AAO-HNS Releases Guideline on Sudden Hearing Loss

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This guideline from the American Academy of Otolaryngology–Head and Neck Surgery (AAO-HNS) focuses on sudden sensorineural hearing loss (SNHL), which affects 5 to 20 per 100,000 persons in the United States. Early diagnosis is essential for effective management.

Recommendations

SNHL should be differentiated from conductive hearing loss in patients with sudden hearing loss.

SNHL and conductive hearing loss are common causes of hearing loss, and distinguishing between them is important for determining treatment options. Diagnosis is based on history, physical examination, tuning fork tests, and audiometry.

Persons with presumptive sudden SNHL should be evaluated for bilateral and recurrent sudden hearing loss, and focal neurologic findings.

Presumptive sudden SNHL can be the result of an underlying disease, including Meniere disease, and systemic, autoimmune, metabolic, and neurologic disorders. Physicians should evaluate for these conditions using history, physical and neurologic examination, and, if available, audiometry. Items to address include prior hearing loss, vertigo, and focal neurologic symptoms. When available, results of previous audiometric and neurologic evaluations should be reviewed. *Table 1* lists conditions that may be associated with bilateral sudden hearing loss.

Computed tomography (CT) of the head/brain should not be performed during initial evaluation of patients with presumptive sudden SNHL.

Although CT does not provide useful information to improve initial management options in patients with presumptive sudden SNHL, this recommendation does not apply to persons who present with focal neurologic findings, history of trauma, or chronic ear disease. It should also be noted that although imaging is not useful in the initial assessment, magnetic resonance imaging (MRI) of the brain or CT of the temporal bone could be beneficial in these patients at a later time.

Diagnosis of presumptive idiopathic sudden SNHL can be confirmed when the patient demonstrates hearing loss of at least 30 dB at three consecutive frequencies on audiometry, assuming an underlying condition is not identified by history and physical examination.

Audiometry differentiates conductive hearing loss from SNHL and establishes frequency-specific hearing thresholds, thereby making it necessary for definitive diagnosis of idiopathic sudden SNHL. Criteria used to diagnose sudden SNHL varies, but hearing loss of at least 30 dB at three consecutive frequencies is used most often in randomized controlled trials. It is also used by the National Institute on Deafness and Other Communication Disorders.

Laboratory testing should not be routinely performed in patients with idiopathic sudden SNHL.

This recommendation refers to automatic, shotgun, or universal testing that is not based on individual patient factors, and testing that is associated with cost and possible harms, including false-positive and false-negative results. Certain tests may still be warranted based on patient history (e.g., Lyme titers in patients in endemic regions).

MRI, auditory brainstem response, or follow-up audiometry should be performed in patients with idiopathic sudden SNHL to evaluate for retrocochlear pathology.

A small but significant number of patients with idiopathic sudden SNHL have an underlying lesion (e.g., schwannoma), indicating possible retrocochlear pathology. Gadolinium-enhanced MRI of the brain, brainstem, and internal auditory canals has the highest sensitivity for detecting retrocochlear pathology; however, persistent abnormalities on auditory brainstem

response or audiometry also suggest retrocochlear pathology, and MRI typically should then be performed. The decision to perform MRI in patients with normal results on auditory brainstem response or stable findings on audiometry should be based on patient preference after discussion with the physician; however, screening for vestibular schwannoma allows for possible earlier tumor identification, which gives the physician the most treatment options and the patient the best likelihood to preserve hearing. ▶

Table 1. Selected Conditions That May Be Associated with Bilateral Sudden Hearing Loss

Cause	*Other features*
Autoimmune inner ear disease	Fluctuation of hearing may sometimes occur; vertigo may occur in some cases
Bilateral synchronous internal auditory artery occlusion associated with vertebrobasilar vascular disease	Vertigo, dysarthria, facial weakness, ataxia, nystagmus, unilateral numbness, abnormal computed tomography or magnetic resonance angiography of the vertebrobasilar vasculature
Cogan syndrome	Nonsyphilitic interstitial keratitis of the cornea, hearing loss, vertigo
Genetic disorders	May be syndromic or nonsyndromic
Herpes zoster oticus (Ramsay-Hunt syndrome)	Otalgia, pinna and/or ear canal vesicles, facial nerve paresis, positive viral titers, positive viral cultures
HIV otitis	Positive HIV titers, altered T-cell counts, and often other cranial neuropathies may be associated with mastoiditis out of proportion to clinical problems
Hyperviscosity syndrome	Mucous membrane bleeding, neurologic and pulmonary symptoms, associated retinopathy
Lead poisoning	Learning disabilities, other stigmata of lead poisoning
Lyme disease	Erythema chronicum migrans, abnormal cerebrospinal fluid studies, fluctuating bilateral audiovestibular symptoms
Meningitis (infectious, inflammatory, neoplastic)	Headache, fever, abnormal cerebrospinal fluid studies, possibly other cranial nerve palsies
Mitochondrial disorders	
MELAS (mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes) syndrome	Periods of confusion, elevated serum lactic acid levels around times of attacks, stroke-like spells, magnetic resonance imaging white matter signal changes, migraine-like headaches, seizures, diabetes mellitus, mitochondrial gene mutation *(MT-RNR1, MT-TS1, POLG genes)*
Other mitochondrial disorders	Variable phenotypes
Neoplastic (neurofibromatosis II, bilateral vestibular schwannomas, intravascular lymphomatosis, others)	Abnormal brain magnetic resonance imaging or cerebrovascular imaging
Ototoxic medications	Vestibular loss, oscillopsia
Sarcoidosis	Pulmonary symptoms, bilateral vestibular loss, elevated serum angiotensin-converting enzyme level or abnormal gallium scan
Syphilis	Abnormal fluorescent treponemal antibody absorption test, bilateral fluctuating hearing loss, tabes dorsalis, multiorgan involvement
Trauma	Significant head trauma, barotrauma, temporal bone fractures

HIV = human immunodeficiency virus.

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Patients with idiopathic sudden SNHL should be provided education on the history of the condition, and the benefits, risks, and limited evidence of effectiveness of treatment.

Providing information about their condition and treatment options can help patients participate fully in shared decision making, which is known to improve compliance and outcomes.

Important points for patient education include:

- The cause of hearing loss is not always evident.
- Sudden SNHL rarely affects both ears.
- Other symptoms can be associated with the hearing loss, including tinnitus and vertigo.
- Some patients may recover some of their hearing within two weeks; those who recover at least one-half have a better prognosis, and those with minimal improvement are not likely to recover.
- Although evidence-based data are lacking, it is thought that early intervention helps with recovery; therefore, early recognition of hearing loss is essential.
- There are many treatment options, but evidence of their effectiveness is limited.
- Treatment benefits include the possibility of quicker and more complete recovery of hearing; however, there also are adverse effects of treatment, and these should be balanced with the benefits when choosing a treatment.
- Watchful waiting is an option, because some patients may recover spontaneously.
- Counseling may be beneficial.
- When hearing loss is diagnosed, audiologic rehabilitation (e.g., counseling, discussion of hearing amplification and restoration options) should be addressed.
- Appropriate follow-up and testing may be ensured by addressing the patient's concerns about cost.

Patients with idiopathic sudden SNHL can be offered corticosteroids as an option for initial therapy.

There have been many studies on the use of corticosteroids to treat idiopathic sudden SNHL; however, most do not meet current criteria for highest quality evidence. There is evidence of an inflammatory cell death cascade in patients with idiopathic sudden SNHL; this can be modified with corticosteroids. Corticosteroids have sites of action in the inner ear, with effectiveness in treating a variety of causes of hearing loss.

Patients with idiopathic sudden SNHL can be offered hyperbaric oxygen therapy within three months of diagnosis.

Hyperbaric oxygen therapy is not approved by the U.S. Food and Drug Administration for treatment of idiopathic sudden SNHL; however, randomized controlled trials and a Cochrane review have been performed. Hyperbaric oxygen therapy is thought to affect immunity, oxygen transport, and hemodynamics, which reduces hypoxia and edema and augments normal physiologic responses to infection and ischemia. It also enables delivery of increased partial pressure of oxygen to the cochlea, which is sensitive to ischemia. Potential adverse effects include damage from pressure changes to the ears, sinuses, and lungs; temporary vision changes; claustrophobia; and oxygen poisoning.

Antivirals, thrombolytics, vasodilators, vasoactive substances, and antioxidants should not be routinely prescribed in patients with idiopathic sudden SNHL.

Despite support for these agents, they have potential adverse effects and no evidence of effectiveness.

Intratympanic steroid therapy should be offered to patients with idiopathic sudden SNHL who have incomplete recovery after initial management.

Intratympanic steroid therapy has been suggested for hearing recovery in patients who do not recover spontaneously or after initial treatment. Studies of this approach exist, and despite having design flaws, most show improved outcomes with intratympanic steroid therapy. The objective of intratympanic steroid therapy is to reduce inflammation in the inner ear that may be causing the hearing loss or inhibiting recovery.

Follow-up audiometry should be performed in patients with idiopathic sudden SNHL within six months of diagnosis.

Follow-up is important to determine if there are other causes of the hearing loss and to determine if rehabilitation would be beneficial. Earlier follow-up audiometry may be indicated in patients in whom treatment has been started to determine the effectiveness of treatment, and in those with incomplete recovery to help guide decisions about initiating salvage therapy.

Patients with incomplete hearing recovery should be counseled about the possible benefits of amplification, hearing assistive technology, and other supportive measures.

Counseling on options for managing hearing loss is helpful. Counseling is also an important component throughout the evaluation and treatment period, and it should be performed by a specialist.

LISA HAUK, Senior Associate Editor, *AFP* Online ■

Answers to This Issue's CME Quiz

Q1. A, C, D	**Q5.** B, C, D	**Q9.** A, D
Q2. A, B, C, D	**Q6.** C	**Q10.** B
Q3. B	**Q7.** A, B, C	
Q4. C	**Q8.** A, B, D	