiogram

The figure depicts the air conduction of a patient diagnosed with sudden SNHL of the right ear. The patient's hearing improved significantly by the February 20, 2009 evaluation, with resolution by March 14, 2009.



Used with permission: Schreiber BE, Agrup C, Haskard DO, Luxon LM. Sudden sensorineural hearing loss. *Lancet*. 2010;375(9721):1203-1211.

If the patient continues to show hearing loss 6 months after initiation of treatment, the hearing loss is generally considered permanent, and the otolaryngologist would initiate a conversation with the patient about amplification and hearing assistive technology.<sup>4</sup> A generous estimate is that 66% of patients experience full recovery; at least 33% of patients experience some degree of permanent hearing loss.<sup>4,23,26</sup>

### ■ Psychosocial implications

There is a paucity of literature regarding the psychological ramifications of sudden SNHL. Reasons may include the low prevalence of the condition and the spontaneous recovery of up to 33% of patients.<sup>32</sup> An extensive literature search found that few scholarly articles with an emphasis on psychological effects of sudden SNHL have been published internationally; the majority have been published in Europe and Asia. Nevertheless, consideration of the psychosocial effects of sudden SNHL is an important part of treatment. The AAO-HNS practice guideline recommends addressing the psychological impact of sudden SNHL when providing patient care and education about sudden SNHL.<sup>4</sup>

Sudden SNHL usually occurs in middle-aged adults ages 43 to 53.<sup>17</sup> This is when adults are often at the peak of their careers, caring for their children or their parents, are leaders in their communities, and are generally otherwise healthy. The suddenness of the disruption in hearing, which affects quality of life and functional ability (often without clear etiology), can be physically and emotionally unsettling.

Patients with idiopathic sudden SNHL display a similar degree of emotional and physical distress compared with patients with bilateral hearing loss, and interpersonal relationships and social functioning become especially impaired.<sup>33</sup> Individuals with idiopathic sudden SNHL often feel more impaired than those with chronic hearing impairment.<sup>34</sup>

In addition to the hearing loss, patients may experience vertigo, tinnitus, psychosocial disturbances, increased levels of depression, and anxiety that another occurrence of hearing loss will occur.<sup>32,33,35</sup> Patients with tinnitus and vertigo after 1 year from the onset of sudden SNHL experience a lower quality of life and take more sick leave from work.<sup>30</sup> The financial costs of treatment, hearing restoration, battery replacement for hearing devices, and specialist consultations present more concern for the patient, contributing to the psychosocial burden. As APRNs, providing holistic care is the backbone of the nursing model. Anticipating patients' physical as well as psychosocial needs is a mainstay of treatment.

### ■ Patient education

Education regarding the necessity for immediate referral to an otolaryngologist, possible pharmacologic treatment, and the importance of follow-up should be provided by the

APRN. Topics such as audiologic rehabilitation, the need for surgical or nonsurgical hearing amplification devices, prognosis, and frequency and duration of continued follow-up should be initiated by a specialist who is well versed in the nuances of these issues.

Psychological aspects of the diagnosis should be addressed by both the otolaryngologist and the APRN. Patient education regarding hearing evolution is especially important because the quality of life of those with sudden SNHL is negatively affected, including patients who experience hearing recovery.<sup>36</sup> Although patients can be expected to be referred to an otolaryngologist, APRNs are an integral part of the treatment team. The APRN often has a long-term relationship with the patient, can detect subtle changes in psychological status, and can assist in coordinating care with specialists.

### ■ Hearing recovery

Hearing loss is a common reason for patients to seek medical care; however, a seemingly benign symptom can have serious consequences if not properly diagnosed. It is incumbent on the APRN to be able to differentiate sudden SNHL from CHL, initiate further diagnostics, and immediately seek consultation with an otolaryngologist. Treatment must not be delayed while waiting for diagnostic testing.

Shared decision making is vital prior to prescribing oral corticosteroids. The consequences of delayed treatment for a patient with idiopathic sudden SNHL can have a profound impact on the patient's quality of life; the physical and psychosocial burdens of idiopathic sudden SNHL are high. APRNs play a key role in early intervention and improved hearing recovery. **NP**

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Elizabeth Ann H. Jensen is a nursing instructor at the Medical University of South Carolina, Charleston, S.C.

Elizabeth D. Harmon is an instructor at the Medical University of South Carolina, Charleston, S.C.

Whitney Smith is an instructor at the Medical University of South Carolina, Charleston, S.C.

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## INSTRUCTIONS

### Early identification of idiopathic sudden sensorineural hearing loss

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