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# Clinical Practice Guideline: Sudden Hearing Loss

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## Abstract

**Objective.** Sudden hearing loss (SHL) is a frightening symptom that often prompts an urgent or emergent visit to a physician. This guideline provides evidence-based recommendations for the diagnosis, management, and follow-up of patients who present with SHL. The guideline primarily focuses on sudden sensorineural hearing loss (SSNHL) in adult patients (aged 18 and older). Prompt recognition and management of SSNHL may improve hearing recovery and patient quality of life (QOL). Sudden sensorineural hearing loss affects 5 to 20 per 100,000 population, with about 4000 new cases per year in the United States. This guideline is intended for all clinicians who diagnose or manage adult patients who present with SHL.

**Purpose.** The purpose of this guideline is to provide clinicians with evidence-based recommendations in evaluating patients with SHL, with particular emphasis on managing SSNHL. The panel recognized that patients enter the health care system with SHL as a nonspecific, primary complaint. Therefore, the initial recommendations of the guideline deal with efficiently distinguishing SSNHL from other causes of SHL at the time of presentation. By focusing on opportunities for quality improvement, the guideline should improve diagnostic accuracy, facilitate prompt intervention, decrease variations in management, reduce unnecessary tests and imaging procedures, and improve hearing and rehabilitative outcomes for affected patients.

**Results.** The panel made *strong recommendations* that clinicians should (1) distinguish sensorineural hearing loss from conductive hearing loss in a patient presenting with SHL; (2) educate patients with idiopathic sudden sensorineural hearing loss (ISSNHL) about the natural history of the condition, the benefits and risks

of medical interventions, and the limitations of existing evidence regarding efficacy; and (3) counsel patients with incomplete recovery of hearing about the possible benefits of amplification and hearing-assistive technology and other supportive measures. The panel made *recommendations* that clinicians should (1) assess patients with presumptive SSNHL for bilateral SHL, recurrent episodes of SHL, or focal neurologic findings; (2) diagnose presumptive ISSNHL if audiometry confirms a 30-dB hearing loss at 3 consecutive frequencies *and* an underlying condition cannot be identified by history and physical examination; (3) evaluate patients with ISSNHL for retrocochlear pathology by obtaining magnetic resonance imaging, auditory brainstem response, or audiometric follow-up; (4) offer intratympanic steroid perfusion when patients have incomplete recovery from ISSNHL after failure of initial management; and (5) obtain follow-up audiometric evaluation within 6 months of diagnosis for patients with ISSNHL. The panel offered as *options* that clinicians may offer (1) corticosteroids as initial therapy to patients with ISSNHL and (2) hyperbaric oxygen therapy within 3 months of diagnosis of ISSNHL. The panel made a *recommendation against* clinicians routinely prescribing antivirals, thrombolytics, vasodilators, vasoactive substances, or antioxidants to patients with ISSNHL. The panel made *strong recommendations against* clinicians (1) ordering computerized tomography of the head/brain in the initial evaluation of a patient with presumptive SSNHL and (2) obtaining routine laboratory tests in patients with ISSNHL.

## Keywords

evidence-based medicine, practice guidelines, sudden hearing loss, sudden sensorineural hearing loss, intratympanic steroids, hyperbaric oxygen

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**S**udden hearing loss (SHL) is a frightening symptom that often prompts an urgent or emergent visit to a physician. This guideline focuses on sudden sensorineural hearing loss (SSNHL), one of many causes of SHL, which, if recognized and managed promptly, may improve hearing recovery and patient quality of life (QOL). Sudden sensorineural hearing loss affects 5 to 20 per 100,000 population, with about 4000 new cases per year in the United States.<sup>1,2</sup> Throughout this guideline, the following definitions are used:

- Sudden hearing loss is defined as a rapid onset, occurring over a 72-hour period, of a subjective sensation of hearing impairment in one or both ears.
- Sudden sensorineural hearing loss (SNHL) is a subset of SHL that (a) is sensorineural in nature and (b) meets certain audiometric criteria.
  - (a) Sensorineural hearing loss indicates an abnormality of the cochlea, auditory nerve, or higher aspects of central auditory perception or processing.
  - (b) The most frequently used audiometric criterion is a decrease in hearing of  $\geq 30$  decibels (dB), affecting at least 3 consecutive frequencies. Because premorbid audiometry is generally unavailable, hearing loss is defined as related to the opposite ear's thresholds.
- Idiopathic sudden sensorineural hearing loss (ISSNHL) is defined as SSNHL with no identifiable cause despite adequate investigation.

The SSNHL definition used throughout this guideline is based on its consistent use in the literature and National Institute on Deafness and Other Communication Disorders (NIDCD) criteria<sup>3</sup>; however, the panel recognizes that in clinical practice, expanding the definition to cases with less

than 30 dB of hearing loss may be considered. The panel recognizes that the NIDCD definition is not universally used, and accordingly, published evidence not using this definition was considered.

The distinction between SSNHL and other causes of SHL is one that should be made by the initial treating health care provider, so that early diagnosis and management can be instituted. Moreover, nonidiopathic causes of SSNHL must be identified and addressed during the course of management; the most pressing of these are vestibular schwannoma (acoustic neuroma), stroke, and malignancy.<sup>4</sup> Up to 90% of SSNHL, however, is idiopathic at presentation and is presumptively attributed to vascular, viral, or multiple etiologies.<sup>5</sup>

A maximum of 32% to 65% of cases of SSNHL may recover spontaneously.<sup>2,6</sup> Clinical experience indicates that even this recovery rate may be an overestimation. Prognosis for recovery is dependent on a number of factors, including patient age, presence of vertigo at onset, degree of hearing loss, audiometric configuration, and time between onset of hearing loss and treatment.<sup>7-9</sup> Treatment options are myriad and include systemic and topical steroids, antiviral agents, rheologic agents, diuretics, hyperbaric oxygen treatment, other medications, middle ear surgery for fistula repair, and observation alone. The comparative efficacy of these treatments, however, is not known, considering that the definitive etiology is also commonly not known.

Long-term follow-up is recommended as some patients will have an underlying cause identified that may not be evident at initial presentation.<sup>10</sup> In addition, the patient with partial or no hearing recovery, or persistent tinnitus, will require ongoing management from otolaryngological, audiological, and psychological perspectives.<sup>11</sup>

This guideline is intended for all clinicians who diagnose or manage adult patients (age 18 years and older) who present with SHL. After addressing causes, diagnosis, and treatments of non-SSNHL briefly, this guideline will address SSNHL in detail. Important points to keep in mind include the following:

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- A cause for SSNHL is identified in only 10% to 15% of patients at the time of presentation.<sup>7,9</sup> Emergency intervention may be needed for rare, life-threatening conditions of which SSNHL is a part. In up to a third of cases, the cause may be identified only after long-term follow-up evaluations.<sup>10</sup>
- In 85% to 90% of cases, despite thorough evaluation, the underlying cause is unknown or uncertain at the time of presentation, and treatment decisions are generally made without knowledge of the etiology.<sup>7,9</sup> It is appropriate, therefore, to approach these idiopathic cases in a common way, understanding that the underlying etiologies may be very dissimilar.<sup>12</sup>
- The primary presenting symptom of SHL is a full or blocked ear. Because this is such a common and nonspecific symptom, both patients and physicians are not sufficiently frightened or worried by it. Thus, evaluation and treatment are often delayed. New onset of ear blockage or fullness can be a symptom of potentially serious conditions and warrants prompt evaluation.
- Conversely, the patient with SHL may be very frightened; the nearly universal accompanying tinnitus seen in SSNHL will frequently contribute intensely to his or her anxiety and depression.<sup>13</sup> All members of the hearing health care team should be cognizant of the psychological response to the sudden loss of a primary sense.
- Familiarity with hearing aids, hearing-assistive technology (HAT), tinnitus management, and implantable hearing solutions is required in the ongoing management of these patients.
- A “team approach” to the overall management of these patients is encouraged.

The incidence of this symptom, the debilitating consequences of missed early diagnosis and management, the presentation of the patient to a variety of health care providers, the abundance of small series and case reports regarding treatment, and the paucity of randomized controlled trials (RCTs) assessing interventions create a pressing need for evidence-based guidelines to aid clinicians in managing SSNHL. Moreover, wide variations in evaluation, treatment, counseling, and follow-up of patients with SSNHL exist worldwide. Such variations are usually ascribed to heterogeneity in clinical practice and training rather than to differences in clinical need. The current lack of consensus, both in the United States and worldwide, on all aspects of care of the patient with SSNHL, further supports the need for an evidence-based clinical practice guideline to highlight best practices.

### Guideline Purpose

The purpose of this guideline is to provide clinicians with evidence-based recommendations in evaluating patients with SHL, with particular emphasis on managing SSNHL. The guideline is intended for all clinicians who see adult patients aged 18 years and older. The recommendations outlined in this guideline are not intended to represent the standard of care for patient management, nor are the recommendations

intended to limit treatment or care provided to individual patients. The guideline is not intended to replace individualized patient care or clinical judgment.

Although the guideline focuses primarily on managing SSNHL, the panel recognized that patients enter the health care system with SHL as a nonspecific, primary complaint. Therefore, the initial recommendations of the guideline deal with efficiently distinguishing SSNHL from other causes of SHL at the time of presentation. The purpose of the guideline is not to present an exhaustive approach to managing SHL, in general, as only a limited number of causes are discussed.

This is the first clinical guideline on SSNHL developed in the United States. Use of this guideline may improve the care of patients and result in improved outcomes. Despite numerous published articles on SSNHL, there remains a paucity of high-quality evidence, creating confusion and practice variations in management. This guideline will provide evidence-based recommendations for clinicians based on multidisciplinary consensus and careful consideration of the benefits vs harms of suggested actions. By focusing on opportunities for quality improvement, the guideline should improve diagnostic accuracy, facilitate prompt intervention, decrease inappropriate variations in management, reduce unnecessary tests and imaging procedures, and improve hearing and rehabilitative outcomes for affected patients.

### Health Care Burden

The incidence of SSNHL is reported as 5 to 20 per 100,000 population, with some estimates ranging as high as 160 per 100,000.<sup>14,15</sup> In most cases, there are multiple physician visits, including emergency physicians, primary care physicians, and otolaryngologists, and still the etiology is not apparent, which can lead to extensive testing. The appropriateness of tests will be examined in this publication but often includes magnetic resonance imaging (MRI), audiometric evaluation (initial and follow-up), and laboratory testing, such as hematologic, serologic, and autoimmune testing.

Because the etiology is usually unknown, treatments have been empiric. The most commonly used treatment has been corticosteroids (systemic and/or intratympanic). A large array of other treatments such as antivirals, antibiotics, diuretics, vasodilators, osmotic agents, plasma expanders, anticoagulants, mineral supplements, and hyperbaric oxygen or carbon dioxide-rich gases, among others, have been used.<sup>16</sup> The lack of one or more uniformly accepted treatment(s) potentially increases the cost of management. Coexistent morbidities such as dizziness and tinnitus are not the subject of this guideline but pose considerable disease burdens for the patient. Dizziness is present in 30% to 40% of cases of SSNHL.<sup>8,17</sup> The associated evaluation and treatment include audiometric testing and follow-up and may include vestibular testing, consultations with neurology and other specialties, and intensive courses of vestibular rehabilitation. Tinnitus is expected to be nearly universal in SSNHL, is difficult to treat, and may pose a significant economic and psychological burden.<sup>18</sup>

The overall audiological burden of SSNHL is considerable. Patients with sudden unilateral hearing loss have immediate difficulty with conversation on the involved side and hearing in

noisy environments, and if they have preexisting hearing loss in the opposite ear from common sources such as presbycusis and noise exposure, SSNHL will only compound the problem. In patients with SSNHL, the asymmetry may often result in the inability to determine where a sound originates, which can be frustrating and even disorienting to the listener. The inability to localize sound may also be very dangerous and put patients at risk for accidents. As a result of these difficulties, rehabilitation of patients with SSNHL can involve hearing aids, surgically implantable hearing devices, or both, with significant resultant expense to the patient and to the health care system.

The significant impact of unilateral sensorineural hearing loss on patients' QOL has been studied in both adults and children.<sup>19,20</sup> The same burden is present in SSNHL, possibly even more so, especially if dizziness and significant tinnitus are suddenly present.<sup>18,21,22</sup> Patients are frustrated that their hearing loss is "not visible" to friends and family and that their physician may not know what caused the problem even after expensive testing. The addition of dizziness and tinnitus associated with SSNHL adds to the diminished QOL.

## Methods

This guideline was developed using an explicit and transparent a priori protocol for creating actionable statements based on supporting evidence and the associated balance of benefit and harm.<sup>23</sup> The guideline development panel comprised representatives from the fields of otolaryngology, otology, neurology, neurotology, family medicine, emergency medicine, audiology, and consumer groups.

All literature searches were performed by an information specialist at the Cochrane ENT Disorders Group through November 27, 2010. Three initial searches were performed to identify clinical practice guidelines, systematic reviews, and RCTs. In addition, a fourth search identified literature relating to the diagnosis of SHL. The searches were performed in multiple databases, including the National Guidelines Clearinghouse (NGC; [www.guideline.gov](http://www.guideline.gov)), The Cochrane Library, the Cumulative Index to Nursing and Allied Health Literature (CINAHL), EMBASE, PubMed, Web of Science, BIOSIS, the Cochrane Central Register of Controlled Trials (CENTRAL), CAB Abstracts, CMA Infobase, NHS Evidence ENT and Audiology, National Library of Guidelines, National Institute of Clinical Excellence (NICE), Scottish Intercollegiate Guidelines Network (SIGN), New Zealand Guidelines Group (NZGG), Australian National Health and Medical Research Council, Tripdatabase, The Database of Abstracts of Reviews of Effects (DARE), HTA Database, and HSTAT.

1. Clinical practice guidelines were identified by a PubMed, EMBASE, CINAHL, Web of Science, CAB Abstracts, BIOSIS, Cochrane Library, DARE, HTA Database, and HSTAT search using *guideline* as a publication type or title word. The search identified 13 guidelines with a topic of SHL. After removing duplicates, clearly irrelevant references, and non-English-language articles, 1 guideline was selected for the panel's attention.

2. Systematic reviews were identified using a validated filter strategy that initially yielded 151 potential articles. The final data set included 29 systematic reviews or meta-analyses on SHL that were distributed to the panel members. Articles were excluded if they were not available in English and did not meet the panel's quality criteria (ie, the review had a clear objective and method, an explicit search strategy, and a valid method of data extraction).
3. Randomized controlled trials were identified through PubMed, EMBASE, Web of Science, BIOSIS, CINAHL, and CENTRAL and totaled 339 trials. The results were then filtered by the guidelines chair and assistant chairs, removing articles that were not relevant to the work of the group. As a result, 136 articles were made available to the guideline panel.
4. Research articles related to the diagnosis of SHL were identified via PubMed. The search was conducted with the following Medical Subject Headings (MESH): "Hearing Loss, Sudden/etiology" and "Hearing Loss, Sudden/diagnosis" and identified 958 possible articles. Articles were removed that were non-English, did not report an abstract, and were tagged with a publication type of "case report." The results were then reviewed by the guidelines' chair and assistant chairs, who removed nonrelevant articles. As a result, 133 articles were made available to the guideline panel.

Results of all literature searches were distributed to guideline panel members, including electronic listings with abstracts (if available) of the searches for clinical guidelines, RCTs, systematic reviews, and other studies. This material was supplemented, as needed, with targeted searches to address specific needs identified in writing the guideline through June 2011.

In a series of conference calls, the working group defined the scope and objectives of the proposed guideline. During the 12 months devoted to guideline development ending in July 2011, the group met twice, with in-person meetings following the format previously described,<sup>23</sup> using electronic decision support (BRIDGE-Wiz) software to facilitate creating actionable recommendations and action statement profiles. Internal electronic review and feedback on each guideline draft were used to ensure accuracy of content and consistency with standardized criteria for reporting clinical practice guidelines.<sup>24</sup>

American Academy of Otolaryngology–Head and Neck Surgery Foundation (AAO-HNSF) staff used the Guideline Implementability Appraisal and Extractor (GLIA)<sup>25</sup> to appraise adherence of the draft guideline to methodological standards, to improve clarity of recommendations, and to predict potential obstacles to implementation. Guideline panel members received summary appraisals in May 2011 and modified an advanced draft of the guideline.

The final guideline draft underwent extensive external peer review. Comments were compiled and reviewed by the panel's chair, and a modified version of the guideline was

**Table 1.** Guideline Definitions for Evidence-Based Statements

Statement	Definition	Implication
Strong recommendation	A strong recommendation means the benefits of the recommended approach clearly exceed the harms (or that the harms clearly exceed the benefits in the case of a strong negative recommendation) and that the quality of the supporting evidence is excellent (grade A or B). <sup>a</sup> In some clearly identified circumstances, strong recommendations may be made based on lesser evidence when high-quality evidence is impossible to obtain and the anticipated benefits strongly outweigh the harms.	Clinicians should follow a strong recommendation unless a clear and compelling rationale for an alternative approach is present.
Recommendation	A recommendation means the benefits exceed the harms (or that the harms exceed the benefits in the case of a negative recommendation), but the quality of evidence is not as strong (grade B or C). <sup>a</sup> In some clearly identified circumstances, recommendations may be made based on lesser evidence when high-quality evidence is impossible to obtain and the anticipated benefits outweigh the harms.	Clinicians should also generally follow a recommendation but should remain alert to new information and sensitive to patient preferences.
Option	An option means that either the quality of evidence that exists is suspect (grade D) <sup>a</sup> or that well-done studies (grade A, B, or C) <sup>a</sup> show little clear advantage to one approach vs another.	Clinicians should be flexible in their decision making regarding appropriate practice, although they may set bounds on alternatives; patient preference should have a substantial influencing role.
No recommendation	No recommendation means there is both a lack of pertinent evidence (grade D) <sup>a</sup> and an unclear balance between benefits and harms.	Clinicians should feel little constraint in their decision making and be alert to new published evidence that clarifies the balance of benefit vs harm; patient preference should have a substantial influencing role.

<sup>a</sup>See **Table 2** for definition of evidence grades.

distributed and approved by the guideline development panel. The recommendations contained in the guideline are based on the best available data published through June 2011. Where data were lacking, a combination of clinical experience and expert consensus was used. A scheduled review process will occur at 5 years from publication or sooner if new compelling evidence warrants earlier consideration.

### Classification of Evidence-Based Statements

Guidelines are intended to produce optimal health outcomes for patients, to minimize harms, and to reduce inappropriate variations in clinical care. The evidence-based approach to guideline development requires that the evidence supporting a policy be identified, appraised, and summarized and that an explicit link between evidence and statements be defined. Evidence-based statements reflect both the quality of evidence and the balance of benefit and harm that is anticipated when the statement is followed. The definitions for evidence-based statements are listed in **Tables 1** and **2**.<sup>26</sup> As much of the guideline dealt with evidence relating to diagnostic tests, **Table 2** was adapted to include current recommendations from the Oxford Centre for Evidence-Based Medicine.<sup>27</sup>

Guidelines are not intended to supersede professional judgment; rather, they may be viewed as a relative constraint on individual clinician discretion in a particular clinical circumstance. Less frequent variation in practice is expected for a “strong recommendation” than might be expected with a “recommendation.” “Options” offer the most opportunity for

practice variability.<sup>28</sup> Clinicians should always act and decide in a way that they believe will best serve their patients’ interests and needs, regardless of guideline recommendations. They must also operate within their scope of practice and according to their training. Guidelines represent the best judgment of a team of experienced clinicians and methodologists addressing the scientific evidence for a particular topic.<sup>26</sup>

Making recommendations about health practices involves value judgments on the desirability of various outcomes associated with management options. Values applied by the guideline panel sought to minimize harm and diminish unnecessary and inappropriate therapy. A major goal of the panel was to be transparent and explicit about how values were applied and to document the process.

### Financial Disclosure and Conflicts of Interest

The cost of developing this guideline, including travel expenses of all panel members, was covered in full by the AAO-HNSF. Potential conflicts of interest for all panel members in the past 5 years were compiled and distributed before the first conference call. After review and discussion of these disclosures,<sup>29</sup> the panel concluded that individuals with potential conflicts could remain on the panel if they (1) reminded the panel of potential conflicts before any related discussion, (2) recused themselves from a related discussion if asked by the panel, and (3) agreed not to discuss any aspect of the guideline with industry before publication. Last, panelists were reminded that conflicts of interest extend beyond financial relationships and may include personal experiences, how

**Table 2.** Evidence Quality for Grades of Evidence

Grade	Evidence Quality for Diagnostic Tests	Evidence Quality for All Other Studies
A	Systematic review of cross-sectional studies with consistently applied reference standard and blinding	Well-designed randomized controlled trials performed on a population similar to the guideline's target population
B	Individual cross-sectional studies with consistently applied reference standard and blinding	Randomized controlled trials; overwhelmingly consistent evidence from observational studies
C	Nonconsecutive studies, case control studies, or studies with poor, nonindependent, or inconsistently applied reference standards	Observational studies (case control and cohort design)
D	Mechanism-based reasoning or case reports	
X	Exceptional situations where validating studies cannot be performed and there is a clear preponderance of benefit over harm	

a participant earns a living, and the participant's previously established "stake" in an issue.<sup>30</sup>

### Guideline Key Action Statements

Each evidence-based statement is organized in a similar fashion: an evidence-based statement in bold, followed by the strength of the recommendation in italic. Several paragraphs subsequently discuss the evidence base supporting the statement. In addition, each evidence-based statement is followed by an "action statement profile" of aggregate evidence quality, benefit-harm assessment, and statement of costs. Last, there is an explicit statement of the value judgments, the role of patient preferences, clarification of any intentional vagueness by the panel, and a repeat statement of the strength of the recommendation. An overview of the evidence-based statements in the guideline is shown in **Table 3**.

The role of patient preference in making decisions deserves further clarification. For some statements, the evidence base may demonstrate clear benefit, which would minimize the role of patient preference. If the evidence is weak or benefits are unclear, however, not all informed patients might decide to follow the suggestion. In these cases, the practice of shared decision making, where the management decision is made by a collaborative effort between the clinician and the informed patient, becomes more useful. Factors related to patient preference include (but are not limited to) absolute benefits (number needed to treat), adverse effects (number needed to harm), cost of drugs or procedures, and frequency and duration of treatment.

**STATEMENT 1. EXCLUSION OF CONDUCTIVE HEARING LOSS: Clinicians should distinguish sensorineural hearing loss (SNHL) from conductive hearing loss (CHL) in a patient presenting with sudden hearing loss.** *Strong recommendation based on evidence with a preponderance of benefit over harm.*

#### Action Statement Profile for Statement 1

- Aggregate evidence quality: Grade B, based on evidence that a common cause of CHL, cerumen impaction, can be treated effectively to improve hearing.

Grade C, for evidence that CHL and SNHL can be distinguished from history, examination, and tuning fork tests

- Benefit: Guide the choice of appropriate diagnostic tests, identify patients with more serious underlying conditions, avoid misdiagnosis, improve diagnostic accuracy, ensure treatment is consistent with diagnosis, guide patient expectations, identify conductive hearing loss that can be treated and resolved
- Risk, harm, cost: Adverse effects of cerumen removal, if required; time required for cerumen removal, if required; misdiagnosis
- Benefit-harm assessment: Preponderance of benefit
- Value judgments: Panel consensus that despite a lack of systematic research evidence supporting this action, distinguishing these types of hearing loss was an essential first step in determining subsequent management
- Intentional vagueness: The panel intentionally decided not to specify the time frame to distinguish CHL from SNHL because of inconclusive evidence of the importance of early intervention but agreed that the distinction should be made as promptly as possible to allow intervention if a diagnosis of SSNHL is confirmed. Ideally, the determination should be made at the time of initial presentation.
- Role of patient preferences: No role
- Exclusions: None
- Policy level: Strong recommendation

#### Supporting Text

The purpose of this statement is to emphasize that the differentiation of CHL from SNHL is essential for defining potential treatments and prognosis. These 2 common causes of hearing loss can be diagnosed by a combination of history, physical examination, tuning fork tests, and audiometry. Conductive hearing loss and SNHL have markedly different management strategies, and there is good evidence that CHL, such as that from cerumen impaction, can be treated effectively.<sup>31</sup> A delay in treatment of SSNHL may result when a clinician assumes a patient has CHL without considering a diagnosis of SNHL.<sup>5</sup>

Hearing loss is classified as conductive, sensorineural, or mixed. Conductive hearing loss is a result of abnormalities of

**Table 3.** Summary of Evidence-Based Statements

Management of Patients with Sudden Hearing Loss (Evidence-Based Statement)	Statement Strength
<i>Diagnosis</i>	
Exclusion of conductive hearing loss (Statement 1)	Strong recommendation
Modifying factors (Statement 2)	Recommendation
Computed tomography (Statement 3)	Strong recommendation against
Audiometric confirmation of idiopathic sudden sensorineural hearing loss (Statement 4)	Recommendation
Laboratory testing (Statement 5)	Strong recommendation against
Retrocochlear pathology (Statement 6)	Recommendation
<i>Shared decision making</i>	
Patient education (Statement 7)	Strong recommendation
<i>Treatment</i>	
Initial corticosteroids (Statement 8)	Option
Hyperbaric oxygen therapy (Statement 9)	Option
Other pharmacologic therapy (Statement 10)	Recommendation against
Salvage therapy (Statement 11)	Recommendation
<i>Follow-up</i>	
Outcomes assessment (Statement 12)	Recommendation
Rehabilitation (Statement 13)	Strong recommendation

the external ear, tympanic membrane, middle ear air space, or ossicles—that is, structures that “conduct” sound waves to the cochlea. Sensorineural hearing loss is a result of abnormalities of the cochlea, auditory nerve, or other structures that translate neural impulses to the auditory cortex of the brain. Mixed hearing loss is a combination of both CHL and SNHL.

### History

Clinicians should ask patients about a history of trauma, external ear and canal pain, ear drainage, fever, or other systemic symptoms. (Also see Statement 2 for additional key elements of the patient history.) Patients cannot accurately distinguish subjective hearing loss as either CHL or SNHL. Patients with SSNHL often report tinnitus, ear fullness or pressure, and vertigo.<sup>8,17</sup> These symptoms, however, may also be present in CHL. Therefore, a focused physical examination is required.

### Physical Examination

Inspection of the ear canals and visualization of the tympanic membranes are essential in SHL to distinguish CHL from SNHL. Causes of CHL include cerumen impaction, middle ear fluid, otitis media, foreign bodies, perforated tympanic membrane, canal edema from otitis externa, otosclerosis, trauma, and cholesteatoma. Many of these conditions can be diagnosed by otoscopy. Pneumatic otoscopy, audiometry, and tympanometry can also help guide diagnosis. Patients with SNHL will almost always have a normal otoscopic examination, whereas examination of patients with CHL will often show abnormalities.<sup>32</sup> Impacted cerumen, if present, must be removed prior to establishing a diagnosis in patients with SHL.<sup>31</sup>

The Weber and Rinne tuning fork tests have been used traditionally to differentiate CHL and SNHL (**Table 4**).<sup>5,33</sup>

Research evidence, however, is sparse regarding the utility of the Weber and Rinne tuning fork tests,<sup>34</sup> and no published studies specifically evaluate the use of tuning fork tests in the diagnosis of SHL. Several authors have noted that the Weber and Rinne tuning fork tests can be misleading.<sup>32,35,36</sup> Despite the limitations in the literature, the panel agreed that tuning fork tests should be used to confirm audiometric findings. Furthermore, when Weber and Rinne results are unequivocal, they can still help clinicians make a preliminary diagnosis of CHL or SNHL if audiometry has not been performed.

**STATEMENT 2. MODIFYING FACTORS: Clinicians should assess patients with presumptive sudden sensorineural hearing loss for bilateral sudden hearing loss, recurrent episodes of sudden hearing loss, or focal neurologic findings.** *Recommendation based on observational studies with a preponderance of benefit over harm.*

### Action Statement Profile for Statement 2

- Aggregate evidence quality: Grade C, observational studies and case series
- Benefit: Identification of patients with a high likelihood of alternative and potentially serious underlying cause, who require specialized assessment and management
- Risk, harm, cost: None
- Benefit-harm assessment: Preponderance of benefit
- Value judgments: None
- Intentional vagueness: None
- Role of patient preferences: Limited
- Exclusions: None
- Policy level: Recommendation

**Table 4.** Recommended Technique for Weber and Rinne Testing

Weber Test	Rinne Test
<ol style="list-style-type: none"> <li>Place vibrating tuning fork (256 or 512 Hz) at midline of forehead or on maxillary teeth (not false teeth)</li> <li>Ask where the sound is heard; it is normal to hear at the midline or “everywhere”</li> <li>If the sound lateralizes to one ear then:               <ol style="list-style-type: none"> <li>There is a CHL in that ear, OR</li> <li>There is SNHL in the opposite ear</li> </ol> </li> </ol>	<ol style="list-style-type: none"> <li>Place vibrating tuning fork (256 or 512 Hz) over the mastoid bone of one ear; then move the tuning fork to the entrance of the ear canal (not touching the ear)</li> <li>The sound should be heard better via air conduction (at the entrance to the ear canal).</li> <li>If the sound is heard better by bone conduction, then there is a CHL in that ear. Repeat for the other ear.</li> </ol>

Abbreviations: CHL, conductive hearing loss; SNHL, sensorineural hearing loss.

**Table 5.** Checklist of Features Often Associated with Specific Disorders Underlying Hearing Loss

Sudden onset of bilateral hearing loss
Antecedent fluctuating hearing loss on one or both sides
Isolated low-frequency hearing trough suggesting Meniere disease
Concurrent onset of severe bilateral vestibular loss with oscillopsia
Accompanying focal weakness, dysarthria, hemiataxia, encephalopathy, severe headaches, diplopia
Downbeating or gaze-evoked nystagmus
Brain imaging indicating stroke or structural lesion likely to explain the hearing loss
Severe head trauma coincident with the hearing loss on one or both sides
Recent acoustic trauma
A history of concurrent or recent eye pain, redness, lacrimation, and photophobia

### Supporting Text

The purpose of this statement is to encourage clinicians to look for clinical features in patients with presumptive SSNHL that may be associated with a definable underlying disease at presentation. Among such causes are systemic disorders, autoimmune disorders, metabolic disorders, bilateral Meniere disease, and certain neurological disorders.<sup>37</sup> If any of these are identified, the patient may not have SSNHL as defined in this guideline and should be managed in accordance with the suspected diagnosis.

The clinician should assess the patient for these conditions by history, general physical and neurologic examination, and audiometry when available. The clinician should ask about prior episodes of unilateral or bilateral hearing loss, vertigo, and focal neurological symptoms (**Table 5**). Prior audiometric test results and neurological workup, when available, should be reviewed.

### Bilateral Sudden Hearing Loss

The sudden onset of bilateral sensorineural hearing loss is relatively rare<sup>14,38</sup> and should raise concern for certain specific causes, some of which are outlined in **Table 6**.

The cause of disorders leading to sudden bilateral sensorineural hearing loss may be vascular, metabolic, autoimmune, infectious, neoplastic, toxic, traumatic, or inflammatory. Acute bilateral hearing loss may occur by any of these mechanisms, but on rare occasions, these same mechanisms may also produce unilateral hearing loss.

### Prior Episodes of SHL or Fluctuating Hearing Loss

Most cases of SSNHL are not preceded by fluctuating hearing, so this feature in the history should raise suspicion for

other causes. Patients with a prior history of a fluctuating hearing loss presenting with SSNHL should be evaluated for causes such as Meniere disease, autoimmune inner ear disease, Cogan syndrome, and hyperviscosity syndromes. Meniere disease is by far the most common disease in this category encountered in clinical practice.<sup>39</sup> The prior history of fluctuation suggests a process that has been ongoing that culminates in an abrupt hearing loss that is usually unilateral and, less often, bilateral. Autoimmune inner ear disease and Cogan syndrome may be exceptions in which bilateral involvement is common at onset.<sup>40,41</sup> In all these conditions, hearing declines in a stepwise or fluctuating manner but may occasionally decline suddenly and thus present as SSNHL.

### SHL with Focal Neurological Findings

SHL in the presence of new focal neurological symptoms or signs indicates a central nervous system process. There are no RCTs specifically pertaining to strokes presenting with SHL. There are, however, ample data indicating that early recognition and treatment of stroke improve outcome,<sup>42-44</sup> so proper recognition of SHL as part of a broader cerebrovascular event is important.

Occlusion of the internal auditory artery may be the most common mechanism for acute unilateral hearing loss with a stroke. Because the internal auditory artery derives its blood supply from larger vessels, often the anterior inferior cerebellar artery (AICA), atherosclerotic disease or vascular dissection or thrombosis in the distal vertebral arteries or proximal basilar artery may also lead to stroke in the AICA distribution. The affected areas include the middle cerebellar peduncle and

**Table 6.** Selected Conditions That May Be Associated with Bilateral Sudden Hearing Loss

Cause	Other Features
Meningitis (infectious, inflammatory, neoplastic)	Headache, fever, abnormal cerebrospinal fluid (CSF) studies, possibly other cranial nerve palsies <sup>211</sup>
Autoimmune inner ear disease	Fluctuation of hearing may sometimes occur; vertigo may occur in some cases. <sup>41</sup>
Lyme disease	Erythema chronicum migrans, abnormal CSF, fluctuating bilateral audiovestibular symptoms <sup>212</sup>
Syphilis	Abnormal fluorescent treponemal antibody absorption (FTA-abs) test, bilateral fluctuating hearing loss, tabes dorsalis, multiorgan involvement <sup>213</sup>
Ototoxic medications	Vestibular loss, oscillopsia <sup>214,215</sup>
Trauma	Significant head trauma, barotrauma, temporal bone fractures <sup>214</sup>
Herpes zoster oticus (Ramsay-Hunt syndrome)	Otalgia, pinna and/or ear canal vesicles, facial nerve paresis, positive viral titers, positive viral cultures <sup>216</sup>
Human immunodeficiency virus (HIV) otitis	Positive HIV titers, altered T cell counts, and often other cranial neuropathies may be associated with mastoiditis out of proportion to clinical complaints. <sup>217,218</sup>
Lead poisoning	Learning disabilities, other stigmata of lead poisoning <sup>219</sup>
Genetic disorders	May be syndromic or nonsyndromic <sup>220,221</sup>
MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes)	Periods of confusion, elevated serum lactic acid levels around times of attacks, stroke-like spells, magnetic resonance imaging (MRI) white matter signal changes, migraine-like headaches, seizures, diabetes, mitochondrial gene mutation (Mt-RNRI, Mt-TSI, POLG genes) <sup>222,223</sup>
Other mitochondrial disorders	Variable phenotypes <sup>224</sup>
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 angiogram of the vertebrobasilar vasculature <sup>48,50,225-227</sup>
Cogan syndrome	Nonsyphilitic interstitial keratitis of the cornea, hearing loss, vertigo <sup>40</sup>
Neoplastic (neurofibromatosis II, bilateral vestibular schwannomas, intravascular lymphomatosis, others)	Abnormal brain MRI or cerebrovascular imaging study <sup>228-230</sup>
Sarcoidosis	Pulmonary symptoms, bilateral vestibular loss, elevated serum angiotensin-converting enzyme level or abnormal Gallium scan <sup>231,232</sup>
Hyperviscosity syndrome	Mucous membrane bleeding, neurologic and pulmonary symptoms, associated retinopathy <sup>233</sup>

portions of the cerebellum and lateral pons.<sup>45,46</sup> Most cases of labyrinthine infarction tied to AICA distribution vascular disease are associated with both acute unilateral hearing loss and vestibular loss,<sup>47</sup> and unilateral hearing loss can occasionally be a manifestation of transient ischemic attacks in the AICA distribution.<sup>48</sup> Vestibular symptoms can also be the result of peripheral vestibular ischemia, infarction of the central vestibular structures in the lateral pontomedullary junction, or a combination of both.<sup>47,49</sup>

Features that may accompany ischemic hearing loss due to AICA occlusion include ipsilateral Horner syndrome (oculosympathetic paresis: miosis, ptosis, and anhidrosis), diplopia, nystagmus, ipsilateral facial weakness and numbness, vertigo, slurred speech, nausea and vomiting, ataxia, unilateral limb clumsiness, and contralateral loss of pain and temperature sensation.<sup>45,46,48</sup> Sudden bilateral hearing loss may also be a prodrome to a stroke in the AICA distribution when there is underlying severe atherosclerotic narrowing of the vertebrobasilar vessels.<sup>50,51</sup>

A unilateral stroke affecting the primary auditory cortex in the posteromedial temporal cortex of the brain (Heschl gyrus) does not typically lead to symptomatic hearing loss.<sup>52</sup> Bilateral strokes affecting the primary auditory cortex are rare but may

cause transient bilateral hearing loss. Patients may present with a prior history of fluctuating hearing loss with this condition, which may lead to reduced word recognition or auditory agnosia.<sup>53-55</sup> Strokes affecting territories rostral to the cochlear nuclei do not affect hearing as measured by conventional pure-tone audiometry and word recognition unless the stroke is bilateral. Many such extensive bilateral cerebral events are lethal.<sup>56</sup>

Studies suggest that SSNHL is associated with both acute and increased long-term risk of stroke. In a prospective series of 364 patients with acute posterior circulation stroke, hearing loss occurred in 8% of cases, sometimes preceding the stroke by several days.<sup>57</sup> One study quoted a 12.8% risk of stroke over the next 5 years in patients admitted with SSNHL vs 7.8% in controls; however, after adjustment for other confounding factors, such as hypertension, hypercholesterolemia, and diabetes mellitus, the hazard was 1.64 times that for non-SSNHL admissions (in this case, appendectomy).<sup>58</sup> The data do not meet our guidelines criteria for significance; however, the clinician should be aware of these studies and be prepared to discuss them with the patient.

Other central nervous system disorders that may infrequently present with SHL include multiple sclerosis,

carcinomatous meningitis, lymphomatous meningitis, and, very rarely, central nervous system (CNS) intravascular lymphomatosis and migrainous infarction.<sup>59</sup> Features that suggest multiple sclerosis would be unilateral weakness or numbness, visual loss, diplopia, or paraparesis. The MRI of the brain would likely show white matter signal abnormalities, particularly on fluid-attenuated inversion recovery (FLAIR) images. Meningitis, whether infectious, neoplastic, or inflammatory, will show elevated protein, increased cerebrospinal fluid (CSF) white blood cells (pleocytosis), and possibly other CSF abnormalities. Tumors or other structural lesions of the cerebellopontine angle that present with SSNHL may sometimes exhibit unilateral limb clumsiness, hemiataxia, and facial weakness. Vestibular schwannomas typically present with slowly progressive hearing loss but may sometimes present with SSNHL. The tumor size does not correlate with the abruptness of the hearing loss.<sup>8,37,60</sup>

**STATEMENT 3. COMPUTED TOMOGRAPHY:** Clinicians should not order computerized tomography of the head/brain in the initial evaluation of a patient with presumptive SSNHL. Strong recommendation against based on systematic reviews with a preponderance of benefit over harm for not obtaining CT.

### Action Statement Profile for Statement 3

- Aggregate evidence quality: Grade B, systematic reviews and appropriateness criteria from the American College of Radiology (ACR), plus observational studies clearly documenting the potential harms of radiation and side effects of intravenous contrast
- Benefit: Avoidance of radiation, cost savings, reduced incidental findings, less inconvenience for the patient, avoiding false sense of security from false-negative scan
- Risk, harm, cost: None
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: The panel recognizes that the term *initial evaluation* is vague, but the intent is to discourage the routine use of CT scanning of the head/brain when patients initially present with SSNHL
- Role of patient preferences: Very limited
- Exclusions: Patients with focal neurologic findings
- Policy level: Strong recommendation against

### Supporting Text

The purpose of this statement is to avoid inappropriate use of CT of the head/brain in the initial assessment of patients with presumptive SSNHL. Computed tomography scanning has potential significant adverse events, which include radiation

exposure and side effects of intravenous contrast, while offering no useful information that would improve initial management. This statement does not apply to patients with focal neurological findings (as identified in the preceding statement), a history of trauma, or chronic ear disease who may require CT scanning. This statement also does not imply that imaging studies are of no use in managing SSNHL patients, who may eventually benefit from MRI of the brain or fine-cut, high-resolution CT scanning of the temporal bone (not routine head/brain; see Statement 6).<sup>37,61</sup>

The ACR has defined evidence-based Appropriateness Criteria (ACR-AC) for imaging studies with a rating of 1 to 3 for “usually not appropriate,” 4 to 6 for “may be appropriate,” and 7 to 9 for “usually appropriate.”<sup>62</sup> A head CT, with or without contrast, in the scenario of acute hearing loss and vertigo receives only a rating of 3, meaning that under most circumstances, the study or procedure is unlikely to be indicated in these specific clinical settings or the risk-benefit ratio for patients is likely to be unfavorable.<sup>63</sup> None of the ACR scenarios, however, are limited to isolated sudden hearing loss, which would achieve an even lower rating of appropriateness. In the current guideline criteria, the panel would assume that the history and physical examination would have determined whether a cholesteatoma or other pathologic condition was present and, if so, a targeted temporal bone CT would then be more appropriate.

The ACR, as part of the Appropriateness Criteria, introduced a radiation dose assessment and relative radiation levels (RRLs) associated with different diagnostic tests.<sup>64</sup> The RRL is expressed in a dose range of milliSieverts (mSv), which is a measure of absorbed radiation. The RRLs range from 0 to 5. An ultrasound or MRI scan offers no radiation exposure, so its RRL is 0; a chest X-ray in an adult has an RRL of 1, with a radiation dose estimate of less than 0.1 mSv; and a head CT scan has an RRL of 3, with a radiation dose of 1 to 10 mSv. Therefore, a nontargeted head/brain CT should be considered not only inappropriate but, in fact, unnecessarily harmful in the evaluation of SSNHL.

The principal differential diagnosis in the patient with suspected SSNHL is an inner ear vs an audiovestibular nerve or brainstem abnormality. No imaging modality currently shows the fine details of the inner ear, so the concern becomes differentiating possible central etiologies. The MRI scan has long replaced CT, or CT with air contrast, as the study of choice for detecting cerebellopontine angle tumors.<sup>61,65-74</sup> Also, the CT scan does not have the resolution to detect brainstem infarcts in the early stages, and emergent MRI is preferred when the clinical situation warrants emergency imaging.

There are other situations where this guideline recommendation would not apply. A CT would be used in situations where an MRI could not be obtained, such as patients with pacemakers, severe claustrophobia, or even financial constraints. Other considerations could be patients with known bone disease, such as Paget disease, fibrous dysplasia, or bone metastasis to the temporal bone, although the history would be used as a guide in these cases.<sup>75</sup>

In summary, the decision to seek imaging in patients with presumptive SSNHL may come early in the evaluation and before audiometric evaluation. In patients with no etiology found on history or physical examination and in whom SSNHL is suspected, CT scanning will be a very low-yield examination with significant cost and radiation exposure and is not recommended.

**STATEMENT 4. AUDIOMETRIC CONFIRMATION OF ISSNHL: Clinicians should diagnose presumptive ISSNHL if audiometry confirms a 30-dB hearing loss at 3 consecutive frequencies AND an underlying condition cannot be identified by history and physical examination. Recommendation based on randomized controlled trials with a preponderance of benefit over harm.**

#### Action Statement Profile for Statement 4

- Aggregate evidence quality: Grade C, based on criteria used in RCTs assessing the benefits for intervention for SSNHL
- Benefit: Guiding treatment, identifying urgent conditions that require prompt management, ensuring that interventions for ISSNHL are limited to those patients who meet appropriate audiometric criteria for diagnosis
- Risk, harm, cost: Potential delay in treatment until audiometry is obtained; direct cost of audiometry
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: Although there is limited evidence as to the audiometric cut points for the definition of SSNHL, this definition has been used widely
- Intentional vagueness: None
- Role of patient preferences: None
- Exclusions: When audiometry is not available, clinical judgment should be used, based on history, examination, and tuning fork evaluation. Lack of audiometry should not preclude discussion of, and initiation of, treatment.
- Policy level: Recommendation

#### Supporting Text

The purpose of this statement is to identify objective, reproducible criteria for diagnosing a patient with ISSNHL. Audiometry is mandatory for definitively diagnosing SSNHL because it distinguishes CHL from SNHL and establishes frequency-specific hearing thresholds. Varying criteria have been used in the literature to diagnose SSNHL, but a hearing loss  $\geq 30$  dB at 3 consecutive frequencies is the definition adopted by the NIDCD and the definition used in most RCTs.<sup>3,76</sup> The adoption of the criteria proposed in this statement will increase the generalization of research findings

by ensuring that patients are similar to those studied by the investigators.

The guideline panel adopted the following definition of SSNHL: a hearing loss of  $\geq 30$  dB affecting at least 3 contiguous frequencies occurring over a 72-hour period.<sup>3,76</sup> This definition assumes that the premorbid hearing level in each ear was either normal just prior to the episode of SSNHL or that premorbid hearing loss was symmetrical in each ear. Clinicians must decide the *degree of certainty* they are comfortable with when making a decision that the hearing loss in the poorer ear is “new.”<sup>77</sup> There are 4 levels of “certainty” about the “newness” of the hearing loss in the effected ear:

1. **Very certain:** patient had previous audiometric evaluation.
2. **Certain:** patient had no prior otologic history and feels his or her premorbid hearing was normal bilaterally.
3. **Fairly certain:** patient had a longstanding hearing problem and reports that the current episode of SSNHL is subjectively poorer.
4. **Uncertain:** the clinician feels there was some pre-existing hearing loss, but the hearing loss was never documented.

Accurate audiometric evaluations initially and during follow-up are essential for proper management of patients with sudden hearing loss. Thus, initial audiometric evaluations should follow *Preferred Practice Patterns*<sup>78</sup> that include all of the following components:

- a. A thorough case history
- b. Otoscopy with removal of excessive or obstructive cerumen
- c. Current American National Standards Institute (ANSI) standards should be met regarding maximum allowable ambient noise levels in the test environment<sup>79</sup>; calibration of the audiometer<sup>80</sup>; audiogram documentation, including use of the proper aspect ratio<sup>80-82</sup>; and symbols.<sup>83</sup> Ear-specific, masked air and bone conduction thresholds, speech recognition threshold (SRT), and word recognition scores (WRS) should be obtained. Reliability and validity of test results should be documented. Air conduction (AC) thresholds should be measured at 250 to 8000 Hz. Additional mid-octave frequencies that may be helpful include 750, 1500, 3000, and 6000 Hz and should be measured if differences in thresholds at 500 and 1000 or 1000 and 2000 Hz are  $\geq 20$  dB hearing level (HL). Bone conduction (BC) thresholds should be measured at 250 to 4000 Hz.
- d. Ear-specific SRT in dB HL should be measured using standardized spondee word lists (eg, CID W-1), preferably recorded, but monitored-live voice (MLV) is acceptable. Agreement between pure-tone average

(PTA) and SRT is helpful in discriminating the presence of a legitimate from questionable SSNHL.

- e. Ear-specific masked WRS (in %) should be measured at a presentation level of a 30- to 40-dB sensation level regarding SRT using recorded versions of monosyllabic word lists (ie, NU-6, W-22, etc) and different word lists for each ear. The clinician managing the patient with SSNHL will of necessity rely on the results of serial audiometric evaluations. As such, there is a need for proper audiologic documentation, not only of AC and BC thresholds, as well as SRT and WRS, but also of masking levels, reliability, validity, words lists used, method of presentation (MLV or recorded), and type of transducer, in order for ongoing comparisons to be useful.<sup>84,85</sup>
- f. Ear-specific *immittance measurements* may be completed on each ear using equipment calibrated to current ANSI standards. Immittance measures may include the following:
  1. Ear-specific *tympanograms*
  2. Ear-specific *contralateral acoustic reflex thresholds* (dB HL) at 500 to 4000 Hz
  3. Ear-specific *ipsilateral acoustic reflex thresholds* (dB HL) at 500 to 4000 Hz
  4. Ear-specific *acoustic reflex decay* (dB HL) at 500 and 1000 Hz

In situations of limited resources and/or access to audiometry, automated audiometry can be considered a secondary alternative.

**STATEMENT 5. LABORATORY TESTING: Clinicians should not obtain routine laboratory tests in patients with ISSNHL.** *Strong recommendation against based on large cross-sectional studies showing a preponderance of benefit over harm.*

#### Action Statement Profile for Statement 5

- Aggregate evidence quality: Grade B, based on small cross-sectional studies showing no benefit as well as case series
- Benefit: Cost containment, avoidance of stress and anxiety of patient, avoidance of false positives, avoidance of delay of diagnosis, avoidance of delayed treatment
- Risk, harm, cost: Missed diagnosis
- Benefit-harm assessment: Preponderance of benefit
- Value judgments: Minimizing testing and the risks of false positives outweigh the value of finding a potential cause, especially when it has not been shown that early treatment affects prognosis
- Intentional vagueness: The word *routine* was to discourage a nontargeted approach to use of laboratory assessment. It is recognized that specific laboratory

tests may be useful in assessing these patients based on specific individual patient conditions.

- Role of patient preferences: Limited
- Exclusions: None
- Policy level: Strong recommendation against

#### Supporting Text

The purpose of this statement is to discourage routine laboratory tests that do not improve management or care of patients with ISSNHL but nonetheless have associated cost and potential harms related to false-positive results, false-negative results, or both. The word *routine* is used in this context to define automatic, shotgun, or universal testing done without consideration of specific patient or geographic risk factors. The panel recognizes that specific tests may be warranted in selected patients if pertinent history suggests that a specific laboratory test might be useful for identifying a specific potential cause of the hearing loss, such as drawing Lyme titers in endemic regions.

The evidence regarding the use of routine laboratory tests in patients with SSNHL is limited to observational and case control studies. Most studies are limited by a small sample size and the lack of evidence that knowing the result of the test would improve outcomes.

Possible etiologies of SSNHL include viral infection, vascular impairment, autoimmune disease, inner ear pathology, and central nervous system anomalies, although the cause in most patients is never identified.<sup>37</sup> Serologic studies of viral or mycoplasma infection or rheumatologic disease with sudden deafness found varying associations with SSNHL and inconsistent correlation with response to steroids.<sup>86,87</sup> There is some evidence of an association of autoimmune disease with ISSNHL.<sup>88</sup> The antibody response was transient in most patients, which led those authors to suggest that a transient phenomenon may trigger antibody activity that produces the hearing loss. In a study of 48 patients, researchers found no association between ISSNHL and abnormal levels of anti-thrombin III, protein C, D-dimer, or fibrinogen or activated protein C resistance.<sup>89</sup>

Another study evaluated serum and CSF markers in 19 patients with SSNHL.<sup>90</sup> However, the failure to show a therapeutic benefit by acting on the laboratory results limits the usefulness of the data. Similarly, ISSNHL co-occurring with diabetes, hypertension, and hyperlipidemia in older patients has been shown to be associated with MRI evidence of cerebral microangiopathy and prognosis, but the association's clinical significance is unclear. Although a low level of thyroid-stimulating hormone (TSH) was shown to be a positive prognostic factor in a study of 133 patients with SSNHL, the study did not take into account its multiple comparisons performed, and so the results lack statistical significance.<sup>91</sup> A case control study showed a relationship between low folate and SSNHL (all 44 cases had low levels), but the clinical implications of the study are not clear.<sup>92</sup> Other factors that have been associated with hearing loss are fatty acids, coenzyme-Q, uronic acid, and C3b.<sup>93,94</sup>

As noted above, any test may lead to an evaluation of a false-positive result. This evaluation carries medical, psychological, and financial costs. Unless there is evidence of potential gain from a specific test, the potential harm will outweigh any potential benefits of performing the test. Currently, there is insufficient evidence that any routine laboratory test will result in changes to the diagnosis, treatment, or prognosis. All studies listed in this section are limited by sample size or their observational nature. Positive studies, such as the association between low TSH and prognosis, should lead to more research to confirm the association and then to evaluate the clinical ramifications of the finding.

**STATEMENT 6. RETROCOCHLEAR PATHOLOGY: Clinicians should evaluate patients with ISSNHL for retrocochlear pathology by obtaining an MRI, auditory brainstem response (ABR), or audiometric follow-up.** *Recommendation based on observational studies with a preponderance of benefit over harm.*

#### Action Statement Profile for Statement 6

- Aggregate evidence quality: Grade C
- Benefit: Identify brain tumors, identify conditions that might benefit from early treatment, patient peace of mind, supporting idiopathic diagnosis
- Risk, harm, cost: Procedure-specific risks/costs, anxiety, and stress
- Benefit-harm assessment: Preponderance of benefit
- Value judgments: Although the panel agreed that the MRI is the most sensitive means for diagnosing retrocochlear pathology, there was no consensus that identifying this pathology would in all cases influence outcomes. The panel therefore concluded that ABR and follow-up audiometry would be acceptable alternatives for initial follow-up of ISSNHL as long as there is appropriate counseling about the limitations of these modalities.
- Intentional vagueness: None
- Role of patient preferences: Limited in deciding whether or not to assess for retrocochlear pathology but substantial in making shared decisions with the clinician for using MRI, ABR, or audiology as the diagnostic test
- Exclusions: None
- Policy level: Recommendation

#### Supporting Text

The purpose of this statement is to ensure that clinicians detect retrocochlear pathology in patients with ISSNHL because a small but significant percentage of such patients have an underlying lesion, most often a vestibular schwannoma. Retrocochlear pathology is defined as a structural lesion of the vestibulocochlear nerve, brainstem, or brain. An

MRI of the brain, brainstem, and internal auditory canals (IACs) with gadolinium is the most sensitive test for detecting retrocochlear pathology, but persistent abnormalities on ABR or audiometry would also be indicative and usually require an MRI for further assessment. Patients with normal ABR results or stable findings on audiometric follow-up may decide whether to pursue additional testing with MRI based on shared decision making with the clinician. However, screening ISSNHL patients for vestibular schwannoma represents an opportunity for early identification of the tumor, affording them the most options for management and potentially the best chances of preserving hearing and facial nerve function.

#### Risk of Vestibular Schwannoma

Ten to twenty percent of patients with a vestibular schwannoma will report a sudden decrease of hearing at some point in their history,<sup>95</sup> but the rate of vestibular schwannoma in patients who present with SHL is somewhat lower, but still remarkable, with several studies demonstrating a relatively high prevalence of cerebellopontine angle tumors in SHL patients ranging from 2.7% to 10.2% of patients who are evaluated with MRI.<sup>4,10,60,96-98</sup>

Testing with MRI, ABR, or follow-up audiometry is important for detecting vestibular schwannoma because no clinical features can reliably distinguish SSNHL caused by an underlying tumor from the more common idiopathic variety.<sup>4</sup> Tinnitus in the affected ear prior to the onset of the SHL, associated otalgia, or paresthesias are more common in patients with vestibular schwannoma; however, these symptoms are too rare for their absence to reliably rule out a retrocochlear lesion. Although the risk of underlying tumor is lower in patients with low-frequency hearing loss, all types of audiometric patterns have been found in SSNHL patients with vestibular schwannomas.<sup>4,97</sup>

Associated events or diseases (eg, barotrauma or recent viral infection) that were presumed to cause the SSNHL are also present in approximately one-third of patients with vestibular schwannoma. Hearing recovery has not been shown to predict whether a patient's SHL is the result of a tumor.<sup>4,98</sup> Sudden hearing loss may be the presenting symptom in a variety of tumor sizes. The mean tumor size in one large study was 2.1 cm, with 10% of tumors over 3 cm in size.<sup>4</sup> Therefore, all patients should be apprised of the risk of a vestibular schwannoma and counseled regarding the various diagnostic strategies and management options.

There are no RCTs comparing a strategy of investigation vs no investigation for vestibular schwannoma in patients with SSNHL. Vestibular schwannomas are mostly slow-growing tumors; one-third to one-half of tumors do not grow on serial follow-up examinations.<sup>99</sup> Many patients do well with no intervention, "undisturbed by their tumors, ultimately dying with them but not because of them."<sup>99</sup> The early diagnosis of vestibular schwannoma is associated with smaller tumor size, which may have advantages regardless of the management strategy. The treatment of smaller tumors is associated with better outcomes with both surgical<sup>100-102</sup> and radiotherapy treatment.<sup>103-105</sup> Smaller tumors are also more suitable for conservative management.<sup>106</sup> The conservative approach may be

a particularly good option in patients with small tumors; only 20% to 25% of patients in selected populations will fail conservative treatment.<sup>106,107</sup> Although surgical, radiosurgical, and conservative approaches are often offered as choices for the treatment of vestibular schwannoma, no RCTs have compared these various approaches.<sup>108</sup>

The costs of screening tests for vestibular schwannoma compare favorably to the additional cost of treating larger tumors.<sup>109</sup> Given this advantage and the higher prevalence of tumors in patients with SSNHL, all patients with SSNHL should be evaluated for vestibular schwannoma. The clinician should not be dissuaded from a workup for retrocochlear pathology by the presence of associated diseases, the audiometric pattern, normal electronystagmography (ENG), or hearing recovery.

### Magnetic Resonance Imaging

An MRI is the gold standard for vestibular schwannoma diagnosis and is more cost-effective than ABR followed by MRI.<sup>99</sup> The specific MRI protocol used will often depend on the neuro-radiological resources available. Magnetic resonance imaging with gadolinium enhancement is extremely sensitive and widely available. High-resolution fast-spin echo or gradient echo MRI imaging (eg, FIESTA protocol) of the internal auditory canal has been shown to be both sensitive in the diagnosis of vestibular schwannoma in patients with SSNHL and more cost-effective than gadolinium-enhanced MRI.<sup>99,110</sup> Fast-spin echo techniques may require technological and radiographic expertise that is not always available in the community.

Magnetic resonance imaging has the added advantage of identifying other causes of SSNHL (eg, cochlear inflammation or multiple sclerosis) or findings that imply an underlying etiology for the SSNHL (eg, small vessel cerebral ischemia) (Table 7). The overall rate of pathogenic MRI abnormalities directly related to the SSNHL ranges from 7% to 13.75%.<sup>60,98,111-113</sup> Therefore, MRI has the highest yield of any diagnostic test in the setting of SSNHL.

For patients in whom MRI is contraindicated (ie, pacemakers, other metallic implants, claustrophobia), a fine-cut CT of the temporal bones with contrast may be used.

One disadvantage of MRI is the possibility of incidental findings not related to the hearing loss that may result in patient anxiety or additional evaluation. In one study of patients with SHL, 57% of the MRI studies revealed some abnormality, but only 20% of these findings were directly related to the hearing loss.<sup>113</sup> In another study, the overall rate of abnormal findings was 34.5%, with 36% of these directly related to the hearing loss.<sup>112</sup> In general, the rate of incidental findings in patients with audiovestibular symptoms is significant (47.5%), but only a small fraction of these (2.5%) required additional referral or investigation.<sup>114</sup> The cost and consequences of these incidental findings on MRI are difficult to assess. A second concern with MRI is the potential for rare immediate reactions to gadolinium (<1%) or gadolinium-induced nephrogenic systemic fibrosis.<sup>115,116</sup> Fortunately, the latter is rare in patients without preexisting renal disease. These contrast-related risks can be avoided with fast-spin

**Table 7.** Number of SSNHL Patients with MRI Abnormalities (n = 82)

	No.	%
<b>Obvious etiology for SSNHL</b>		
Vestibular schwannoma	4	5
• Intracanalicular	(2)	
• CPA	(2)	
Obliterated internal carotid artery	1	1
Infarction (pons)	1	1
<b>Possible etiology for SSNHL</b>		
Blood vessel anomalies	4	5
• Prominent vertebral artery	(1)	
• Elongated basilar artery	(1)	
• Carotidocavernous fistula	(1)	
• Venous angioma	(1)	
Demyelinating processes	2	2
<b>Unknown causal relationship for SSNHL</b>		
Capillary hemangioma (pons)	1	1
White matter diseases	4	5
Asymmetry of the cerebellum	1	1
Meningioma originating from the tentorium	1	1
Parasagittal meningioma	1	1
Cochlear deformity in the contralateral ear	1	1
AICA loop	3	4
Frontal posttraumatic changes	1	1
Enhancement of mastoid cells	3	4
Enhancement of endolymphatic duct	1	1

Adapted from Aarnisalo et al.<sup>60</sup> Abbreviations: AICA, anterior inferior cerebellar artery; CPA, cerebellopontine angle; MRI, magnetic resonance imaging; SSNHL, sudden sensorineural hearing loss.

echo MRI. Clearly, the patient and clinician should discuss these issues thoroughly before proceeding with an MRI scan in this setting.

### Auditory Brainstem Response

The ABR test may be used to initially evaluate these patients in the appropriate scenario (eg, older patients in whom the missed diagnosis of a small tumor may be less consequential). The ABR test is highly sensitive for a vestibular schwannoma greater than 1 cm in size<sup>99</sup>; however, ABR testing has limits.<sup>117</sup> The reported sensitivity of ABR for small vestibular schwannomas varies widely from 8% to 42%,<sup>118-120</sup> and ABR is not possible when the hearing loss exceeds 80 dB in the 2000- to 4000-Hz range and may be problematic with even lesser degrees of hearing loss. The sensitivity of ABR is proportional to the degree of hearing loss; therefore, mild hearing losses or those that have recovered will be more likely to yield false-negative ABR results.<sup>121</sup>

### Audiometric Follow-up

Although ABR and MRI are generally indicated to evaluate for retrocochlear pathology in patients with SSNHL, serial audiometry is an option in selected patients. Obviously, patients with a complete hearing loss are not eligible for this strategy. For patients with some degree of residual hearing after the episode of SSNHL, repeated hearing tests looking

for progression can be used as an indicator of patients with higher likelihoods of retrocochlear pathology. Serial audiometry will not identify retrocochlear pathology directly and is not as effective as either MRI or ABR. In addition, for patients with a vestibular schwannoma, growth is possible without immediate progression of hearing loss. Nonetheless, given the benign nature of the vast majority of retrocochlear lesions and the relatively low incidence of retrocochlear pathology in patients with SHL, it is an option. With shared decision making, serial audiometric follow-up may be appropriate for older patients in whom aggressive treatment of a retrocochlear lesion is less likely, patients unable to tolerate an MRI, or patients with financial or other concerns leading them to select a less definitive evaluation strategy with the understanding that it could lead to a delay in diagnosis.

For patients electing this method, a follow-up hearing test should be performed in 6 months. In the panel's opinion, a progressive loss of hearing of greater than 10 dB (HL) in 2 or more frequencies or a drop in word recognition scores of greater than 10% should trigger an evaluation with an ABR or MRI.<sup>122</sup>

**STATEMENT 7. PATIENT EDUCATION: Clinicians should educate patients with ISSNHL about the natural history of the condition, the benefits and risks of medical interventions, and the limitations of existing evidence regarding efficacy. Strong recommendation based on systematic reviews with a preponderance of benefit over harm.**

### Action Statement Profile for Statement 7

- Aggregate evidence quality: Grade B
- Benefit: Facilitate shared decision making, increase patient adherence to proposed therapy, empower patients, informed consent, link evidence to clinical decisions
- Risk, harm, cost: Time spent, miscommunication, patients get overwhelmed, patient anxiety
- Benefit-harm assessment: Preponderance of benefit
- Value judgments: Shared decision making is beneficial
- Intentional vagueness: None
- Role of patient preferences: Large
- Exclusions: None
- Policy level: Strong recommendation

### Supporting Text

The purpose of this statement is to emphasize the importance of shared decision making in developing a plan of care for patients with ISSNHL. Clinicians are encouraged to provide patients with the information necessary to participate fully in shared decision making (**Table 8**).

Patient involvement in making decisions with regard to their treatment plan is known to facilitate better compliance and desired outcomes and is now widely accepted in the United States.<sup>123</sup> Shared decision making refers to more comprehensive

patient counseling in which the clinician gives the patient personalized treatment options and outcomes, including the efficacy and probabilities for success. Patients share their values and the relative importance of the potential benefit or harm associated with the various options. By working together, they can reach agreement on the best treatment strategy.<sup>124</sup> There are 3 key elements to true shared decision making:

1. An involved patient and/or family
2. Full disclosure about the risk and benefits of all viable options
3. A shared process involving the clinician and the patient/family

Shared decision making may be limited by practical barriers such as time constraints rather than attitude or lack of interpersonal skills. A study demonstrated better patient confidence in the decision made and compliance with the treatment plan when consultations were conducted in settings with more time and fewer interruptions. These findings support the need for appropriate consultation time.<sup>125</sup> Maximizing the time available for successful shared decision making can be accomplished with the use of various decision aids.

Using decision aids to provide information can make health care decisions less difficult. These pamphlets or videos are designed to promote patient/family understanding of available options, consider the personal importance of possible benefit or harm, and participate in decision making. An updated review of 55 trials found that the use of decision aids improved patient/family knowledge of the options, created accurate risk perceptions of the associated benefits and harms, reduced difficulty with decision making, and increased participation in the process.<sup>126</sup>

A basic protocol for management would include a discussion of the following:

1. The diagnosis including the possible causes
2. The available treatment options
3. The risks and benefits associated with each form of treatment
4. Shared decision making. The clinician should use his or her expertise in assisting patients to evaluate the risk/benefit of treatment options in the context of their medical history. The clinician should focus on QOL and functional health status in addition to objective treatment outcomes.

**STATEMENT 8. INITIAL CORTICOSTEROIDS: Clinicians may offer corticosteroids as initial therapy to patients with ISSNHL. Option based on systematic reviews of randomized control trials with a balance between benefit and harm.**

### Action Statement Profile for Statement 8

- Aggregate evidence quality: Grade B, systematic reviews of randomized trials with methodological limitations
- Benefit: Hearing improvement

**Table 8.** Patient Education Discussion Points for Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL)

1. The cause of sudden sensorineural hearing loss (SSNHL) is often not readily apparent and thus called idiopathic. It rarely affects both ears and can be associated with other symptoms such as tinnitus, vertigo, and fullness in the ear.
2. Approximately one-third to two-thirds of patients with ISSNHL may recover some percentage of their hearing within 2 weeks.<sup>2</sup> Those who recover half of their hearing in the first 2 weeks have a better prognosis.<sup>234</sup> Patients with minimal change within the first 2 weeks are unlikely to show significant recovery.
3. Early recognition of ISSNHL is important. Although there is a lack of evidence-based research, it is generally accepted that early intervention may increase recovery.
4. Many treatments have been proposed for ISSNHL, but research about their effects is limited by small sample size and varying experimental designs. The benefits of therapy may include more prompt and complete recovery of hearing, but side effects also must be considered when choosing among the available options.
5. Watchful waiting is an alternative to active treatment as between one-third and two-thirds of patients may recover hearing on their own and can be monitored with repeat hearing tests.
6. Sudden hearing loss can be frightening and may result in embarrassment, frustration, anxiety, insecurity, loneliness, depression, and social isolation. Individual or group counseling can be helpful in supporting patients with ISSNHL.
7. Audiologic rehabilitation needs to be addressed as soon as the hearing loss is identified. This includes counseling and discussion of nonsurgical and surgical amplification and hearing restoration options.
8. Financial concerns should be addressed to ensure appropriate follow-up and testing in an effort to attain the best possible outcome.

- Risk, harm, cost: Oral corticosteroids: suppression of hypothalamic-pituitary-adrenal axis and Cushing-like syndrome, minimal with 10- to 14-day treatment; low cost. Intratympanic corticosteroids: Minimal systemic effect; local reactions of pain, tympanic membrane perforation, transient dizziness; high cost and multiple office visits
- Benefit-harm assessment: Balance of benefit vs harm
- Value judgments: Even a small possibility of hearing improvement makes this a reasonable treatment to offer patients, considering the profound impact on QOL a hearing improvement may offer
- Intentional vagueness: None
- Role of patient preferences: Large role for shared decision making with patients
- Exclusions: Oral steroids: medical conditions affected by corticosteroids such as insulin-dependent or poorly controlled diabetes, tuberculosis, and peptic ulcer disease, among others
- Policy level: Option

### Supporting Text

The purpose of this statement is to clarify the role of corticosteroids, a commonly employed treatment modality. Many trials have been published investigating the use of corticosteroids in patients with ISSNHL; however, these trials adopted a variety of methodologies and drew varying conclusions. There is laboratory evidence of an inflammatory cell death cascade in ISSNHL, which is modified by steroid therapy. The term *corticosteroid* refers to common synthetic glucocorticoids delivered via the oral, intravenous, and/or intratympanic routes. These steroids include prednisone, methylprednisolone, solumedrol, and dexamethasone. Corticosteroids are known to have sites of action in the inner ear, with efficacy in viral, vascular, syphilitic, autoimmune, endolymphatic hydrops (Meniere disease), and other etiologies of hearing loss.<sup>127,128</sup> Most studies, however, do not meet present-day criteria in terms of highest quality evidence, as identified by RCTs, systematic reviews, meta-analyses, or evidence reports.

### Systematic Reviews of Randomized Controlled Trials

A Cochrane review, first published in 2006 and updated in 2009, found only 2 trials that met their inclusion criteria, and both were of low methodological quality and with small numbers of subjects.<sup>16</sup> One trial showed a lack of effect of oral steroids compared with placebo, and one study showed a significant improvement in 61% of patients receiving corticosteroids compared with 32% in the control group (placebo and untreated patients). The authors concluded that the value of corticosteroid treatment for ISSNHL remained unclear due to the conflicting results of the studies.

In another systematic review, Conlin and Parnes (2007)<sup>7</sup> found no valid RCTs to determine the effectiveness of corticosteroids in SSNHL and pointed out limitations in landmark studies that such treatment has been traditionally based on. In a separate treatment meta-analysis reviewing 5 studies that met their inclusion criteria, the same authors concluded that there was no evidence that corticosteroid treatment was better than a placebo.<sup>6</sup>

A recent meta-analysis of various medical treatments, including corticosteroids, showed a slight but not statistically significant improvement with medical therapy compared with placebo.<sup>129</sup>

### Benefits vs Risks of Oral Corticosteroid Therapy for Individual Patients

On the basis of the studies cited above, the clinician might choose not to prescribe corticosteroids for ISSNHL. However, faced with a patient with the serious consequences of a severe to profound SSNHL, corticosteroid treatment is one of the few treatment options that has data showing efficacy, although even those data are somewhat equivocal.<sup>1,9,38,76,130-135</sup>

The greatest spontaneous improvement in hearing occurs during the first 2 weeks<sup>2</sup>; late recovery has been reported but is a rare event. In a similar fashion, treatment with corticosteroids appears to offer the greatest recovery in the first 2 weeks, with little benefit after 4 to 6 weeks.<sup>1,5,8,91,134,136-140</sup>

**Table 9.** General Guidelines for Corticosteroid Therapy for Idiopathic Sudden Sensorineural Hearing Loss (SSNHL)<sup>a</sup>

	Oral Corticosteroids	Intratympanic Corticosteroids
Timing of treatment	Immediate, ideally within first 14 days. Benefit has been reported up to 6 weeks following onset of sudden sensorineural hearing loss (SSNHL)	Immediate Salvage (rescue) after systemic treatment fails
Dose	Prednisone 1 mg/kg/d (usual maximal dose is 60 mg/d) or Methylprednisolone 48 mg/d or Dexamethasone 10 mg/d	Dexamethasone 24 mg/mL or 16 mg/mL (compounded), or 10 mg/mL (stock)Methylprednisolone 40 mg/mL or 30 mg/mL
Duration/frequency	Full dose for 7 to 14 days, then taper over similar time period	Inject 0.4 to 0.8 mL into middle ear space every 3 to 7 days for a total of 3 to 4 sessions
Technique	Do not divide doses	Anterosuperior myringotomy after topical anesthetic Inject solution into the posterior inferior quadrant via narrow-gauge spinal needle to fill middle ear space Keep head in otologic position (one side down, affected ear up) for 15 to 30 minutes
Monitoring	Audiogram at completion of treatment course and at delayed intervals	Audiogram before each subsequent injection, at completion of treatment course, and at delayed intervals Inspect tympanic membrane (TM) to ensure healing at completion of treatment course and at a delayed interval
Modifications	Medically treat significant adverse drug reactions, such as insomnia Monitor for hyperglycemia, hypertension in susceptible patients	May insert pressure-equalizing tube if planning multiple injections, but this increases risk of TM perforation May consider adding round window transport facilitator

<sup>a</sup>This table is designed to provide guidance for systemic and intratympanic steroid treatment for SSNHL. Treatment is routinely individualized by provider and per patient. The most important principles pertain to early institution of high enough dosages of treatment. Prednisone 1 mg/kg/d or its equivalent and/or adequate concentration of intratympanic dexamethasone or solumedrol should be administered.

For maximal treatment outcomes, recommended treatment doses of oral prednisone are given at 1 mg/kg/d in a single (not divided) dose, with the usual maximum dose of 60 mg daily, and treatment duration of 10 to 14 days.<sup>141</sup> Data comparing treatment protocols are limited, but one representative regimen uses the maximum dose for 4 days, followed by a 10-mg taper every 2 days.<sup>5</sup> The basis of selecting this dose rests on the maximum adrenal output of hydrocortisone (cortisol) of 200 to 300 mg/d during stress. Prednisone is 4 times, methylprednisolone is 5 times, and dexamethasone is 25 times more powerful than hydrocortisone. The equivalent dose of prednisone 60 mg is 48 mg for methylprednisolone and 10 mg for dexamethasone. Underdosage, by the above standards, is a possibility if attention is not given to these ratios. For example, the commonly prescribed methylprednisolone dose pack, which contains 4-mg tablets, provides 6 tablets the first day and 1 less on each subsequent day, for a total dose of 84 mg over 6 days.<sup>142</sup> This only gives the equivalent of 105 mg prednisone, compared with a total dose of 540 mg prednisone over 14 days for a 60-kg adult using the formula above. As noted above, early treatment is important, so the clinician should ensure that the patient is initially adequately dosed.

Potential side effects of systemic corticosteroid therapy are reported in many organ systems. Corticosteroids are hormones and have access to, as well as an effect on, all organ systems. The commonly used glucocorticoids, such as prednisone, have little mineralocorticoid, androgenic, or estrogenic effect, and the major systemic side effects are suppression of hypothalamic-pituitary-adrenal function and signs and symptoms of Cushing syndrome.<sup>143</sup> An exhaustive list of side effects is beyond the scope of this guideline, but common side effects of prednisone include insomnia, dizziness, weight gain, increased sweating, gastritis, mood changes, photosensitivity, and hyperglycemia. Severe (but rare) side effects include pancreatitis, bleeding, hypertension, cataracts, myopathy, opportunistic infections, osteoporosis, and osteonecrosis manifesting as fractures and aseptic necrosis of the femoral and humeral heads.<sup>142-146</sup> To minimize the risk of treatment, patients with systemic medical conditions such as insulin-dependent or poorly controlled diabetes, labile hypertension, tuberculosis, peptic ulcer disease, and prior psychiatric reactions to corticosteroids, among others, may not be able to receive systemic corticosteroids.

The lack of clear evidence supporting this treatment, as well as the existence of potential adverse treatment effects, supports a large role for shared decision making with

patients.<sup>147</sup> Most serious side effects, however, occur with chronic use, and adverse events are usually acceptable and manageable for the short 10- to 14-day course of steroids recommended for SSNHL. Alexander et al<sup>148</sup> reviewed the safety of high-dose steroids taken for up to 22 weeks for autoimmune inner ear disease and found that most patients completed the course, with the most frequent adverse events being hyperglycemia and weight gain. There is also evidence that osteonecrosis and fractures occur more commonly in patients with preexisting bone or joint problems in conditions such as systemic lupus erythematosus and rheumatoid arthritis.<sup>149</sup>

### *Intratympanic Corticosteroids*

A more recent method of corticosteroid delivery is the intratympanic (IT) route. The use of IT corticosteroids in patients who do not recover with systemic corticosteroids (“salvage therapy”) will be covered in a different section of this guideline; IT corticosteroids will be reviewed here in the context of initial treatment. Parnes et al<sup>137</sup> published the first animal data and clinical series and demonstrated higher inner ear steroid levels following IT steroid application, with benefit in one-third of patients, and higher percentages of benefit in certain otologic conditions. Subsequent laboratory data have substantiated the claim of higher perilymph steroid concentrations after IT steroid application.<sup>150</sup> Since those publications, a large number of small series without controls, and usually retrospective in nature, have shown inconsistent results for IT steroids. One regimen of initial treatment combining oral and IT steroids for patients with profound hearing loss, in an effort to improve the poor prognosis, had a positive effect in only 3 of 25 patients.<sup>151</sup> However, a combination of a high-dose prednisone taper with IT steroids resulted in partial or complete hearing recovery in 14 of 16 patients.<sup>152</sup> Another study combining oral and IT corticosteroids did not show a difference in hearing recovery compared with corticosteroids alone.<sup>153</sup> A recent study proposed IT treatment as the sole initial treatment.<sup>154</sup> The protocol consisted of early injections for 3 consecutive days, with only 3 of 34 patients failing to improve. A systematic review concluded that IT steroids can be a valuable solution for patients with SSNHL who either cannot tolerate systemic steroid therapy or are refractory to it.<sup>155</sup> For patients with diabetes who cannot take systemic corticosteroids, IT steroids may be an alternative.<sup>156,157</sup>

Intratympanic steroids are usually administered as either dexamethasone or solumedrol.<sup>137</sup> Agents such as histamine and hyaluronic acid have been shown to facilitate transport of the corticosteroid across the round window membrane in laboratory studies.<sup>158,159</sup> Intratympanic corticosteroids appear to affect both immune suppression and ion homeostasis.<sup>160</sup> Corticosteroid concentrations vary widely between studies; most studies on IT corticosteroids refer to dexamethasone 10 to 24 mg/mL and solumedrol 30 mg/mL and higher. Higher concentrations appear to have better outcomes.

The frequency of IT steroid administration also varies widely between studies, from self-administration by the patient across a pressure-equalizing tube (PET) several times

per day to physician administered for several consecutive days to once weekly or less. Moreover, IT corticosteroids have been reported as primary, secondary, or salvage treatment. As such, the myriad studies on IT steroids are difficult to assess, but based on reasonable success in initial reports, more rigorous studies are indicated.<sup>9,160-164</sup> Although with less potential toxicity than systemic corticosteroid treatment, IT corticosteroids can also have adverse effects. These are infrequent but include pain, transient dizziness, infection, persistent tympanic membrane perforation, possible vasovagal or syncopal episode during injection, cost, and multiple office visits.

The only RCT on oral vs IT steroid therapy for SSNHL was conducted at 16 centers and enrolled 250 patients.<sup>13</sup> All patients were enrolled within 14 days of onset of their SSNHL. For primary therapy of SSNHL, promptly administered and equivalently dosed oral and IT steroid appeared to be equally effective, with hearing improvement seen in more than 75% of treated patients. Because the hearing outcomes in these 2 groups of patients were equivalent, the clinician’s judgment about the choice of therapy can and should be based on other considerations, such as risk of side effects and cost. Adverse effects were reported by 88% of the oral group, such as elevated blood sugar, increased thirst, and sleep or appetite changes, and 90% of the IT group, such as transient pain at the injection site and brief caloric vertigo. The adverse effects were the anticipated manageable side effects, most of which were resolved within 1 to 2 weeks, with rare outlying persistent tympanic membrane perforations lasting up to 6 months.

### *Harm vs Benefit of Corticosteroid Therapy*

Despite the uncertain balance of benefit vs harm for steroid therapy based on existing RCTs, there is also insufficient evidence to conclude the treatment is ineffective. Moreover, a large volume of observational studies suggests a treatment benefit, although to what degree this exceeds spontaneous resolution is not known.<sup>9</sup> Considering the devastation of SSNHL and the profound impact on QOL that a hearing improvement may offer, the panel concludes that, therefore, even a small possibility of hearing improvement makes this a reasonable treatment to offer to patients (**Table 9**).

**STATEMENT 9. HYPERBARIC OXYGEN THERAPY: Clinicians may offer hyperbaric oxygen therapy within 3 months of diagnosis of SSNHL. Option based on systematic reviews of randomized control trials with a balance between benefit and harm.**

### *Action Statement Profile for Statement 9*

- Aggregate evidence quality: Grade B, systematic review of RCTs with methodological limitations
- Benefit: Hearing improvement
- Risk, harm, cost: Costs, patient time/effort, patient anxiety and stress, barotraumas, otitis media, oxygen toxicity, worsening of cataracts, fatigue, death
- Benefit-harm assessment: Equilibrium

**Table 10.** Summary of Hyperbaric Oxygen Therapy for Idiopathic Sudden Sensorineural Hearing Loss

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Younger patients respond better to hyperbaric oxygen therapy (HBOT) than older patients (the age cutoffs varied from 50-60 years).<sup>173,235-238</sup>  
 Early HBOT is better than late HBOT (early is defined from 2 weeks to 3 months).<sup>173,177,235,236,238-241</sup>  
 Patients with moderate to severe hearing loss benefit more from HBOT than those with mild hearing loss (moderate hearing loss cutoff was usually at 60 dB).<sup>168,170-172,242-244</sup>  
 Results of studies detailing effectiveness of HBOT depend on the choice of outcome measures.<sup>166</sup>

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- Value judgments: Although hyperbaric oxygen therapy (HBOT) is not widely available in the United States and is not recognized by many US clinicians as an intervention for ISSNHL, the panel felt that the level of evidence for hearing improvement, albeit modest and imprecise, was sufficient to promote greater awareness of HBOT as an intervention for ISSNHL
- Intentional vagueness: None
- Role of patient preferences: Large role for shared decision making
- Exclusions: None
- Policy level: Option

### Supporting Text

The purpose of this statement is to evaluate the role of HBOT (**Table 10**), recognizing that, although this is not a popular therapy in the United States and is not currently approved by the Food and Drug Administration (FDA) for the treatment of ISSNHL, there have been RCTs and a Cochrane review performed on this topic. Vascular compromise and associated cochlear ischemia are thought to be contributory to SSNHL in some cases or could be a final common pathway to hearing loss. Hyperbaric oxygen therapy exposes a patient to 100% oxygen at a pressure level higher than 1 atmosphere absolute (ATA) in a specially designed sealed chamber. This allows for delivery of greatly increased partial pressure of oxygen to the tissues—in this case the cochlea, which is very sensitive to ischemia. Furthermore, HBOT is thought to have complex effects on immunity, oxygen transport, and hemodynamics, reducing hypoxia and edema and potentiating normal host responses to infection and ischemia.<sup>165</sup>

Hyperbaric oxygen therapy has been implemented for the treatment of ISSNHL, typically as an adjunctive treatment. The most recent Cochrane review on this topic reports that HBOT was first used to treat SHL in the late 1960s by French and German workers.<sup>166</sup> Since that time, numerous studies (n = 91) have reported or evaluated the use of HBOT in SHL, but only a small fraction are prospective RCTs.

Two important issues to consider in the evaluation of potential treatments are the outcome measures used to assess benefit and the risk of adverse events. Evaluation of outcome is particularly challenging for ISSNHL, as there is no widely accepted standard,<sup>84</sup> and each method of measuring outcome has limitations.<sup>84</sup>

The Cochrane review included 7 identified RCTs, published between 1985 and 2004.<sup>167-173</sup> The criterion used for

determination of significant benefit was 50% improvement in hearing. Although the chance of a 50% improvement was not significantly increased following HBOT, the chance of a 25% increase was. Data indicated that a physician would need to treat 5 patients with HBOT therapy to improve 1 person's hearing by 25%. Whether this is truly clinically significant is debatable. Although the small total numbers of subjects in this pooled group (n = 392) precluded extensive subgroup analysis, data suggested that improvement may be related to the severity of the hearing loss on presentation. Results were better if HBOT was performed within 2 weeks of acute onset. However, both of these issues should be explored further in future RCTs.

Since this review, the panel found only one other prospective RCT of HBOT for SSNHL.<sup>174</sup> Thirty-six patients were randomized into the study arm that consisted of HBOT plus “standard” medical therapy with prednisolone and compared them with 21 patients who were treated only with prednisolone. Success was defined as hearing regained completely (>50-dB improvement) or moderately (10- to 50-dB improvement). Seventy-nine percent of patients in the HBOT arm had success compared with 71.3% of the control group, a nonsignificant difference. The study offers no evidence that would support the addition of HBOT to a medical regimen for the treatment of SSNHL.

Although risk of serious side effects with HBOT is small, some risks do exist. These include damage to ears, sinuses, and lungs from pressure changes; temporary worsening of short-sightedness; claustrophobia; and oxygen poisoning. Major adverse events were not reported in most of the studies reviewed.

In a population of 782 patients with 11,376 sessions receiving HBOT for a variety of indications, the primary complication of HBOT was difficulty equalizing pressure in the middle ear, which occurred in 17% of patients.<sup>175</sup> Another study found that 45% of patients undergoing HBOT for a variety of indications had eustachian tube dysfunction.<sup>176</sup> Patients undergoing HBOT for SSNHL may have fewer complications as the use of concurrent systemic steroids is common and may decrease the inflammation or edema that may lead to difficulty in pressure equalization or eustachian tube dysfunction. In a study of 80 patients undergoing HBOT for SSNHL, 5 (6.25%) had ear or sinus barotrauma.<sup>177</sup> In addition, patients may suffer from some degree of confinement anxiety while undergoing HBOT.<sup>175,177</sup>

Finally, HBOT is an expensive and time-consuming intervention. Therapy typically involves multiple 1- to 2-hour sessions over days to weeks. Although costs may vary considerably among facilities, queries by the committee showed that typical

fees in academic institutions are approximately \$600 to \$700 per session, including both technical and professional fees. Typical treatments have consisted of 5 to 10 sessions.

Given the small number of patients in the trials reviewed, methodological shortcomings, and poor reporting, the reported findings of benefit should be interpreted cautiously. The substantial cost, the potential adverse effects (including barotrauma), a question of the clinical significance of reported benefits, and the confounding effect of cointerventions (steroids, antivirals, rheologic agents) make it difficult to weigh benefits and harms. The evidence supports possible benefit of HBOT as an adjuvant treatment in cases of acute SSNHL when used within 3 months of the onset of the hearing loss, with potentially more benefit noted in cases of severe to profound loss.

**STATEMENT 10. OTHER PHARMACOLOGIC THERAPY: Clinicians should not routinely prescribe antivirals, thrombolytics, vasodilators, vasoactive substances, or antioxidants to patients with ISSNHL. Recommendation against based on systematic reviews of RCTs with a preponderance of harm over benefit.**

#### Action Statement Profile for Statement 10

- Aggregate evidence quality: Grade B
- Benefit: Avoidance of unnecessary treatment, avoid adverse events of unnecessary treatment, cost saving
- Risk, harm, cost: None as the recommendation is against the use of these therapies
- Benefit-harm assessment: Preponderance of benefit
- Value judgments: None
- Intentional vagueness: The word *routine* is used to avoid setting a legal standard, recognizing that patient-specific indications for 1 or more of these therapies may be reasonable to try on an individualized basis, with shared decision making
- Role of patient preferences: None
- Exclusions: None
- Policy level: Recommendation against

#### Supporting Text

The purpose of this statement is to discourage clinicians from using pharmacologic agents that have potential side effects and no documented efficacy, despite the fact that many of these agents are advocated.

One of the proposed etiologies of SSNHL is inflammation caused by a viral infection. Proposed mechanisms of action include direct viral invasion of the cochlea or cochlear nerve, reactivation of a latent virus within the spiral ganglion, and immune-mediated mechanisms once an infection becomes systemic.<sup>178</sup> Theoretically, initiation of antiviral agents may be valuable for aiding in the recovery of hearing. Because direct sampling of inner ear fluids is impractical and potentially harmful to the patient, hematologic serologic testing is the only avenue for viral testing.

Multiple trials have been carried out and failed to find any benefit of the addition of antiviral therapies. Conlin and Parnes

performed both a systematic review<sup>7</sup> and meta-analysis<sup>6</sup> of treatments for SSNHL and found only 4 RCTs<sup>179-182</sup> comparing antiviral therapy and steroid therapy vs placebo and steroid therapy. None of the studies reported statistically significant results. In addition, antiviral agent use is not without consequences, and reported side effects include nausea, vomiting, photosensitivity, and, rarely, reversible neurologic reactions, including mental status changes, dizziness, and seizures.

Another proposed etiology of SSNHL is cochlear ischemia. Because the blood supply to the inner ear has no collateral circulation, it is tenuous at best. As with most vascular disorders, hemorrhage, embolism, and vasospasm may affect the inner ear negatively and cause damage, resulting in SSNHL. Fisch et al<sup>183</sup> demonstrated a 30% reduction of perilymphatic oxygen tension in patients with SSNHL and demonstrated that treatment with carbogen resulted in a mean increase in perilymph oxygen tension of 175%. Hypercoagulability has also been seen in blood samples of patients with SSNHL. There is contradictory histopathological and clinical evidence against the vascular theory of SSNHL.<sup>12,184-186</sup> Most patients with SSNHL probably do not have a solely ischemic etiology, which is difficult to disprove based on clinical features and testing.

Vasoactive agents have been tried to enhance cochlear blood flow. Prostaglandin E<sub>1</sub> has shown benefit as a vasodilator and an inhibitor of platelet aggregation. Naftidrofuryl acts to dilate vessels by antagonizing the effect of serotonin and thromboxane A<sub>2</sub>. Calcium antagonists act to dilate vessels by antagonizing contraction of the smooth muscle cells in the vessel walls. *Ginkgo biloba* extract contains flavones and terpenes, which prevent the development of free radicals in cases of ischemic-related metabolic disturbances and thus counteract secondary vessel contraction. Antihypoxidotic and antiedematous effects, as well as platelet-activating factor (PAF)-antagonistic properties, have been described. Pentoxifylline increases the flexibility of erythrocytes and leukocytes and thus improves blood viscosity, particularly in the capillaries. In addition, pentoxifylline also inhibits platelet aggregation by means of prostaglandin synergy. Dextran may improve microcirculation due to an antithrombotic effect. Hydroxyethyl starch carrier solution reduces the hematocrit level and platelet aggregation.<sup>187,188</sup>

These therapies have considerable side effects. Different types of treatment entail different risks. The clinician should be aware of these potential adverse drug events, including allergic reactions, bleeding, hypotension, arrhythmias, seizures, circulatory collapse, and drug interactions.

The use of vasodilators and vasoactive substances for ISSNHL was reviewed by the Cochrane Collaborative in 2009.<sup>189</sup> Only 3 RCT studies were worthy of evaluation. All 3 of these were considered to have a high risk of bias because their overall methodology was poor and sample sizes were small. The reviewers noted differences in the type, dosage, and duration of vasodilator treatment used in each of these studies. Because of the degree of heterogeneity in methodology and outcomes assessment, the results could not be

combined to reach a conclusion of efficacy. Another review found no clinically significant benefit of rheologic agents over placebo.<sup>129</sup>

Another proposed therapy for SSNHL is defibrinogenation therapy, which leads to a decrease in the peripheral blood viscosity and thereby to augmentation of the blood circulation. Multiple poorly controlled studies have failed to show clinical improvement of this form of therapy.<sup>190</sup>

Finally, diatrizoate meglumine (Hypaque) is an intravenous contrast agent that has been postulated to improve hearing in patients with SSNHL. An analysis of Hypaque vs steroids vs vasodilator showed no better results with any of those treatments than the published spontaneous recovery rate of 65%.<sup>191</sup> Fatal reactions to intravenous contrast agents have been reported to be as high as 1 per 10,000.<sup>192</sup>

In summary, no data support the use of antiviral agents. Interventions that improve cochlear blood flow through a variety of mechanisms to treat SSNHL have limited evidence supporting their use and high risk of adverse events. Also, no data support the use of thrombolytics, vasodilators, vasoactive substances, or antioxidants in the treatment of SSNHL.

In addition to the therapies discussed above, a host of other therapies have been used to treat SSNHL (ie, vitamins, minerals, interferon, nitroglycerine, and other complementary and alternative medications). The evidence base for these therapies was insufficient to review in this guideline, and no comment is made on their use.

**STATEMENT 11. SALVAGE THERAPY: Clinicians should offer IT steroid perfusion when patients have incomplete recovery from ISSNHL after failure of initial management.** *Recommendation based on RCTs with a preponderance of benefit over harm.*

#### Action Statement Profile for Statement 11

- Aggregate evidence quality: Grade B, RCTs with limitations
- Benefit: Hearing recovery
- Risk, harm, cost: Perforation, discomfort, cost, patient anxiety
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: Patients qualifying for salvage therapy have failed to respond to initial management or have had an incomplete response. Failure of initial management is not clearly defined as there is limited guidance from the literature as to what level of residual hearing loss qualifies a patient for salvage. The guideline panel recognized that varying degrees of hearing loss will affect patients differently. This may govern the aggressiveness of the decision to pursue further therapy.
- Role of patient preferences: Significant role for shared decision making regarding treatment options

depending on various perceived levels of hearing impairment

- Exclusions: None
- Policy level: Recommendation

#### Supporting Text

The purpose of this statement is to discuss the role of IT steroids as salvage therapy for patients with incomplete hearing recovery (**Table 11**). This is in contrast to the previous statement (Statement 8) regarding steroid therapy, which dealt only with IT steroids in the context of initial management.

For patients who fail to recover spontaneously or after initial management, including corticosteroid treatment and/or observation, IT delivery of steroids has been proposed by a number of authors as an option to obtain additional hearing recovery.<sup>9,137,150,151,193-196</sup> There is now a significant body of research investigating the use of IT steroid therapy in this setting, consisting of numerous case series and 4 RCTs. Although most of these studies suffer from considerable design flaws, the majority do show improved hearing outcomes after IT steroid therapy.<sup>155</sup>

Similar to the concept of parenteral steroids for ISSNHL, IT steroid therapy aims to reduce inflammation in the inner ear that may be contributing to or preventing recovery from hearing loss. There is experimental evidence from animal models indicating that a considerably higher concentration of steroid can be delivered to the inner ear when the medication is delivered through a transtympanic route compared with systemic administration.<sup>137,150</sup>

The steroids are delivered to the middle ear and then absorbed and diffused through the round window membrane into the inner ear. Intratympanic steroids may be delivered via a needle through the tympanic membrane or may be placed into the middle ear through a tympanostomy tube or a myringotomy (incision in the eardrum). Steroids may also be delivered to the round window via a microcatheter, a MicroWick,<sup>193</sup> hydrogel applications, and nanoparticles. Transtympanic needle or tympanostomy tubes are the most frequently used.<sup>128</sup>

The IT delivery route has the additional benefit of avoiding the considerable side effects of further systemic steroid therapy. Intratympanic steroids very rarely cause changes in serum glucose levels in patients with diabetes.<sup>156</sup> They may also be given to patients with cataracts, myasthenia gravis, and glaucoma.<sup>138</sup> The principal risk appears to be a persistent tympanic membrane perforation at the injection site. This complication, however, is rare and frequently resolves spontaneously or with a paper patch myringoplasty in the office.

Existing studies showed considerable variability in the dose and concentration of steroids administered; the timing, frequency, and total number of injections (ranging from one to several to continuous); and drug selection (dexamethasone and methylprednisolone).<sup>155</sup> This high degree of variability makes it difficult to compare results across studies.

Despite this variability, 3 of the 4 RCTs evaluating intratympanic steroids as salvage therapy found that IT steroids

**Table 11.** Summary of Intratympanic Steroid as Salvage Therapy

Study/No.	Therapy Begins	Dose/Method of Injection	Monitoring of Patient	% Improvement
Ahn et al (2008), <sup>194</sup> 49 patients	<2 weeks after oral failure 2 weeks to 1 month 1 to 2 months	0.3-0.4 mL of 5 mg/mL Dexamethasone 2 times per week for 2 weeks	Final hearing evaluation was performed 3 months after the outbreak of SSNHL	7 of 16 (43.8%) early ITD 6 of 20 (30.0%) mid-ITD 2 of 13 (15.4%) late ITD
Ho et al (2004), <sup>195</sup> 22 patients	Within 2 weeks after methylprednisolone	1 mg/mL dexamethasone once per week for 3 weeks	PTA Successful treatment defined as hearing improvement of >30 dB HL	8 of 15 (53.3%)
Slattery et al (2005), <sup>134</sup> 20 patients	Up to 3 months after oral steroids	62.5 mg/mL methylprednisolone, 4 injections over a 2-week period	PTA >10 dB HL WRS 12%	55%
Choung et al (2006), <sup>245</sup> 33 patients	<28 days after oral steroids	5 mg/mL dexamethasone, 2 injections per week for 2 weeks	10 dB HL PTA WRS 15%	38.2%
Dallan et al (2006), <sup>246</sup> 8 patients	Unknown	40 mg/mL methylprednisolone, single injection	PTA >15 dB HL	75%
Xenellis et al (2006), <sup>196</sup> 19 patients	<2 weeks after IV prednisolone	0.5 mL of 40 mg/mL, 4 injections over 2 weeks	PTA >10 dB HL performed after injections and at 1.5 months	47%
Haynes et al (2007), <sup>9</sup> 40 patients	<40 days	24 mg/mL dexamethasone, single injection	PTA >20 dB HL WRS 20%	26.7%
Roebuck and Chang (2006), <sup>247</sup> 31 patients	After 5 to 7 days of oral steroids	24 mg/mL dexamethasone, single injection	PTA >10 dB HL WRS > 15%	33% 38.7%
Plaza et al (2007), <sup>248</sup> 9 patients	<5 days after IV methylprednisolone	20 mg/mL methylprednisolone, 3 injections over 1 week	PTA >15 dB HL WRS >15% performed after injections and at 1.5 months	55%
Kilic et al (2007), <sup>201</sup> 19 patients	After a 3-week course of high-dose systemic corticosteroid where hearing gain was PTA <10 dB	0.5 mL of 62.5 mg/mL, 5 injections over 12 days	PTA >10 dB HL Performed at 1 and 3 months	73.6%
Gouveris et al (2005), <sup>249</sup> 21 patients	<2 weeks after oral steroids	0.3-0.4 mL of 8 mg/mL dexamethasone every 2 days	PTA >10 dB HL	33%
Silverstein (1996), <sup>250</sup> 8 patients	<30 days	Dexamethasone via microwick, 3 times per week for 3 to 4 weeks	PTA >10 dB HL	25%
Kopke (2001), <sup>251</sup> 6 patients	<6 weeks	62.5 mg/mL methylprednisolone, catheter for 14 days	PTA >10 dB HL	83%

Abbreviations: HL, hearing level; ITD, intratympanic dexamethasone; IV, intravenous; PTA, pure-tone average; SSNHL, sudden sensorineural hearing loss; WRS, word recognition scores.

improved hearing outcomes beyond placebo. Hearing improvement occurred in 53% to 90% of patients.<sup>195,196</sup> The other RCT compared a continuous infusion of steroids to the middle ear for 14 days with an infusion of saline.<sup>197</sup> This study, with only 23 patients, was underpowered to detect meaningful differences, and all patients in the control group received IT steroids after 1 week, making it impossible to differentiate longer term differences. This study failed to find a statistically significant difference in improvement in PTA between the treatment and placebo groups (mean [SD], 13.9 [21.3] dB HL vs 5.4 [10.4] dB HL, respectively;  $P = .07$ ), although word

recognition trended toward better in the treatment group (24.4% improvement in one group vs 4.5% in the other group;  $P = .07$ ).

The majority of non-RCTs and noncontrolled trials of IT steroids as salvage therapy reported a hearing improvement in the treatment group ranging from 8% to 100%.<sup>9,153,155,157,198-202</sup> One critical problem in the individual trials is how an improvement in hearing was defined. Most authors used a 10-dB HL improvement in the PTA or an improvement in WRS of 15% or 20% as indicative of successful treatment. Others used a 30-dB HL PTA improvement or 50% recovery as the

definition of successful therapy. Depending on the degree of hearing loss, these thresholds may or may not indicate an improvement to useable hearing. Hence, these outcomes, although statistically significant, may not be of clinical significance.

Despite the limitations of the existing research, the majority of studies evaluating IT steroids as salvage therapy for ISSNHL, including both nonrandomized and RCTs, demonstrated a consistent benefit of some degree of additional hearing recovery beyond that afforded by initial therapy. Because salvage IT steroid therapy has been found to be beneficial for some degree of hearing recovery, treatment may be applicable for those who have persistent hearing loss despite conventional treatment with oral or intravenous steroids. The decision to perform this treatment should be based on whether a significant degree of hearing loss still exists and patient preference. The decision to treat is often subjective but should take into consideration the risks and benefits of the treatment being considered.

**STATEMENT 12. OUTCOMES ASSESSMENT: Clinicians should obtain follow-up audiometric evaluation within 6 months of diagnosis for patients with ISSNHL.**

*Recommendation based on observational studies with a preponderance of benefit over harm.*

**Action Statement Profile Statement 12**

- Aggregate evidence quality: Grade C, based on observation studies
- Benefit: Assess outcome of intervention, identify patients who may benefit from audiologic rehabilitation, identify cause of hearing loss, identify progressive hearing loss, improve counseling
- Risk, harm, cost: Procedural cost
- Benefit-harm assessment: Preponderance of benefit
- Value judgments: The patient perception of hearing recovery is not always completely accurate, and patients may be unaware of a residual hearing impairment that could be identified through audiometric assessment. Patients who report subjective hearing improvement may still derive additional benefits from objective testing
- Intentional vagueness: None
- Role of patient preferences: Some
- Exclusions: None
- Policy level: Recommendation

**Supporting Text**

The purpose of this statement is to highlight the importance of audiometric follow-up in patients with ISSNHL to assess for other etiologies in patients with progressive hearing loss and to identify patients who might benefit from rehabilitation options. If treatment is initiated, then earlier audiometric follow-up may be indicated to assess the benefit of the intervention and guide decision making regarding salvage therapy if incomplete recovery occurs.

In patients having an episode of ISSNHL, it is important to obtain a follow-up audiometric evaluation to determine if therapy was successful in improving hearing. Long-term follow-up was reported in 156 patients diagnosed with ISSNHL.<sup>203</sup> Of those patients who showed recovery, 54.5% showed recovery within a 10-day course of combined therapy. Although the majority of patients did not improve completely, final hearing levels were reached by 1 month in 78% of patients and by 3 months in 97% of patients. Of all patients, only 1 (0.6%) showed any recovery beyond 6 months. Beyond this, there are no data to guide the timing of follow-up. With the constraint of 3 months on the short end, the chosen time of 6 months used in the statement was based on expert opinion. Slightly shorter or longer durations of follow-up would not be unreasonable.

If the hearing loss is permanent, the disability may require auditory rehabilitation. In a patient with residual hearing loss, a discussion should be undertaken of the benefits of hearing aids or assistive listening devices to manage the hearing loss. There is benefit to initiating these discussions when a hearing loss is first discovered, as temporary measures for hearing assistance may be beneficial and awareness of long-term rehabilitative options may alleviate some anxiety.

**Follow-up Audiometric Measures to Assess the Effectiveness of Treatment(s) for ISSNHL**

Clinicians agree that the most accurate and cost-efficient method to monitor the effectiveness of medical intervention(s) to treat SSNHL is to compare pure-tone thresholds, PTA, SRT, and/or WRS at follow-up audiometric evaluations with the initial audiometric evaluation.

**Outcome Measures Used to Assess Effectiveness of Treatment of ISSNHL**

A meta-analysis reviewed 20 studies using placebos, steroids, antiviral agents, other active therapies, and IT dexamethasone injections to treat ISSNHL.<sup>6</sup> Although the treatments were quite diverse, the common thread was that all the studies used pure-tone thresholds, PTA, and/or WRS to monitor the effectiveness of treatment leading to recovery of hearing.

There have been many definitions of *recovery* to define improvement in hearing attributable to treatment. One of the landmark early studies on ISSNHL defined recovery as follows<sup>76</sup>:

- a. **Complete:** if the follow-up PTA (dB HL) or SRT (dB HL) improved to within 10 dB of pre-sudden hearing loss hearing levels
- b. **Partial:** if the follow-up PTA (dB HL) or SRT (dB HL) improved to within 50% of pre-sudden hearing loss hearing levels
- c. **No recovery:** if the follow-up PTA (dB HL) or SRT (dB HL) was less than 50% of recovery of pre-sudden hearing loss hearing levels

Other definitions of recovery were outlined nearly 30 years later, after reviewing 25 studies to provide clinicians with definitions provided by investigators using IT steroid injections.<sup>161</sup> The “recovery” ranged from:

- a. **10- to 30-dB HL improvement in PTA (dB HL) from pretreatment hearing levels:** no measure of change in WRS was provided. Using a 10-dB HL change in PTA is worrisome because this magnitude of change in PTA (dB HL) is within the test-retest reliability of measuring pure-tone thresholds.
- b. **10- to 30-dB HL improvement in PTA (dB HL) and 10% to 20% improvement in WRS:** Using a *fixed* 10% to 20% criterion is worrisome, as will be shown in the next section.
- c. **Calculate individual PTA (dB HL) recovery** and determine how this improvement falls into **complete, partial, and no recovery** categories.<sup>76</sup> These include the following:
  1. **Complete:** PTA (dB HL) within 10 dB HL of initial HL or within 10 dB HL of the HL of the unaffected ear
  2. **Partial:** PTA (dB HL) within 50% of initial HL or >10-dB HL improvement of the HL
  3. **No recovery:** <10-dB HL improvement in HL relative to the initial HL
- d. **Improvement to 50% of baseline** difference between the treated and untreated ear
- e. **Improvement in WRS (%) and decrease in PTA (dB HL)**
- f. **Hearing is within normal limits** (–10 to 15 dB HL) **and hearing is serviceable**

When comparing *follow-up* HL with *initial* HL, it is important that any change must exceed 10 dB HL to be considered significant.<sup>81,82</sup>

When determining significant changes in WRS, the clinician should consult the binomial distribution table (**Table 12**) to compare the *posttreatment* WRS relative to the *initial* WRS. For example, if a WRS of 20% is measured initially for a 50-word list, the follow-up WRS must exceed 36% to be considered significant improvement and be less than 8% to be considered a significant reduction ( $P > .05$ ). As an alternative, the clinician should consult the test material manual to determine if differences between initial and follow-up WRS exceed the 95% confidence interval or other statistical approaches.

Finally, the clinician should document the patient's comments concerning changes in hearing, tinnitus, sensation of fullness, vertigo, or nausea following treatment.

### Recommendations for Outcomes Assessment in Future Studies

**Current limitations.** All of the above strategies suffer from 2 main limitations. First, although an absolute improvement in pure-tone thresholds or WRS may be statistically significant, they may not be clinically significant. Second, in most patients, the pre-SSNHL hearing levels in the affected ear are not known.

**Recommendations.** To address these issues, this guideline panel proposes the following measures for future outcomes assessment (note: in the absence of guidance from the literature, clinical expert opinion was also used in making these

recommendations): (1) unless a preevent asymmetry of hearing was known or suspected, the unaffected ear should be used as the standard against which recovery should be compared; (2) a complete recovery requires return to within 10 dB HL of the unaffected ear *and* recovery of word recognition scores to within 5% to 10% of the unaffected ear; (3) partial recovery should be defined in 2 ways based on whether or not the degree of initial hearing loss after the event of SSNHL rendered the ear nonserviceable (based on the AAO-HNSF definition); and (4) anything less than a 10-dB HL improvement should be classified as no recovery.

**Partial recovery.** For ears that were rendered nonserviceable by the episode of SSNHL, return to serviceable hearing should be considered a significant improvement, and whether or not this level of recovery occurs should be recorded. Recovery to a serviceable level typically indicates that after recovery, the ear would be a candidate for traditional hearing amplification. Recovery to less than serviceable levels indicates an ear that would in most circumstances not benefit from traditional amplification. For ears with SSNHL to hearing levels that are still in the serviceable range, a 10-dB HL improvement in pure-tone thresholds (the smallest recordable improvement outside of the range of error for most audiograms) or an improvement in WRS of  $\geq 10\%$  (approximate lower limit for a statistically significant change based on binomial tables for WRS of  $>50\%$  at baseline) should be considered partial recovery and recorded.

This guideline panel recognizes that these criteria still have limitations in that the impact of an absolute 10-dB HL improvement in pure-tone sound detection or of an absolute 10% improvement in WRS may have different benefits for different patients. Nonetheless, this standard better captures whether or not a meaningful change has occurred with or without treatment.

**STATEMENT 13. REHABILITATION: Clinicians should counsel patients with incomplete recovery of hearing about the possible benefits of amplification and hearing-assistive technology (HAT) and other supportive measures.** *Strong recommendation based on systematic reviews and observational studies with a preponderance of benefit over harm.*

### Action Statement Profile for Statement 13

- Aggregate evidence quality: Grade B, based on systematic reviews and observational studies
- Benefit: Improved quality of life, improved functionality, emotional support, improved hearing
- Risk, harm, cost: Time and cost of counseling
- Benefit-harm assessment: Preponderance of benefit
- Value judgments: None
- Intentional vagueness: None
- Role of patient preferences: Patient may decline counseling
- Exclusions: None
- Policy level: Strong recommendation

### Supporting Text

The purpose of this statement is to increase awareness that counseling and education for patients on the options

**Table 12.** Binomial Distribution Table<sup>85</sup>

% Score	n = 50	n = 25	n = 10	% Score	n = 100 <sup>a</sup>
0	0-4	0-8	0-20	50	37-63
2	0-10			51	38-64
4	0-14	0-20		52	39-65
6	2-18			53	40-66
8	2-22	0-28		54	41-67
10	2-24		0-50	55	42-68
12	4-26	4-32		56	43-69
14	4-30			57	44-70
16	6-32	4-40		58	45-71
18	6-34			59	46-72
20	8-36	4-44	0-60	60	47-73
22	8-40			61	48-74
24	10-42	8-48		62	49-74
26	12-44			63	50-75
28	14-46	8-52		64	51-76
30	14-48		10-70	65	52-77
32	16-50	12-56		66	53-78
34	18-52			67	54-79
36	20-54	16-60		68	55-80
38	22-56			69	56-81
40	22-58	16-64	10-80	70	57-81
42	24-60			71	58-82
44	26-62	20-68		72	59-83
46	28-64			73	60-84
48	30-66	24-72		74	61-85
50	32-68		10-90	75	63-86
52	34-70	28-76		76	64-86
54	36-72			77	65-87
56	38-74	32-80		78	66-88
58	40-76			79	67-89
60	42-78	36-84	20-90	80	68-89
62	44-78			81	69-90
64	46-80	40-84		82	71-91
66	48-82			83	72-92
68	50-84	44-88		84	73-92
70	52-86		30-90	85	74-93
72	54-86	48-92		86	75-94
74	56-88			87	77-94
76	58-90	52-92		88	78-95
78	60-92			89	79-96
80	64-92	56-96	40-100	90	81-96
82	66-94			91	82-97
84	68-94	60-96		92	83-98
86	70-96			93	85-98
88	74-96	68-96		94	86-99
90	76-98		50-100	95	88-99
92	78-98	72-100		96	89-99
94	82-98			97	91-100
96	86-100	80-100		98	92-100
98	90-100			99	94-100
100	96-100	92-100	80-100	100	97-100

<sup>a</sup>If score is less than 50%, find Score = 100 – observed score and subtract each critical difference limit from 100.

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**Table 13.** Common Issues Raised by Individuals with Sudden Sensorineural Hearing Loss

Counseling Topic	Type of Counseling	Suggestions
Is there anything I can do to restore my hearing?	Informational counseling	Discuss various treatment options and possible outcomes
What are the risks of treatment?	Informational counseling	Benefits and risks of treatment options
Will I lose hearing in my other ear?	Personal adjustment counseling	Address the emotional components of hearing loss
Is there anything I can do to restore my hearing?	Personal adjustment/informational counseling	Introduce amplification and rehabilitation options
How will I be able to manage with hearing in just one ear?	Personal adjustment counseling	Discuss support groups such as Hearing Loss Association of American (HLAA)
Do I have to wear a hearing aid?	Personal adjustment/informational counseling	Discuss types of hearing aids and contralateral routing of signal (CROS and BiCROS) option if appropriate
Is there any surgery I can have to get my hearing back?	Personal adjustment/informational counseling	Discuss surgical options if a candidate

available to manage their existing hearing loss are beneficial. Counseling is a critical component of all aspects of patient care. Although this action statement emphasizes its importance for patients with incomplete recovery, it should be noted that counseling is an integral focus throughout the assessment and treatment process for SSNHL and should be done by a specialist.

The presence of hearing loss during the course of the illness commands immediate attention. Waiting until it is determined if medical treatments have been successful, either completely or partially, or if no recovery is achieved at all does not adequately address the common concerns many patients and their communication partners experience. Patients fear loss of hearing in their better ear, how long they will have to live with the hearing loss, and if they will need to wear a hearing aid. Although these questions cannot be answered during the initial treatment period, a continuous dialogue, sharing of information, and listening will assist the patient's adjustment to the changes that have occurred and, in some cases, may be permanent.<sup>204</sup> Carlsson et al<sup>21</sup> recommend that these patients require extensive multidisciplinary rehabilitation to contend with the multifaceted problems associated with SSNHL. **Table 13** highlights some issues that may need to be addressed when counseling your patient through the process of managing SSNHL.

Although the vast majority of hearing loss associated with SSNHL is unilateral, this does not diminish the handicapping effect it may have on an individual's functioning and QOL. A retrospective study of adults with unilateral SSNHL found that 86% (n = 21) reported hearing handicap as determined through the use of the Hearing Handicap Inventory for Adults (HHIA).<sup>18,205</sup> For those who reported the presence of tinnitus, 56% demonstrated handicap as measured by the Tinnitus Handicap Inventory.<sup>206</sup>

Self-assessment measurement tools, such as the Hearing Handicap Inventory for the Elderly (HHIE)<sup>207</sup> and the modified version for use with adults, the HHIA, have long been available to assist in determining the impact of hearing loss

on QOL. These tools have frequently been used as outcome measures to determine success with amplification. The management of the patient with SSNHL may require addressing the need for hearing aid(s) or HAT systems either as a means of bridging the period of time that hearing is impaired during treatment or as an option if recovery is not possible. A systematic review of health-related QOL and hearing aids determined that amplification improves the QOL for individuals with SSNHL by aiding in a major reduction of psychosocial and emotional manifestations.<sup>208</sup>

There are a variety of amplification options available for the management of unilateral impairment. Traditional recommendations are the CROS (contralateral routing of signal)—style hearing aids that require the use of a microphone placed on the ear with hearing impairment that transmits the auditory signal to the better ear. CROS instruments have previously been large and cosmetically unappealing. Recent digital developments, however, have resulted in smaller behind-the-ear or custom instruments. For individuals who may have a preexisting hearing loss in the better hearing ear, bilateral contralateral routing of signals (BiCROS) hearing aids are recommended that will allow both CROS and hearing aid characteristics as necessary. Monaural hearing aid options may also be recommended for those who can benefit from amplification in the poorer ear without the need for crossover. More and more options are being used and investigated for amelioration of single-sided deafness (SSD). Osseointegrated bone conductive devices use bone conduction as a means of transferring sound from the affected side to the better hearing cochlea. Although the device is a surgical option, head band placement is available for those individuals who may not be surgical candidates. Deep intracanal devices and dental bridges accompanied by ear-level devices also employ bone conduction sound transmission for the treatment of single-sided deafness. There is ongoing clinical research on the utility of cochlear implantation in SSD. In the laboratory, inner ear hair cell regeneration remains a major goal in ear research.

In addition to hearing aids or, as an alternative for some, HAT systems can provide the patient with SSNHL a means of improving communication in specific listening conditions and can be very useful during the initial stages of medical treatment. Hearing-assistive technologies typically require the use of headphones and a handheld or lapel-worn microphone. Sound is transmitted from the source directly to the listener either through hardwire or wireless technologies such as infrared and frequency modulated (FM). Other considerations for assistive technology include auditory, visual, and tactile alerting systems. For additional information regarding the rehabilitative options for adults with hearing loss, the reader is referred to the evidenced-based guidelines of the American Academy of Audiology.<sup>209</sup>

Coping with the issues resulting from the sudden and at times permanent loss of hearing may require more than professional intervention. Consumer-based organizations may be a valuable resource for support and information. The Hearing Loss Association of America (HLAA) is the largest, but by no means the only, consumer-driven organization for adults with hearing loss. Many patients rely on the information they receive from these types of organizations as they develop their mechanisms for coping with hearing loss.

Some patients, depending on the handicapping effects of the hearing loss and their perceived communication deficits, may require therapeutic interventions such as counseling, speech reading, and auditory training. A systematic review of the effectiveness of counseling-based group aural rehabilitation for patients with SSNHL found reasonably good evidence for the reduction of self-perceived hearing handicap.<sup>210</sup> Availability of these rehabilitation services either for a group or an individual, however, may be difficult to locate or find locally. In such cases, patients can be directed to a variety of computer-based interactive treatment programs. Further information regarding online/DVD self-study programs can be obtained by contacting the following organizations: Academy of Rehabilitative Audiology ([www.audrehab.org](http://www.audrehab.org)), American Academy of Audiology ([www.audiology.org](http://www.audiology.org)), American Speech-Language and Hearing Association ([www.asha.org](http://www.asha.org)), and the American Academy of Otolaryngology–Head and Neck Surgery ([www.entnet.org](http://www.entnet.org)).

Counseling and rehabilitative services are necessary to allow the patient with SSNHL to cope with the loss of hearing and manage independently to the best of his or her ability. Combining many of the items contained in this action statement may help address these very significant communication needs.

## Implementation Considerations

The complete guideline is published as a supplement to *Otolaryngology–Head and Neck Surgery*, which will facilitate reference and distribution. A full-text version of the guideline will also be accessible free of charge for a limited time at <http://www.entnet.org>. The guideline will be presented to AAO-HNSF members as a miniseminar at the AAO-HNSF annual meeting and OTO EXPO. Existing brochures and publications by the AAO-HNSF will be updated to reflect the guideline recommendations.

To distinguish SNHL from CHL, the guideline panel recommends a combination of history, physical examination, tuning fork tests, and audiometry. To aid the clinician's implementation of this recommendation, a description of both the Weber and Rinne tests has been provided.

As a supplement to clinicians, the panel created a checklist of features associated with specific disorders underlying hearing loss. This checklist can be incorporated into future educational materials developed by the AAO-HNSF.

The panel believes that patient education and shared decision making are an important component in the successful management of patients with ISSNHL. As such, it is important for both clinicians and patients to be aware of the possible etiology of their hearing loss, available treatments and their associated benefits and risks, and rehabilitation services. A basic protocol has been developed for the management of patients with ISSNHL along with a list of discussion points. The panel believes these resources can be incorporated into a patient leaflet that can be made available through the AAO-HNSF.

To assist clinicians in determining an appropriate course of treatment, summary tables have been provided for corticosteroid therapy, hyperbaric oxygen therapy, and IT steroids as salvage therapy. As a reference aid, these summary tables, as part of the shared decision-making process, will help guide the clinician's management of ISSNHL.

To aid patients in managing their SSNHL, **Table 13** (Counseling Issues Raised by Patients with Sudden Sensorineural Hearing Loss) will be adapted into a patient leaflet. The AAO-HNSF will seek the assistance of the consumer groups represented on the guideline panel when developing this tool.

## Research Needs

This guideline was developed based on the current body of evidence regarding the diagnosis, treatment, and ongoing management of patients with SHL. As determined by the guideline panel's review of the literature, assessment of current clinical practices, and determination of evidence gaps, research needs were determined as follows:

1. Determine a standardized and evidence-based definition of SSNHL.
2. Investigate the effectiveness of corticosteroid treatment vs a placebo. The panel believes that such a clinical trial should be conducted due to the equipoise of existing data.
3. Further investigate the benefit of HBOT. Current evidence regarding this treatment option looks promising; however, there is a bias among US physicians and payers not to offer this therapy. Standardized treatment protocols are needed for HBOT for ISSNHL.
4. Develop standardized outcome criteria to aid the comparison of clinical studies.
5. Further study the use of IT steroids as salvage therapy, particularly the optimal drugs, dosage,

concentrations, and administration schedules for IT therapy.

6. Develop criteria to determine at what level of hearing recovery IT steroids would be offered as salvage.
7. Determine the percentage of patients who gain serviceable hearing as a result of treatment.
8. Investigate the use of “combined therapy” (ie, oral and IT steroids) in patients with ISSNHL.
9. Develop long-term follow-up protocols for patients with ISSNHL.
10. Evaluate therapies using standardized definitions and treatment protocols across studies.

## Disclaimer

This clinical practice guideline is not intended as the sole source of guidance in managing patients with SHL. Rather, it is designed to assist clinicians by providing an evidence-based framework for decision-making strategies. The guideline is not intended to replace clinical judgment or establish a protocol for all individuals with this condition and may not provide the only appropriate approach to managing this problem.

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