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Clinical Practice Guideline: Sudden Hearing Loss (Update)

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Abstract

Objective. Sudden hearing loss is a frightening symptom that often prompts an urgent or emergent visit to a health care provider. It is frequently but not universally accompanied by tinnitus and/or vertigo. Sudden sensorineural hearing loss affects 5 to 27 per 100,000 people annually, with about 66,000 new cases per year in the United States. This guideline update provides evidence-based recommendations for the diagnosis, management, and follow-up of patients who present with sudden hearing loss. It focuses on sudden sensorineural hearing loss in adult patients aged $\geq$18 years and primarily on those with idiopathic sudden sensorineural hearing loss. Prompt recognition and management of sudden sensorineural hearing loss may improve hearing recovery and patient quality of life. The guideline update is intended for all clinicians who diagnose or manage adult patients who present with sudden hearing loss.

Purpose. The purpose of this guideline update is to provide clinicians with evidence-based recommendations in evaluating patients with sudden hearing loss and sudden sensorineural hearing loss, with particular emphasis on managing idiopathic sudden sensorineural hearing loss. The guideline update group recognized that patients enter the health care system with sudden hearing loss as a nonspecific primary complaint. Therefore, the initial recommendations of this guideline update address distinguishing sensorineural hearing loss from conductive hearing loss at the time of presentation with hearing loss. They also clarify the need to identify rare, nonidiopathic sudden sensorineural hearing loss to help separate those patients from those with idiopathic sudden sensorineural hearing loss, who are the target population for the therapeutic interventions that make up the bulk of the guideline update. By focusing on opportunities for quality improvement, this 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.

Methods. Consistent with the American Academy of Otolaryngology–Head and Neck Surgery Foundation's "Clinical Practice Guideline Development Manual, Third Edition" (Rosenfeld et al. *Otolaryngol Head Neck Surg.* 2013;148[1]:S1-S55), the guideline update group was convened with representation from the disciplines of otolaryngology–head and neck surgery, otology, neurotology, family medicine, audiology, emergency medicine, neurology, radiology, advanced practice nursing, and consumer advocacy. A systematic review of the literature was performed, and the prior clinical practice guideline on sudden hearing loss was reviewed in detail. Key Action Statements (KASs) were updated with new literature, and evidence profiles were brought up to the current standard. Research needs identified in the original clinical practice guideline and data addressing them were reviewed. Current research needs were identified and delineated.

Results. The guideline update group made *strong recommendations for* the following: (KAS 1) Clinicians should distinguish sensorineural hearing loss from conductive hearing loss when a patient first presents with sudden hearing loss. (KAS 7) Clinicians should educate patients with sudden sensorineural hearing loss about the natural history of the condition, the benefits and risks of medical interventions, and the limitations of existing evidence regarding efficacy. (KAS 13) Clinicians should counsel patients with sudden sensorineural hearing loss who have residual hearing loss and/or tinnitus about the possible benefits of audiologic rehabilitation and other supportive measures. These strong

recommendations were modified from the initial clinical practice guideline for clarity and timing of intervention.

The guideline update group made *strong recommendations against the* following: (KAS 3) Clinicians should not order routine computed tomography of the head in the initial evaluation of a patient with presumptive sudden sensorineural hearing loss. (KAS 5) Clinicians should not obtain routine laboratory tests in patients with sudden sensorineural hearing loss. (KAS 11) Clinicians should not routinely prescribe antivirals, thrombolytics, vasodilators, or vasoactive substances to patients with sudden sensorineural hearing loss.

The guideline update group made *recommendations for* the following: (KAS 2) Clinicians should assess patients with presumptive sudden sensorineural hearing loss through history and physical examination for bilateral sudden hearing loss, recurrent episodes of sudden hearing loss, and/or focal neurologic findings. (KAS 4) In patients with sudden hearing loss, clinicians should obtain, or refer to a clinician who can obtain, audiometry as soon as possible (within 14 days of symptom onset) to confirm the diagnosis of sudden sensorineural hearing loss. (KAS 6) Clinicians should evaluate patients with sudden sensorineural hearing loss for retrocochlear pathology by obtaining magnetic resonance imaging or auditory brainstem response. (KAS 10) Clinicians should offer, or refer to a clinician who can offer, intratympanic steroid therapy when patients have incomplete recovery from sudden sensorineural hearing loss 2 to 6 weeks after onset of symptoms. (KAS 12) Clinicians should obtain follow-up audiometric evaluation for patients with sudden sensorineural hearing loss at the conclusion of treatment and within 6 months of completion of treatment. These recommendations were clarified in terms of timing of intervention and audiometry and method of retrocochlear workup.

The guideline update group offered the following KASs as *options*: (KAS 8) Clinicians may offer corticosteroids as initial therapy to patients with sudden sensorineural hearing loss within 2 weeks of symptom onset. (KAS 9a) Clinicians may offer, or refer to a clinician who can offer, hyperbaric oxygen therapy combined with steroid therapy within 2 weeks of onset of sudden sensorineural hearing loss. (KAS 9b) Clinicians may offer, or refer to a clinician who can offer, hyperbaric oxygen therapy combined with steroid therapy as salvage therapy within 1 month of onset of sudden sensorineural hearing loss.

Differences from Prior Guideline

- Incorporation of new evidence profiles to include quality improvement opportunities, confidence in the evidence, and differences of opinion
- Included 10 clinical practice guidelines, 29 new systematic reviews, and 36 new randomized controlled trials
- Highlights the urgency of evaluation and initiation of treatment, if treatment is offered, by emphasizing the time from symptom occurrence
- Clarification of terminology by changing potentially unclear statements; use of the term *sudden sensorineural hearing loss* to mean idiopathic sudden sensorineural hearing loss to emphasize that >90% of sudden sensorineural hearing loss is idiopathic sudden sensorineural hearing loss and to avoid confusion in nomenclature for the reader
- Changes to the KASs from the original guideline:
  - KAS 1—When a patient first presents with sudden hearing loss, conductive hearing loss should be distinguished from sensorineural.
  - KAS 2—The utility of history and physical examination when assessing for modifying factors is emphasized.
  - KAS 3—The word ''routine'' is added to clarify that this statement addresses nontargeted head computerized tomography scan that is often ordered in the emergency room setting for patients presenting with sudden hearing loss. It does not refer to targeted scans, such as temporal bone computerized tomography scan, to assess for temporal bone pathology.
  - KAS 4—The importance of audiometric confirmation of hearing status as soon as possible and within 14 days of symptom onset is emphasized.
  - KAS 5—New studies were added to confirm the lack of benefit of nontargeted laboratory testing in sudden sensorineural hearing loss.
  - KAS 6—Audiometric follow-up is excluded as a reasonable workup for retrocochlear pathology. Magnetic resonance imaging, computerized tomography scan if magnetic resonance imaging cannot be done, and, secondarily, auditory brainstem response evaluation are the modalities

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recommended. A time frame for such testing is not specified, nor is it specified which clinician should be ordering this workup; however, it is implied that it would be the general or subspecialty otolaryngologist.
    - KAS 7—The importance of shared decision making is highlighted, and salient points are emphasized.
    - KAS 8—The option for corticosteroid intervention within 2 weeks of symptom onset is emphasized.
    - KAS 9—Changed to KAS 9A and 9B. Hyperbaric oxygen therapy remains an option but only when combined with steroid therapy for either initial treatment (9A) or salvage therapy (9B). The timing of initial therapy is within 2 weeks of onset, and that of salvage therapy is within 1 month of onset of sudden sensorineural hearing loss.
    - KAS 10—Intratympanic steroid therapy for salvage is recommended within 2 to 6 weeks following onset of sudden sensorineural hearing loss. The time to treatment is defined and emphasized.
    - KAS 11—Antioxidants were removed from the list of interventions that the clinical practice guideline recommends against using.
    - KAS 12—Follow-up audiometry at conclusion of treatment and also within 6 months posttreatment is added.
    - KAS 13—This statement on audiologic rehabilitation includes patients who have residual hearing loss and/or tinnitus who may benefit from treatment.
- Addition of an algorithm outlining KASs
- Enhanced emphasis on patient education and shared decision making with tools provided to assist in same

Keywords

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

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Introduction

Sudden hearing loss (SHL) is a frightening symptom that often prompts an urgent or emergent visit to a clinician. This guideline update focuses on sudden sensorineural hearing loss (SSNHL), the majority of which is idiopathic and which, if not recognized and managed promptly, may result in persistent hearing loss and tinnitus and reduced patient quality of life (QOL).[1] SSNHL affects 5 to 27 per 100,000 people annually, with about 66,000 new cases per year in the United States.[2-4] Throughout this guideline the following definitions are used (see **Table 1**):

- SHL is defined as a rapid-onset subjective sensation of hearing impairment in one or both ears. The hearing loss in SHL may be a conductive hearing loss (CHL), sensorineural hearing loss (SNHL), or mixed hearing loss, defined as both CHL and SNHL occurring in the same ear. CHL and the conductive component of mixed hearing loss may be due to an abnormality in the ear canal, tympanic membrane (''ear drum''), or middle ear.
    - Physical examination will help determine if there is obstructing cerumen or a foreign body in the ear canal, if there is a perforation of the tympanic membrane, or if there is fluid in the middle ear.
    - Tuning fork testing will enable the initial treating clinician to distinguish CHL from SNHL so that the SSNHL evaluation and management pathway can be triggered appropriately.
- SSNHL is a subset of SHL that is (a) sensorineural in nature, (b) occurs within a 72-hour window, and (c) meets certain audiometric criteria.
    - SNHL is sometimes referred to colloquially as ''nerve hearing loss'' and indicates abnormal functioning of the cochlea, auditory nerve, or higher aspects of central auditory perception or processing.
    - The most frequently used audiometric criterion for SSNHL is a decrease in hearing of $\geq$30 decibels affecting at least 3 consecutive frequencies. Because premorbid audiometry is generally unavailable, hearing loss is often defined in relation to the opposite ear's thresholds.
    - The guideline update group (GUG) acknowledges that in both clinical practice and in research studies, less stringent criteria for SSNHL are employed.
    - SSNHL is often but not always accompanied by tinnitus and/or vertigo. The tinnitus may persist and may be disturbing to the patient.
- Idiopathic SSNHL (ISSNHL) is defined as SSNHL with no identifiable cause despite adequate investigation. This is the situation in 90% of patients with SSNHL and is the primary focus of this clinical practice guideline (CPG) update. The use of SSNHL in this document is equivalent to ISSNHL, as determined after the appropriate workup denoted in Key Action Statement 1 (KAS 1) and KAS 2.

The SSNHL definition used throughout this guideline is $\geq$30-dB SNHL at 3 consecutive frequencies, based on its consistent use in the literature and National Institute on Deafness and Other Communication Disorders (NIDCD)

Table 1. Definitions of Common Terminology.

Sudden hearing loss (SHL)	A rapid-onset subjective sensation of hearing impairment in one or both ears.
Sensorineural hearing loss (SNHL)	Hearing loss resulting from abnormal function of the cochlea, auditory nerve, or higher aspects of central auditory perception or processing.
Conductive hearing loss (CHL)	Hearing loss resulting from a problem conducting sound waves anywhere along the route through the outer ear, tympanic membrane, or middle ear.
Mixed hearing loss (MHL)	Hearing loss resulting from both SNHL and CHL occurring in the same ear.
Sudden sensorineural hearing loss (SSNHL)	A subset of SHL that (a) is sensorineural in nature, (b) occurs within a 72-hour window, and (c) consists of a decrease in hearing of $\geq$30 decibels affecting at least 3 consecutive frequencies.
Idiopathic sudden sensorineural hearing loss (ISSNHL)	SSNHL with no identifiable cause despite adequate investigation.
Salvage therapy	Any therapy offered after 2 weeks from symptom onset (even if initial therapy was observation).

criteria[5]; however, the GUG recognizes that in clinical practice, expanding the definition to cases with <30 decibels of hearing loss may be considered. The GUG recognizes that the NIDCD definition is not universally used and, accordingly, published evidence not using this definition was considered.

The distinction between SSNHL and sudden conductive or mixed hearing loss is one that should be made by the initial treating health care provider for early diagnosis and management to 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, malignancy, noise, and ototoxic medications.[6-9]

Much of the literature indicates that 32% to 65% of cases of SSNHL may recover spontaneously.[4,10] Clinical experience, however, shows that these numbers may be an overestimation. It is important to remember that tinnitus is a frequent comorbidity that may persist and, with time, may become the patient's primary concern. Details on tinnitus management can be found in the American Academy of Otolaryngology–Head and Neck Surgery Foundation (AAO-HNSF) ''Clinical Practice Guideline: Tinnitus.''[11] The 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.[10,12,13] Additionally, the psychological and communicative detriment experienced during an acute episode of SHL and then in potentially unrecovered hearing loss and persistent tinnitus creates a strong desire for treatment.[14,15]

Treatment options that have been proffered for SSNHL are myriad and include systemic and topical steroids, antiviral agents, hyperbaric oxygen therapy (HBOT), rheologic agents, diuretics, other medications, herbal and other complementary and alternative treatments, middle ear surgery for fistula repair, and observation alone.

Long-term follow-up is recommended, as some patients (up to one-third) will have an underlying cause that is eventually identified but was not evident at initial presentation.[16] Additionally, the patient with partial or no hearing recovery and/or persistent tinnitus will require ongoing management from otolaryngologic, audiologic, and psychological perspectives.[17]

This multidisciplinary guideline update is intended for all clinicians who diagnose or manage adult patients (aged $\geq$18 years) who present with SHL. After addressing causes, diagnosis, and treatments of sudden conductive/mixed hearing loss briefly, this guideline update will go on to address SSNHL in detail.

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 created a pressing need for the original evidence-based guideline[10] in 2012 to aid clinicians in managing SSNHL and for this update now. Moreover, wide variations in evaluation, treatment, counseling, and follow-up of patients with SSNHL continue to exist within the United States and worldwide. Such variations are usually ascribed to heterogeneity in clinical practice and training rather than to differences in clinical need.

Data show that, since publication of the initial SHL CPG, adherence to KAS recommendations is not universal.[18] Among otolaryngologists, there is high adherence to the recommendations to rule out CHL, to avoid routine head computed tomography (CT) scan, and to perform a retrocochlear workup. There is moderate adherence to the recommendations to avoid routine laboratory assessment and avoid other treatments (nonsteroid/non-HBOT). In this specialty group, however, there is low adherence to the recommendations for patient education regarding the natural history of SSNHL and for counseling regarding hearing rehabilitation. As for the original CPG's statements regarding systemic steroid therapy as an option for primary treatment and intratympanic (IT) steroid therapy as a recommendation for salvage treatment, otolaryngologists in this

study opted to prescribe both interventions—initial oral steroid therapy and salvage IT steroid therapy.

Nonotolaryngologists more commonly ordered routine head CT and performed routine nontargeted (often called ''shotgun'') laboratory assessments despite recommendations against these actions. They did not pursue retrocochlear workup or provide patient education as recommended.

Ten research needs were identified in the original SHL CPG:

1. Determine a standardized and evidence-based definition of SSNHL.
2. Investigate the effectiveness of corticosteroid treatment versus a placebo.
3. Further investigate the benefit of HBOT and standardized HBOT treatment protocols for ISSNHL.
4. Develop standardized outcome criteria to aid the comparison of clinical studies.
5. Further study IT steroids as salvage therapy with particular attention to the optimal medications, dosage, concentrations, timing, 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,'' such as oral and IT steroids, in patients with ISSNHL.
9. Develop long-term follow-up protocols for patients with ISSNHL
10. Evaluate therapies with standardized definitions and treatment protocols across studies.

The current CPG update addresses the research questions that have been answered and the research needs that remain. Additionally, novel agents in trials are mentioned so that the reader may be alerted to new developments in the field.

The incomplete adoption of CPG recommendations and the ongoing lack of consensus on all aspects of the care of the patient with SSNHL further support the need for a user-friendly evidence-based CPG update to highlight and encourage use of best practices.

Guideline Scope and Purpose

The purpose of this multidisciplinary guideline update is to provide clinicians with evidence-based recommendations in evaluating patients with SHL, with emphasis on managing SSNHL. The guideline update is intended for all clinicians who are likely to diagnose and manage adults aged $\geq$18 years with SHL and applies to any setting in which an adult with SHL would be identified, managed, or monitored. The recommendations outlined in this guideline update 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 update is not intended to replace individualized patient care or clinical judgment. Information for patients is also provided, in appropriate language, to facilitate understanding and shared decision making.

Although the guideline update focuses primarily on managing SSNHL, the GUG recognized that patients enter the health care system with SHL as a nonspecific primary complaint. Therefore, the initial recommendations of the guideline update address an efficient manner by which to distinguish SNHL from CHL at the time of presentation. This distinction will often fall to the primary care or emergency room physician or other health care provider. Therefore, there is detailed discussion in this CPG of what otolaryngologists might consider obvious in the physical examination, including the use of tuning forks to distinguish CHL from SNHL. The purpose of the guideline update is not to present an exhaustive approach to managing CHL.

This is a multidisciplinary and interprofessional (herewith ''multidisciplinary'') update of the first CPG on ISSNHL developed in the United States. This guideline update will provide evidence-based recommendations for clinicians based on multidisciplinary consensus and careful consideration of the benefits versus harms of suggested actions. By focusing on opportunities for quality improvement, the update should further 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 27 per 100,000 people annually (United States), with some estimates ranging as high as 160 per 100,000 (Germany).[2,19,20] The US data may be underestimated, as individuals who experience mild SHL and/or a quick, spontaneous resolution may not seek medical care.

For most patients with SHL, their medical journey often starts at an emergency room, walk-in or urgent care clinic, or primary care physician's office. Even with the lower incidences quoted earlier, this represents between 15,000 and 60,000 patients seen in US urgent/emergency care or primary care clinics for this problem annually.

Coexistent morbidities, such as dizziness and tinnitus, pose considerable disease burdens for the patient with SSNHL. Dizziness is present in 30% to 60% of cases of SSNHL.[12,21,22] The presence of dizziness or vertigo at time of onset of SSNHL is seen often in more severe cases and is frequently associated with poorer prognosis for hearing recovery.[21] Tinnitus is nearly universal in SSNHL and, if persistent and bothersome, may pose a significant economic and psychological burden.[11,23] Recovery of hearing after SSNHL is often accompanied by improvement of the attendant tinnitus. Residual tinnitus may exacerbate or supersede the psychological and functional burden of nonrecovered hearing loss in SSNHL.[24]

The audiologic needs of patients with SSNHL are considerable and can be costly. Patients with sudden unilateral

hearing loss have immediate difficulty with conversation on the involved side and overall hearing in noisy environments. If they have preexisting hearing loss from common causes, such as presbycusis and noise exposure, SSNHL will compound the problem. In patients with SSNHL, the hearing asymmetry often results 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 make it difficult for patients who rely on hearing to avoid dangerous situations, such as crossing a busy street, thereby posing safety concerns. Repeated audiometric assessment with continued follow-up is needed. Rehabilitation of patients with persistent hearing loss following 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 SNHL on patients' QOL has been shown in adults and children.[25,26] Sudden SNHL, particularly when accompanied by tinnitus and dizziness, can result in even greater decrements in QOL.[10,14,23] Patients may experience fear and frustration at the inability to identify a cause for their hearing loss.

The cumulative weight of this disease burden underlies the importance of an updated CPG to optimize care of patients with this debilitating condition.

Methods

General Methods and Literature Search

In developing this update of the evidence-based CPG, the methods outlined in the AAO-HNSF ''Clinical Practice Guideline Development Manual, Third Edition'' were followed explicitly.[27]

An executive summary of the original ''Clinical Practice Guideline: Sudden Hearing Loss''[10] was sent to a panel of expert reviewers from the fields of general otolaryngology, otology, neurotology, neurology, family practice, advanced practice nursing, emergency medicine, radiology, and audiology, who assessed the KASs to decide if they should be kept in their current form, revised, or removed and to identify new research that might affect the guideline recommendations. The reviewers concluded that the original guideline action statements remained valid but should be updated with minor modifications. Suggestions were also made for new KASs.

The recommendations in this CPG are based on systematic reviews identified by a professional information specialist using an explicit search strategy. Additional background evidence included RCTs as needed to supplement the systematic review or to fill gaps when a review was not available. An information specialist conducted a systematic literature search using a validated filter strategy to identify CPGs, systematic reviews, and RCTs published since the prior guideline (2012). Search terms used were as follows: (''Hearing Loss, Sudden''[Mesh] OR ''sudden hearing loss''[ti] OR ''sudden deafness''[ti] OR ''sudden sensorineural hearing loss''[ti] OR ''idiopathic sudden hearing loss''[ti]). These search terms were used to capture all evidence on the population, incorporating all relevant treatments and outcomes. In certain instances, targeted searches for lower-level evidence were performed to address gaps from the systematic searches identified in writing the guideline. The original search was updated from January 2011 to July 2017 to include MEDLINE, EMBASE, Web of Science, Cumulative Index to Nursing and Allied Health Literature, Cochrane Database of Systematic Reviews, National Guideline Clearinghouse, Allied and Complementary Medicine Database, Canadian Medical Association Infobase, and National Institute for Health and Care Excellence.

1. The initial search for CPGs identified 20 guidelines. After removal of duplicates, irrelevant references, and non–English language articles, 2 guidelines were reviewed for inclusion. Quality criteria for including guidelines were (a) an explicit scope and purpose, (b) multidisciplinary stakeholder involvement, (c) systematic literature review, (d) explicit system for ranking evidence, and (e) explicit system for linking evidence to recommendations. Additional targeted searches were performed, which resulted in the inclusion of 10 CPGs to the CPG update; this includes the prior version of this AAO-HNSF CPG.
2. The initial search for systematic reviews identified 127 systematic reviews or meta-analyses. After removal of duplicates, irrelevant references, and non–English language articles, 32 articles were reviewed for inclusion. Quality criteria for including reviews were (a) relevance to the guideline topic, (b) clear objective and methodology, (c) explicit search strategy, and (d) valid data extraction methods. After the public review process, 1 further systematic review and 1 further meta-analysis were included. The final data set retained was 29 systematic reviews or meta-analyses that met inclusion criteria.
3. The initial search for RCTs identified 83 RCTs that were distributed to GUG members for review. After removal of duplicates, irrelevant references, and non–English language articles, 30 articles were reviewed for inclusion. Quality criteria for including RCTs were (a) relevance to the guideline topic, (b) publication in a peer-reviewed journal, and (c) clear methodology with randomized allocation to treatment groups. The total final data set retained 36 RCTs that met inclusion criteria.

The AAO-HNSF assembled a GUG representing the disciplines of otolaryngology–head and neck surgery, otology, neurotology, family medicine, audiology, emergency medicine, neurology, radiology, advanced practice nursing, and consumer advocacy. The GUG had 3 conference calls and 1 in-person meeting, during which it defined the scope and

Table 2. Aggregate Grades of Evidence by Question Type.[a]

Grade	OCEBM Level	Treatment	Harm	Diagnosis	Prognosis
A	1	Systematic review[b] of randomized trials	Systematic review[b] of randomized trials, nested case-control studies, or observational studies with dramatic effect	Systematic review[b] of cross-sectional studies with consistently applied reference standard and blinding	Systematic review[b] of inception cohort studies[c]
B	2	Randomized trials or observational studies with dramatic effects or highly consistent evidence	Randomized trials or observational studies with dramatic effects or highly consistent evidence	Cross-sectional studies with consistently applied reference standard and blinding	Inception cohort studies[c]
C	3-4	Nonrandomized or historically controlled studies, including case-control and observational studies	Nonrandomized controlled cohort or follow-up study (postmarketing surveillance) with sufficient numbers to rule out a common harm; case-series, case-control, or historically controlled studies	Nonconsecutive studies, case-control studies, or studies with poor, nonindependent, or inconsistently applied reference standards	Cohort study, control arm of a randomized trial, case series, or case-control studies; poor-quality prognostic cohort study
D	5	Case reports, mechanism-based reasoning, or reasoning from first principles			
X	N/A	Exceptional situations where validating studies cannot be performed and there is a clear preponderance of benefit over harm			

Abbreviation: OCEBM, Oxford Centre for Evidence-Based Medicine.
[a]Adapted from Oxford Centre for Evidence-Based Medicine Work Group.[30]
[b]A systematic review may be downgraded to level B because of study limitations, heterogeneity, or imprecision.
[c]A group of individuals identified for subsequent study at an early uniform point in the course of the specified health condition or before the condition develops.

objectives of updating the guideline, reviewed comments from the expert panel review for each KAS, identified other quality improvement opportunities, and reviewed the literature search results.

The evidence profile for each statement in the earlier guideline was then converted into an expanded action statement profile for consistency with our current development standards.[27] Information was added to the action statement profiles regarding the quality improvement opportunity to which the action statement pertained, the guideline panel's level of confidence in the published evidence, differences of opinion among panel members, and the feasibility of measurability and implementability. New KASs were developed with an explicit and transparent a priori protocol for creating actionable statements based on supporting evidence and the associated balance of benefit and harm. Electronic decision support software (BRIDGE-Wiz, Yale Center for Medical Informatics; New Haven, Connecticut) was used to facilitate creating actionable recommendations and evidence profiles.[28]

The updated guideline then underwent GuideLine Implementability Appraisal to assess adherence to methodologic standards, to improve clarity of recommendations, and to predict potential obstacles to implementation.[29] The GUG received summary appraisals and modified an advanced draft of the guideline based on the appraisal. That modified draft of the updated CPG was again revised per the comments received during multidisciplinary peer review, open public comment, and journal editorial peer review, resulting in the final manuscript. 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 harm, 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 **Table 2** and **Table 3**.[30,31]

Guidelines are not intended to supersede professional judgment but rather may be viewed as a relative constraint on individual clinician discretion in a particular clinical

Table 3. Strength of Action Terms in Guideline Statements and Implied Levels of Obligation.

Strength	Definition	Implied obligation
Strong recommendation	A strong recommendation means that the benefits of the recommended approach clearly exceed the harms (or, in the case of a strong negative recommendation, that the harms clearly exceed the benefits) and that the quality of the supporting evidence is high (grade A or B).[a] 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 that the benefits exceed the harms (or, in the case of a negative recommendation, that the harms exceed the benefits), but the quality of evidence is not as high (grade B or C).[a] 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 is suspect (grade D)[a] or well-done studies (grade A, B, or C)[a] show little clear advantage to one approach versus 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.

[a]Adapted from the American Academy of Pediatrics classification scheme.[31] (See **Table 2** for definitions of evidence grades.)

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.[32] 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.[31] 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 2 years were compiled and distributed before the first conference call. After review and discussion of these disclosures,[33] 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 a participant earns a living, and the participant's previously established ''stake'' in an issue.[34] Conflicts were again delineated at the start of the in-person meeting and at the start of each teleconference meeting, with the same caveats followed. All conflicts are disclosed at the end of this document.

Guideline KASs

Each evidence-based statement is organized in a similar fashion: a KAS is in bold, followed by the strength of the recommendation in italics. Each KAS is followed by an ''action statement profile'' that explicitly states the quality improvement opportunity, aggregate evidence quality, level of confidence in evidence (high, medium, low), benefit, harms, risks, costs, and a benefits-harm assessment. Additionally, there are statements of any value judgments, the role of patient preferences, clarification of any intentional vagueness by the panel, exceptions to the statement, any differences of opinion, and a repeat statement of the strength of the recommendation. Several paragraphs subsequently discuss the evidence supporting the statement. An overview of each evidence-based statement in this guideline can be found in **Table 4**.

For the purposes of this guideline, *shared decision making* refers to the exchange of information regarding treatment risks and benefits, as well as the expression of

Table 4. Summary of Guideline Key Action Statements.

Statement	Action	Strength
1. Exclusion of conductive hearing loss	Clinicians should distinguish sensorineural hearing loss (SNHL) from conductive hearing loss (CHL) when a patient first presents with SHL.	Strong recommendation
2. Modifying factors	Clinicians should assess patients with presumptive SSNHL through history and physical examination for bilateral SHL, recurrent episodes of SHL, and/or focal neurologic findings.	Recommendation
3. Computed tomography	Clinicians should **not** order routine computed tomography (CT) of the head in the initial evaluation of a patient with presumptive SSNHL.	Strong recommendation against
4. Audiometric confirmation of SSNHL	In patients with SHL clinicians should obtain, or refer to a clinician who can obtain, audiometry as soon as possible (within 14 days of symptom onset) to confirm the diagnosis of SSNHL.	Recommendation
5. Laboratory testing	Clinicians should **not** obtain routine laboratory tests in patients with SSNHL.	Strong recommendation against
6. Retrocochlear pathology	Clinicians should evaluate patients with SSNHL for retrocochlear pathology by obtaining an MRI or auditory brainstem response (ABR).	Recommendation
7. Patient education	Clinicians should educate patients with SSNHL about the natural history of the condition, the benefits and risks of medical interventions, and the limitations of existing evidence regarding efficacy.	Strong recommendation
8. Initial corticosteroids	Clinicians may offer corticosteroids as initial therapy to patients with SSNHL within 2 weeks of symptom onset.	Option
9a. Initial therapy with hyperbaric oxygen therapy	Clinicians may offer, or refer to a clinician who can offer, hyperbaric oxygen therapy (HBOT) combined with steroid therapy within 2 weeks of onset of SSNHL.	Option
9b. Salvage therapy with hyperbaric oxygen therapy	Clinicians may offer, or refer to a clinician who can offer, hyperbaric oxygen therapy (HBOT) combined with steroid therapy as salvage within 1 month of onset of SSNHL.	Option
10. Intratympanic steroids for salvage therapy	Clinicians should offer, or refer to a clinician who can offer, intratympanic steroid therapy when patients have incomplete recovery from SSNHL 2 to 6 weeks after onset of symptoms.	Recommendation
11. Other pharmacologic therapy	Clinicians should **not** routinely prescribe antivirals, thrombolytics, vasodilators, or vasoactive substances to patients with SSNHL.	Strong recommendation against
12. Outcomes assessment	Clinicians should obtain follow-up audiometric evaluation for patients with SSNHL at the conclusion of treatment and within 6 months of completion of treatment.	Recommendation
13. Rehabilitation	Clinicians should counsel patients with SSNHL who have residual hearing loss and/or tinnitus about the possible benefits of audiologic rehabilitation and other supportive measures.	Strong recommendation

patient preferences and values, which result in mutual responsibility in decisions regarding treatment and care.[35] The role of patient preferences in making decisions deserves further clarification. The GUG classified the role of patient preference per consensus among the group as ''none, small, moderate, or large.'' For some statements where the evidence base demonstrates clear benefit, although the role of patient preference for a range of treatments may not be relevant (eg, with tuning fork testing), clinicians should provide patients with clear and comprehensible information on the benefits to facilitate understanding and shared decision making, which in turn leads to better patient adherence and outcomes. In cases where evidence is weak or benefits unclear, the practice of shared decision making, where the management decision is made by a collaborative effort between the clinician and an informed patient, is extremely useful. Factors related to patient preference include (but are not limited to) absolute benefits, adverse effects, cost of medications or procedures, and frequency and duration of treatment, as well as certain less tangible factors, such as religious and/or cultural beliefs or personal levels of desire for intervention. As with all counseling, documentation of

the patient discussion and shared decision making should be entered into the patient chart.

Key Action Statements

STATEMENT 1. EXCLUSION OF CHL: Clinicians should distinguish SNHL from CHL when a patient first presents with SHL. *Strong recommendation based on systematic reviews and cross-sectional studies with a preponderance of benefit over harm.*

Action Statement Profile: 1

- Quality improvement opportunity: Identifying patients who are appropriate for the guideline recommendations and those with CHL who may benefit from other therapies. (National quality strategy: Prevention and Treatment of Leading Causes of Morbidity and Mortality)
- 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 by history, examination, and tuning fork tests
- Level of confidence in the evidence: High
- Benefits: Guide the choice of appropriate diagnostic tests, avoid misdiagnosis, improve diagnostic accuracy, ensure that treatment is consistent with diagnosis, guide patient expectations, identify CHL that can be treated and resolved
- Risks, harms, costs: None
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: Panel consensus that despite a lack of systematic research evidence supporting this action, distinguishing SNHL was an essential first step in appropriate subsequent management
- Intentional vagueness: None
- Role of patient preferences: None
- Exceptions: None
- Policy level: Strong recommendation
- Differences of opinion: None

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 including tuning fork tests, and audiometry. CHL and SNHL have markedly different management strategies, and there is good evidence that CHL, such as that from cerumen impaction or middle ear effusion, can be treated effectively.[36-38] A delay in treatment of SSNHL may result when a clinician assumes that a patient has CHL without considering a diagnosis of SNHL.[39] Hearing loss is classified as conductive, sensorineural, or mixed. CHL results from abnormalities of the external ear, tympanic membrane, middle ear air space, or ossicles—structures that ''conduct'' sound waves to the cochlea. SNHL results from 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. The patient presents with a complaint of SHL, which may be accompanied by tinnitus and/or vertigo. It is the GUG's experience that patients cannot accurately distinguish subjective hearing loss as either CHL or SNHL. Clinicians should ask patients about a history of trauma, ear pain, history of ear canal instrumentation, ear drainage, fever, and other neurologic or systemic symptoms. (See KAS 2 for additional key elements of the patient history.) Patients with SSNHL often report tinnitus, ear fullness or pressure, and vertigo.[18,39] Some of these symptoms, however, may also be present in CHL.[37,38] Hearing loss associated with ear fullness should therefore not be presumed to be CHL.[39] A focused physical examination is required.

Physical Examination. A thorough examination of the ears, including inspection of the ear canals and visualization of the tympanic membranes, is essential in SHL to distinguish CHL from SNHL. Refer to http://www.entnet.org/content/ent-exam-video-series for video instruction of a proper ear examination. 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,[18] while examination of patients with CHL will often but not always show abnormalities.[40,41] Impacted cerumen, if present, must be removed prior to establishing a diagnosis in patients with SHL.[38]

The Weber and Rinne tuning fork tests have been used traditionally to differentiate CHL and SNHL (**Table 5**).[10,39,42] Several authors, however, have noted that the Weber and Rinne tuning fork test results may not be reliably reproduced between examiners and that the results can be misleading.[10,43-45] Those same studies also showed than an abnormal Rinne significantly increases the probability of a CHL with a likelihood ratio between 2.7 to 62.[10] When the Weber and Rinne were consistent with each other, the sensitivity was as high as 95%, but the 2 tests agreed only 50% of the time.[45] Burkey et al showed that the Rinne could correctly distinguish between a sensorineural and conductive loss in 96% of cases with an overall accuracy of 91% in nonexpert users.[44] The only study to have focused on the use of a tuning fork in detecting SSNHL agreed that while the Weber could be unreliable >20% of the time, its sensitivity was 99% for those patients in which the Weber lateralized away from the ear in question, prompting an urgent workup with audiometric testing.[42] As such, the GUG agreed that tuning fork tests should be used in the

Table 5. Recommended Technique for Weber and Rinne Testing.

Weber Test	Rinne Test
1. Vibrate the tuning fork by striking it on your (covered) elbow or knee, not on a hard metallic or wooden surface. 2. Place vibrating tuning fork (256 or 512 Hz) at midline of forehead or on maxillary teeth (not false teeth) 3. Ask where the sound is heard; it is normal to hear at the midline or "everywhere" 4. If the sound lateralizes to one ear, then a. There is CHL in that ear. OR b. There is SNHL in the opposite ear.	1. Vibrate the tuning fork by striking it on your (covered) elbow or knee, not on a hard metallic or wooden surface. 2. 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) with the tines directed toward the ear. 3. The sound should be heard better via air conduction (at the entrance to the ear canal) 4. If the sound is heard better by bone conduction in the same ear, then there is CHL in that ear. 5. If the sound is heard better by bone conduction but in the opposite ear, there is SNHL in the test ear. 6. Repeat for the other ear.

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

immediate setting in conjunction with otoscopic examination to help clinicians make a preliminary diagnosis of either CHL or SNHL prior to audiometry being available. Tuning fork tests, while giving reasonable initial information to distinguish between CHL and SNHL, do not supplant formal audiometric testing, as discussed in KAS 4. It is important to strike the tuning fork correctly, on a protected elbow or knee, to avoid nonharmonic frequencies that are noted when the fork is struck against a wooden or metal surface.[46] Refer to https://www.youtube.com/watch?v=2js72BYjZAw for video instruction of the proper performance of the Weber and Rinne tests. Combining the information gleaned from both Weber and Rinne testing will aid greatly in making this important distinction between CHL and SNHL.

The GUG understands that a tuning fork may not be available to the initial health care provider who encounters the patient who presents with SHL. It is our hope that by emphasizing its utility here and with the accompanying how-to video that we encourage the addition of tuning fork examination to the thorough history taking and physical examination that are performed to exclude cases of CHL. A reasonable alternative to the Weber tuning fork test is the hum test—the patient is asked to hum and if he or she hears one's own hum louder in the affected ear, it is likely CHL in that ear. The sensitivity, specificity, and diagnostic accuracy of the hum test is similar to that of the Weber test.[47] Rinne testing cannot, of course, be done with a hum. Neither of these has the accuracy of audiometry, but each can be used in the initial assessment if better testing methods are not available.

STATEMENT 2. MODIFYING FACTORS: Clinicians should assess patients with presumptive SSNHL through history and physical examination for bilateral SHL, recurrent episodes of SHL, and/or focal neurologic findings. *Recommendation based on observational studies with a preponderance of benefit over harm.*

Action Statement Profile: 2

- Quality improvement opportunity: Identify patients with potentially serious alternative conditions for whom the subsequent guideline recommendations do not apply. (National quality strategy: Prevention and Treatment of Leading Causes of Morbidity and Mortality; Effective Communication and Care Coordination)
- Aggregate evidence quality: Grade C, based on observational studies and case series studies
- Level of confidence in the evidence: High
- Benefits: Identification of patients with a high likelihood of alternative and potentially serious underlying cause, who require specialized assessment and management
- Risks, harms, costs: Time of assessment
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: None
- Role of patient preferences: None
- Exceptions: None
- Policy level: Recommendation
- Differences of opinion: None

Supporting Text

The purpose of this statement is to encourage clinicians to look for clinical features in patients with presumptive ISSNHL that may be associated with an underlying disease. ISSNHL is rarely bilateral or recurrent and is not associated with other focal neurologic symptoms or signs. The clinician should assess the patient for these findings by history and general physical and neurologic examination. If available, audiometry and targeted imaging will likely provide additional diagnostic help. **Table 6** lists some symptoms

Table 6. Some Symptoms and Signs Suggestive of Nonidiopathic Sudden Sensorineural Hearing Loss.

- Sudden onset of bilateral hearing loss
- Antecedent fluctuating hearing loss on one or both sides
- Concurrent severe bilateral vestibular loss with oscillopsia
- Gaze evoked or downbeat nystagmus
- Concurrent eye pain, redness, lacrimation, and photophobia
- Focal neurologic symptoms or signs, such as headache, confusion, diplopia, dysarthria, focal weakness, focal numbness, ataxia, facial weakness
- Recent head trauma
- Recent acoustic trauma
- Recent barotrauma

and signs that would suggest that the hearing loss may be associated with an underlying condition.

Bilateral SHL. Most patients with ISSNHL have unilateral hearing loss; bilateral loss is rare and should prompt consideration of other causes as listed in **Table 7**.[10,19] On occasion, these same mechanisms may produce unilateral hearing loss.

Prior Episodes of SSNHL or Fluctuating Hearing Loss. Most cases of SSNHL are not preceded by fluctuating hearing, so this feature in the history should raise suspicion for this being a nonidiopathic case. Patients with a history of a fluctuating hearing loss presenting with SSNHL should be evaluated for causes such as Ménière's disease, autoimmune inner ear disease, Cogan's syndrome, and hyperviscosity syndrome. Ménière's disease is the most common cause of this presentation.[48] In all these conditions, hearing declines in a stepwise or fluctuating manner but may occasionally decline suddenly and present as SSNHL, most commonly unilateral. Autoimmune inner ear disease and Cogan's syndrome may be exceptions in that bilateral involvement is common at onset.[49-53]

SSNHL With Focal Neurologic Findings. SSNHL in the presence of new focal neurologic symptoms or signs indicates central nervous system involvement. Stroke and transient ischemic attack rarely present as isolated SSNHL. Peripheral vestibular involvement is usually present with brainstem infarct, primarily involving the lateral pontomedullary region, the middle cerebellar peduncle, and the cerebellum.[54-57] Anterior inferior cerebellar artery (AICA) occlusion is a significant cause of otovestibular compromise in stroke.[58] Vestibular symptoms can be the result of peripheral vestibular ischemia, infarction of the central vestibular structures in the region, or a combination of both.[56,59] Central nervous system symptoms and signs include vertigo, dysarthria, ipsilateral Horner's syndrome (miosis, ptosis, and anhidrosis), diplopia, nystagmus, ipsilateral facial numbness, contralateral body numbness, dysmetria, and ataxia.[54,55,57] The vascular supply to the region derives from the vertebrobasilar system through the AICA and internal auditory artery. Atherosclerosis, thrombosis, embolism, or dissection may lead to occlusion of any of these arteries and result in stroke. Sudden bilateral hearing loss has been reported to be a prodrome to a stroke in the AICA distribution when there is underlying severe atherosclerotic narrowing of the vertebrobasilar vessels.[60,61] Bilateral hemispheric stroke involving the auditory cortex and/or associated subcortical areas may rarely cause complaints of deafness and can be associated with other abnormalities, such as auditory agnosia. Unilateral hemispheric stroke involving the auditory cortex may cause subtle hearing dysfunction, but complaints of deafness are exceedingly rare.[62]

Multiple sclerosis has been reported to present with SSNHL.[63,64] However, there are usually other focal neurologic symptoms or signs present simultaneously or previously. Isolated cranial nerve involvement is rare in patients with multiple sclerosis (10.4%), and isolated eighth nerve palsy is extremely rare (<1%). Magnetic resonance imaging (MRI) in multiple sclerosis shows white matter changes, most easily seen on FLAIR images (fluid-attenuated inversion recovery). Meningitis, whether infectious, inflammatory/autoimmune, or neoplastic, is usually associated with other symptoms, such as headache, other cranial nerve palsies, and other focal neurologic symptoms and signs. Lumbar puncture is usually abnormal with elevated cell count, protein, and other evidence of infection, inflammation, or neoplasm. Migraine may rarely cause stroke.[65] Tumors of the cerebellopontine angle, primarily vestibular schwannomas and meningiomas, occasionally present with SHL. Other more common symptoms are progressive hearing loss, dizziness or vertigo, facial weakness, dysmetria, and ataxia. The tumor size of a vestibular schwannoma does not correlate with the abruptness or the hearing loss or the degree of hearing loss and likelihood of hearing recovery.[10,12,66]

STATEMENT 3. COMPUTED TOMOGRAPHY: Clinicians should not order routine computed tomography (CT) of the head 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: 3

- Quality improvement opportunity: Avoiding unnecessary imaging (National quality strategy: Patient Safety)
- Aggregate evidence quality: Grade B, based on 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 (IV) contrast
- Level of confidence in the evidence: High
- Benefits: Avoidance of radiation, cost savings, reduced incidental findings, less inconvenience for the patient, avoiding false sense of security from false-negative scan

Table 7. Selected Conditions That May Be Associated with Bilateral Sudden Sensorineural Hearing Loss.

Cause	Other Features
Infection (viral, including herpes simplex virus, varicella zoster virus, human immunodeficiency virus, and others; bacterial; mycoplasma; Lyme; tuberculosis; syphilis; fungal)	Headache, fever, other cranial nerve palsies, abnormal cerebrospinal fluid commonly seen in meningitis; Pinna or ear canal vesicles and facial weakness are often seen in varicella zoster virus (Ramsay Hunt syndrome/herpes zoster oticus)[246-251]
Autoimmune inner ear disease	Hearing fluctuation, vertigo[49,50]
Ototoxic medication	Vestibular loss, oscillopsia[6,252]
Trauma	Temporal bone fracture with possible Battle's sign[252]; cochlear concussion without visible fracture; barotrauma
Lead poisoning	Learning disabilities, other stigmata of lead poisoning[253]
Genetic disorders	May be syndromic or nonsyndromic and may present later in life[254-256]
Mitochondrial disorders, including MELAS (metabolic encephalopathy, lactic acidosis, and stroke-like episodes) and others	Confusion, stroke like spells, elevated lactate, MRI white matter changes; others with variable phenotypes[257-261]
Stroke	Vertigo, dysarthria, facial weakness, ataxia, nystagmus, unilateral numbness, abnormal CT or MRI or MR angiogram of the vertebrobasilar vasculature[55,58,60,262-264]
Cogan's syndrome	No-syphilitic interstitial keratitis of the cornea, hearing loss, vertigo[53,265]
Neoplastic (neurofibromatosis II, bilateral vestibular schwannomas, carcinomatous meningitis, intravascular lymphomatosis, others)	Abnormal brain MRI, cerebrovascular imaging study, or cerebrospinal fluid[266-269]
Sarcoidosis	Pulmonary symptoms, bilateral vestibular loss, elevated angiotensin-converting enzyme level, abnormal Gallium scan[270,271]
Hyperviscosity syndrome	Mucous membrane bleeding, neurologic and pulmonary symptoms, associated retinopathy[272]

Abbreviations: CT, computed tomography; MR, magnetic resonance; MRI, magnetic resonance imaging.

- Risks, harms, costs: None
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: The word ''routine'' in radiology parlance means thick-cut CT of the head. Additionally, this indicates that while head CT to rule out intracranial bleed is not warranted in the absence of targeted neurologic findings, targeted imaging may be indicated if signs or clinical findings suggest an underlying etiology that is being explored. The panel recognizes that the terms ''initial evaluation'' are 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: Small
- Exceptions: Patients with focal neurologic findings
- Policy level: Strong recommendation
- Differences of opinion: None

Supporting Text

The purpose of this statement is to avoid inappropriate use of routine CT of the head in the initial assessment of patients with presumptive SSNHL. A routine head CT scan in common radiology parlance refers to CT with thick (5 mm) cuts through the brain. This is usually performed for evaluation of possible intracranial bleed or acute stroke. There is no high-resolution cut through the internal auditory canal (IAC) on routine head CT. A CT scan has some risks, which include radiation exposure and side effects of IV contrast, yet rarely offers useful information that would improve initial management. This statement does not apply to patients as identified in KAS 2 with focal neurologic findings, a history of trauma, or chronic ear disease, who may require a CT scan. This statement also does not imply that imaging studies are of no value in managing SSNHL patients, who may eventually benefit from MRI of the brain or a fine-cut, high-resolution CT scan of the temporal bone (not routine head/brain) (see KAS 6).[10,67]

The ACR has defined evidence-based appropriateness criteria for imaging studies with a rating of 1-3 for ''usually not appropriate,'' 4-6 for ''may be appropriate,'' and 7-9 for ''usually appropriate.''[68] 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 not indicated in these specific clinical settings or the risk-benefit ratio for patients is unfavorable.[68] None of the ACR scenarios, however, are limited to isolated SHL, which would achieve an even lower rating of

appropriateness. For patients with a history and physical examination consistent with a cholesteatoma or other identifiable pathologic condition of the ear or temporal bone, targeted temporal bone CT may be appropriate. This KAS makes a recommendation against routine head CT for patients without an identified cause, not for patients with findings on history or physical examination.

The ACR, as part of the appropriateness criteria, introduced a radiation dose assessment and relative radiation levels (RRLs) associated with different diagnostic tests.[68] 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 zero, while a chest x-ray in an adult has an RRL of 1, with a radiation dose estimate of <0.1 mSv, and a head CT scan has an RRL of 3, a radiation dose of 1-10 mSv. Therefore, nontargeted head/brain CT should be considered not only inappropriate but, in fact, unnecessarily harmful in the evaluation of SSNHL.

Other, Potentially Appropriate, Imaging Modalities. The principal differential diagnosis in the patient with suspected SSNHL is an inner ear versus a cochleovestibular nerve or brainstem abnormality. MRI has long replaced CT as the study of choice for detecting cerebellopontine angle tumors.[67,69-77] Also, because a CT scan does not have the resolution to detect brainstem infarcts in the early stages, emergent MRI is preferred, when the clinical situation warrants emergency imaging. The retrocochlear workup in patients with SSNHL is clarified further in KAS 6.

There are other situations where CT can be used in situations in which an MRI cannot be obtained, as in patients with pacemakers, severe claustrophobia, or even financial constraints. CT can also be considered in patients with known bone disease, such as Paget's disease, fibrous dysplasia, or bone metastasis to the temporal bone, although the history typically would have prompted the study.[78]

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 founded on history or physical examination and in whom SSNHL is suspected, routine head CT is a very low-yield examination with significant cost and radiation exposure and is not recommended.

STATEMENT 4. AUDIOMETRIC CONFIRMATION OF SSNHL: In patients with SHL clinicians should obtain, or refer to a clinician who can obtain, audiometry as soon as possible (within 14 days of symptom onset) to confirm the diagnosis of SSNHL. *Recommendation based on RCTs with a preponderance of benefit over harm.*

Action Statement Profile: 4

- Quality improvement opportunity: Ensuring an accurate diagnosis (National quality strategy: Prevention and Treatment of Leading Causes of Morbidity and Mortality; Effective Communication and Care Coordination)
- Aggregate evidence quality: Grade C, based on criteria used in RCTs assessing the benefits and timing for intervention for SSNHL
- Level of confidence in the evidence: High
- Benefits: Guiding treatment, identifying urgent conditions that require prompt management, ensuring that interventions for SSNHL are offered to those patients who meet appropriate audiometric criteria for diagnosis
- Risks, harms, costs: Potential delay in treatment until audiometry is obtained; direct cost of audiometry
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: Treatments are more likely to be effective if offered early. The expediency of diagnosis is necessary to ensure that treatment can be offered within a reasonable therapeutic window.
- Intentional vagueness: Although most of the group felt that earlier is better, the words ''as soon as possible (within 14 days of symptom onset)'' were used, given that barriers to access to care may make it unreasonable to set an earlier time point.
- Role of patient preferences: None
- Exceptions: 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
- Differences of opinion: While everyone in the group agreed that audiometry is necessary, there were differences of opinion regarding how expediently the test should be obtained. Some members felt that it should be within 72 hours while others felt within 2 weeks was adequate. We agreed on the current language that sets an outside limit on how long it can be, while encouraging earlier testing if feasible.

Supporting Text

The purpose of this statement is to encourage timely audiometric testing to identify objective, reproducible criteria for diagnosing a patient with SSNHL.

Audiometry is mandatory for definitively diagnosing SSNHL because it distinguishes CHL from SSNHL and establishes frequency-specific hearing thresholds and word recognition ability.

Varying criteria have been used in the literature to diagnose SSNHL, but a hearing loss $\geq$30 dB at 3 consecutive frequencies occurring within a 72-hour period is the definition adopted by the NIDCD and the definition used in most

RCTs.[5,10] The adoption of these criteria will increase the generalization of research findings by ensuring that patient characteristics are similar to those studied by the investigators in RCTs.

This definition assumes that the hearing level in each ear was either normal just prior to the episode of SSNHL or that hearing loss was symmetrical in each ear prior to the SSNHL. In the absence of premorbid audiometric information, clinicians may consider the ''degree of certainty'' from uncertain to certain with which they are comfortable when making a decision that the hearing loss in the poorer ear is ''new.''[79]

The importance of the accuracy of the initial and all follow-up audiometric evaluations is essential to achieve the goals of the recommendations within this guideline. Thus, all audiometric evaluations should employ adherence to current American National Standards Institute (1987, 2003, 2004) standards for equipment and environment used, age- and ability-appropriate techniques for audiometric assessment procedures for the patient, and appropriate validity checks, as well as follow joint clinical practice guidance and preferred practice patterns.[10,80-82] Key components of the audiologic evaluation may include the following:

a. A thorough hearing-specific case history.
b. Otoscopic examination, including management of excessive or obstructive cerumen.
c. Ear-specific air and bone conduction threshold measures with appropriate masking. Hearing thresholds should be measured at 250-8000 Hz, including 3000 and 6000 Hz with additional interoctaves as appropriate (ie, differences $\geq$20 dB).[10] Bone conduction hearing thresholds should be measured at octave intervals 250-4000 Hz and 3000 Hz.[10]
d. Speech audiometry measures in quiet and noise, with appropriate masking. Recorded stimuli are preferred for evaluation for standardization of outcomes, although monitored live voice may be used when appropriate.[83] Speech recognition threshold or speech detection/awareness thresholds for non–English speaking individuals and adults with developmental delay should be obtained in agreement with pure tone average (PTA), typically with standardized spondee word lists. This serves to quantify a hearing threshold level for speech stimuli as well as a validity check for the pure tone audiogram.
e. Word recognition scores (WRSs) measured in percentage of correct answers should be evaluated at a suprathreshold presentation level, typically 30- to 40-dB sensation level relative to speech recognition threshold with standardized word lists. This confirms performance in agreement with the pure tone audiogram, permits identification of unusual asymmetry that is not predicted by the hearing thresholds, and assists in making therapeutic and, eventually, rehabilitation decisions. Additional measures of words or sentences in background noise may be evaluated to quantify speech-in-noise deficits to further guide clinical and rehabilitation decisions. Results are typically presented in signal-to-noise ratio.

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 of the following for ongoing comparisons to be useful: air and bone conduction thresholds and speech audiometry, masking levels, reliability, validity, word lists used, method of presentation (monitored live voice or recorded), and type of transducer.[84,85]

e. Ear-specific immittance measurements completed with equipment calibrated to current American National Standards Institute standards.[81] Immittance measures may include tympanometry, static immittance, and acoustic reflex measures. These measures quantify middle ear function to rule out CHL. Furthermore, the acoustic reflex response pattern can support a site of lesion; however, this should not be used in isolation as other tests are necessary for confirmation.
   Caveat: The equipment used for acoustic reflex measures is capable of producing high-intensity stimuli (eg, $\geq$120 dB HL). Literature suggests that acoustic reflex measures can cause permanent hearing loss and tinnitus.[86] Although some authors have recommended that presentation levels not exceed 110 dB SPL,[87] there are no standards for safe presentation levels for these pure tone stimuli. The Occupational Safety and Health Administration recommends a limit of 115 dBA for brief duration noise, but a pure tone results in a greater amount of energy concentrated over a smaller area of the basilar membrane compared with noise.[88] It is important to exercise caution regarding performing these tests in the SSNHL population, as these individuals (particularly those with cochlear hearing loss) may be more sensitive to loud sounds as well as potentially more susceptible to noise trauma in the acute and subacute phases.
f. Otoacoustic emission (OAE) measures may be obtained to determine cochlear function. OAEs are sounds generated by the outer hair cells in the cochlea. They are recorded with a probe in the ear canal when the cochlea is stimulated by sound.[89] OAEs can be used to screen for those with a greater-than-mild hearing loss. In patients with greater-than-mild SSNHL, OAEs may help distinguish sensory from neural hearing loss, as normal responses are usually not obtained with hearing loss greater than 30-40 dB except in those with a neural hearing loss. In situations of limited resources and/or access to comprehensive audiometry, automated

audiometry can be considered as a secondary alternative.

STATEMENT 5. LABORATORY TESTING: Clinicians should not obtain routine laboratory tests in patients with SSNHL. *Strong recommendation against based on 1 large cross-sectional study and a large number of other studies as well as a preponderance of benefit over harm.*

Action Statement Profile: 5

- Quality improvement opportunity: Avoidance of unnecessary testing (National quality strategy: Making Quality Care More Affordable)
- Aggregate evidence quality: Grade B, based on cross-sectional studies and case series showing no benefit
- Level of confidence in the evidence: High
- Benefits: Cost containment, avoidance of stress and anxiety of patient, avoidance of false positives
- Risks, harms, costs: Missed diagnosis
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: Minimizing testing and the risks of false positives outweigh the value of finding a potential cause
- 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: Small
- Exceptions: None
- Policy level: Strong recommendation
- Differences of opinion: None

Supporting Text

The purpose of this statement is to discourage routine laboratory tests that do not improve management or care of patients with SSNHL 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, sometimes called ''shotgun,'' or universal testing done as a ''panel'' without consideration of specific patient or geographic risk factors. The GUG recognizes that there may be specific tests that are warranted in selected patients if pertinent history or examination 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.

While there have been some advances in the hypothesis of a common final pathway of insult for hearing loss in patients with SSNHL, there is insufficient evidence to support routine laboratory testing. 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 a lack of evidence that the result of the test would either change the management paradigm or improve outcomes.

Possible etiologies of SSNHL include viral infection, vascular compromise, autoimmune disease, inner ear pathology, and central nervous system pathophysiology, although the cause in most patients is never identified.[10] 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.[90,91] There is evidence of an association of autoimmune disease with SSNHL.[49,50,92] In the Toubi et al study, the antibody response was short-lived in most patients, which led those authors to suggest that a transient phenomenon may trigger antibody activity that produces the hearing loss.[92] In a study of 48 patients, researchers found no association between SSNHL and abnormal levels of antithrombin III, protein C, D-dimer, fibrinogen, or activated protein C resistance.[93] A prospective study to determine the prevalence of rheumatologic and immunologic disorders in patients with SSNHL in Iran looked at 83 patients and found that $<5\%$ had specific positive immunologic test results.[94] A smaller prospective case-controlled study showed lower tumor necrosis factor alpha, no elevation of anti–heat shock protein 70, and more positive antinuclear antibodies and erythrocyte sedimentation rate in SSNHL patients, but none of this correlated with corticosteroid treatment response.[95]

Another study evaluated serum and cerebrospinal fluid markers for Lyme disease in 19 patients with SSNHL.[96] While patients who received antibiotic treatment demonstrated greater recovery of their hearing loss, it was confounded by the fact that the majority of these patients with positive serology for Lyme disease also had greater recovery, making it impossible to determine if it was spontaneous recovery from Lyme disease or antibiotic treatment that improved the prognosis in this small group of patients.

SSNHL co-occurring with diabetes, hypertension, and hyperlipidemia in older patients has been associated with MRI evidence of cerebral microangiopathy and worse prognosis, but the association's clinical significance is unclear. A study of 94 patients with SSNHL showed a significant negative correlation between hearing recovery and total cholesterol levels but no other clinical or blood indices.[97] A database analysis of 400 Swedish patients with SSNHL showed that 65% had hematologic testing. Of the 300 who were designated as having ISSNHL, 24% had ≥ 1 abnormal laboratory or radiographic findings, but no significant correlation was found between these findings and treatment or hearing recovery.[98]

One study of 133 patients with SSNHL seemed to show that a low level of thyroid-stimulating hormone was a positive prognostic factor, but it did not take into account the multiple comparisons performed, and so the results lack clinical significance.[99] A case-control study showed a

relationship between low folate and SSNHL (all 44 patients had low levels), but the clinical implications of the study are not clear.[100] Other factors that have been associated with hearing loss are fatty acids, coenzyme-Q, nervonic acid, and C3b but all in small studies that have not been replicated and may not necessarily be extrapolated to the general SSNHL patient.[101,102] While SHL in children is not the focus of this guideline, a retrospective study of 136 Chinese children with SSNHL revealed varying serologic abnormalities, including white blood cell count elevation in 31%, elevated homocysteine levels in 22%, high alkaline phosphatase in 66%, high immunoglobulin E antibody in 34%, and positive cytomegalovirus in 86%. These findings did not correlate with recovery.[103]

Targeted serologic testing should be performed as indicated by clinical evaluation. For example, there is literature to suggest that SSNHL may coincide with a *Borrelia burgdorferi* infection (Lyme disease).[104] Thus, a patient traveling in a Lyme-endemic area who has a bull's-eye skin lesion and SSNHL should be tested for Lyme disease. However, that same test is not indicated in a patient with no risk of Lyme exposure or suspicious lesion who presents with SSNHL.

Another concern with nontargeted or routine laboratory testing panels is that any false-positive result may lead to further and likely unnecessary evaluation, which carries medical, psychological, and financial costs. Currently, there is insufficient evidence that any routine laboratory/serologic 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. Currently, routine laboratory testing adds cost and harm but offers no benefit and should be avoided.

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

Action Statement Profile: 6

- Quality improvement opportunity: Identify an underlying cause of the hearing loss that may have other implications and treatment recommendations (National quality strategy: Prevention and Treatment of Leading Causes of Morbidity and Mortality)
- Aggregate evidence quality: Grade B for MRI, Grade C for ABR based on observational studies and a meta-analysis
- Level of confidence in the evidence: High
- Benefits: Identify vestibular schwannoma or other tumors in the IAC or cerebellopontine angle, identify conditions that might benefit from early treatment, patient peace of mind, supporting idiopathic diagnosis
- Risks, harms, costs: Procedure specific risks/costs, anxiety, and stress
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: Although the panel agreed that 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 (with follow-up MRI if abnormal) would be an acceptable alternative for initial assessment for retrocochlear pathology in SSNHL as long as there is appropriate counseling about the limitations of this modality.
- Intentional vagueness: The time frame and the health care professional responsible for ordering tests to assess for retrocochlear pathology are not specified. The panel felt that this should happen at some point in the care of the patient and that this would most likely be ordered by the treating otolaryngologist or otologist/neurotologist.
- Role of patient preferences: Small in deciding whether or not to assess for retrocochlear pathology; large in making shared decisions with the clinician for using MRI or ABR as the diagnostic test
- Exceptions: None
- Policy level: Recommendation
- Differences of opinion: None

Supporting Text

The purpose of this statement is to ensure that clinicians look for retrocochlear pathology in patients with SSNHL 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. MRI of the brain, brainstem, and IACs with gadolinium is the most sensitive test for detecting retrocochlear pathology. Patients may opt for ABR testing. ABR is an electrical test of central auditory pathways. It is noted that ABR is called by a number of different names, including brainstem evoked response audiometry, auditory brainstem evoked response, and brainstem auditory evoked response, depending on geographic location in the United States. Sensitivity of ABR for retrocochlear lesions is low, in that ABR alone will miss an average of 20% (range, 8%-42%) of intracanalicular vestibular schwannomas.[105-107] Moreover, an abnormal ABR result will require imaging for confirmation. Contrast-enhanced temporal bone CT scans should be used only in patients who cannot have MRI. Assessing patients with SSNHL for vestibular schwannoma represents an opportunity for early identification of the tumor, affording the most

options for shared decision making, institution of therapeutic management, and potentially the best chances of preserving hearing and facial nerve function. If the patient with SSNHL is found to have a vestibular schwannoma, the management for his or her hearing loss likely includes systemic steroids but not IT steroids or HBOT, and it is no longer covered by this CPG.

Risk of Vestibular Schwannoma. Vestibular schwannoma is a benign tumor of the vestibular nerve that can lead to progressive loss of hearing, balance function, and ultimately compression of the brainstem with severe neurologic sequelae. Ten to twenty percent of patients with a vestibular schwannoma will report a sudden decrease of hearing at some point in their history,[108] but the rate of vestibular schwannoma in patients who present with SSNHL is somewhat lower. Several studies report a relatively high prevalence of cerebellopontine angle tumors in patients with SSNHL, ranging from 2.7% to 10.2% of patients who are evaluated with MRI.[9,16,66,98,109,110] A review of 291 patients with SSNHL and MRI showed 13 abnormalities felt to be causal of the SSNHL, of which 9 were vestibular schwannoma and 4 were other abnormalities. Of the 9 vestibular schwannoma cases, 3 were intracanalicular, and 6 had extrameatal extension but were still small (<1 cm) or medium sized (<2.9 cm).[111]

Evaluating patients with SSNHL for retrocochlear pathology is important because there are no clinical features early in the disease that can reliably distinguish SSNHL caused by an underlying vestibular schwannoma or other cerebellopontine angle tumor from the more common idiopathic variety.[9] 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 not universally present for their absence to reliably rule out a retrocochlear lesion. Although the likelihood of tumor presence is lower in patients with low-frequency hearing loss, all types of audiometric patterns have been found in patients with SSNHL with vestibular schwannomas.[9,110]

Associated events or diseases (eg, barotrauma or recent viral infection) that are presumed to cause SSNHL are also present in approximately one-third of patients with vestibular schwannoma. Hearing recovery has not been shown to predict whether or not a patient's SSNHL is the result of a tumor.[9,98] Accordingly, following hearing outcomes is not adequate to assess for retrocochlear pathology. SSNHL may be the presenting symptom in a variety of tumor sizes. The mean tumor size in 1 large study was 2.1 cm, with 10% of tumors >3 cm in size.[9] Therefore, all patients should be apprised of the risk of a vestibular schwannoma and counseled regarding the various diagnostic strategies and management options as part of shared decision making.

There are no RCTs comparing a strategy of investigation versus 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.[10,112,113] Many patients do well with no intervention, ''undisturbed by their tumors, ultimately dying with them but not because of them.''[10] 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[114-116] and radiotherapy[117-119] treatment. Smaller tumors are also more suitable for conservative management.[120] The conservative approach may be a particularly good option in patients with small tumors; only 20% to 25% of patients with small tumors and 33% of patients with all size tumors at presentation will experience significant tumor growth over time.[10,120,121] While surgical, radiosurgical, and conservative approaches are options for the treatment of vestibular schwannoma, no RCTs have compared these various approaches.[122] However, a large study of 642 patients demonstrated that the diagnosis of vestibular schwannoma had a greater impact on QOL than the treatment strategy, whether it be microsurgery, radiotherapy, or observation.[123] Nevertheless, optimizing QOL is essential in the management of vestibular schwannoma, indicating a high degree of shared decision making and therefore highlighting the importance of retrocochlear evaluation in patients with SSNHL.[112] The costs of screening tests for vestibular schwannoma compares favorably to the additional cost of treating larger tumors.[124] Given this advantage and the higher prevalence of tumors in patients with SSNHL, all patients with SSNHL should be evaluated for vestibular schwannoma.

The recovery of hearing to normal after an episode of SSNHL does not negate the possibility of retrocochlear pathology being present. A retrospective review of 295 patients with SSNHL found that MRI identified vestibular schwannoma in 4%, and all tumors were intrameatal or small to medium sized. A contralateral vestibular schwannoma was found in 3 patients with SSNHL and normal ipsilateral IAC. Four of the 12 cases of SSNHL and vestibular schwannoma showed good recovery after corticosteroid treatment.[125] The clinician should therefore not be dissuaded from performing a retrocochlear workup despite the presence of associated diseases, the audiometric pattern, normal vestibular findings, or hearing recovery.

Magnetic Resonance Imaging. MRI is the gold standard for imaging diagnosis of vestibular schwannoma and is more cost-effective than ABR followed by MRI.[10] The specific MRI protocol utilized will often depend on the neuroradiologic resources available. However, a dedicated MRI IAC protocol with gadolinium enhancement is extremely sensitive and is widely available. High-resolution 3-dimensional (3D) gradient echo or 3D fast spin echo sequences (heavily T2-weighted sequences) such as CISS or FIESTA of the inner ear/IACs as well as contrast-enhanced T1-weighted MRI should be included in the IAC protocol. Studies have shown that high-resolution heavily T2-weighted 3D fast spin echo or gradient echo MRI to be both sensitive in the

diagnosis of vestibular schwannoma in patients with SSNHL and more cost-effective than gadolinium-enhanced MRI.[10,126-129] The limited noncontrast, high-resolution, lower-cost T2 study can be considered an excellent alternative to the full MRI study. However, these techniques require technological and radiographic expertise that is not always available in the community.

Many authors have advocated adding 3D FLAIR sequences pre- and postgadolinium for patients presenting with SSNHL, since they have increased sensitivity to detect enhancement; however, this may be difficult to acquire in a community imaging center.[130] When possible, MRI IAC studies should be performed per protocols supervised and interpreted by a neuroradiologist given the potential subtlety of findings in SSNHL.

MRI 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 8**). The overall rate of pathogenic MRI abnormalities directly related to the SSNHL ranges from 4.4% to 13.75%.[67,93,98,111,131,132] 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) or those with financial constraints, high-resolution dedicated CT of the temporal bones with contrast may be used, although there is a significant risk that small- to medium-sized tumors could be missed by this modality.

One disadvantage of MRI is the possibility of incidental findings that are not related to the hearing loss that may result in patient anxiety or additional evaluation. In 1 study of SSNHL patients, 57% of the MRI studies revealed some abnormality, but only 11% of these findings were directly related to the hearing loss (**Table 8**).[93] In another study, while the overall rate of abnormal findings was 34.5%, only 12.5% of all patients imaged demonstrated findings directly related to the hearing loss.[132] 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.[133] Overall intracranial incidental findings on brain MRI are common, but in the majority of cases, they have no immediate medical consequences. Familiarity with, and ability to refer to neurology, neurosurgery, or other appropriate specialty for, the common incidental findings, their clinical relevance, and recommended management are required to discuss the findings adequately with the patient and to initiate further investigation only if necessary.[134] 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.[10,135] Fortunately, the latter is rare in patients without preexisting renal disease. These contrast-related risks can be avoided with a noncontrast study with only high-resolution heavily T2-weighted 3D gradient echo or fast spin echo sequences. More recently, the Food and Drug Administration (FDA) and ACR jointly with the American Society of Neuroradiology have issued warnings regarding the deposition of gadolinium-based contrast agent in the brain, even in patients with normal renal function. While no known adverse risks have been linked to this, this is an ongoing investigation.[136] Renal function and risks of contrast should be discussed with the patient before proceeding with an MRI scan in this setting.

There are not enough data to give a timeline for ordering MRI or ABR for retrocochlear workup in SSNHL. The purpose of this KAS is to ensure that within some reasonable time frame, whether or not there is hearing recovery, the patient is offered an MRI scan of the IACs/brain. This is usually ordered by the treating otolaryngologist.

Auditory Brainstem Response. ABR is a useful but imperfect tool for identification of retrocochlear pathology. ABR may miss an average of 20% (range, 8%-42%) of intracanalicular vestibular schwannoma tumors.[105-107] It may be considered adequate to initially evaluate patients in the appropriate scenario (eg, older patients in whom the missed diagnosis of a small tumor may be less consequential) but not in those in whom identification of smaller tumors is desired. ABR is highly sensitive for a vestibular schwannoma >1 cm in size and those in the cerebellopontine angle; however, ABR

Table 8. MRI Findings in SSNHL.[a]

	Cases, n (%)
MRI normal	23 of 54 (43)
MRI abnormal	31 of 54 (57)
MRI abnormality directly related to SSNHL	6 of 54 (11)
Labyrinthine hemorrhage	2
Cochlear inflammation	1
Vestibular schwannoma of IAC and CPA	1
Arachnoid cyst of the CPA	1
White matter lesions	1
Incidental MRI finding unrelated to SSNHL	8 of 54 (15)
Chiari anomaly type 1	1
Empty sella	1
Perinatal hypoxic-ischemic insult	1
Variant of temporal lobe venous drainage	1
Parietal meningioma	1
Circle of Willis aneurysm or focal arterial ectasia	3
MRI findings not directly related to SSNHL but possibly expressions of disease	17 of 54 (31)
Microvasculopathic chronic leukoencephalopathy	12
Demyelinating disease	1
Anterior inferior cerebellar artery loop in IAC	4
Artifactual findings on MRI: Artifactual semicircular canal abnormalities	3 of 54 (5.5)

Abbreviations: CPA, cerebellopontine angle; IAC, internal auditory canal; MRI, magnetic resonance imaging; SSNHL, sudden sensorineural hearing loss.

[a]Cadoni et al (*J Otolaryngol*; 2006;35[5]).[93]

testing has limits.[137] ABR is not possible when the hearing threshold exceeds 80 dB at 4000 Hz, and it may be problematic with even lesser degrees of hearing loss, depending on the hearing as well as the caliber of the equipment. The sensitivity of ABR is proportional to the degree of hearing loss; therefore, individuals with SSNHL and vestibular schwannoma with mild or recovered hearing losses will be more likely to yield false-negative ABR results.[138]

ABR evaluation may be offered for patients with SSNHL who do not wish to have MRI. There is a strong role for shared decision making and patient preference, as ABR has excellent sensitivity for medium- and large-sized tumors but may miss up to 42% of small or intracanalicular tumors. Multiple approaches and applications of ABR may be utilized for diagnostic purposes, including varied stimulus parameters to facilitate suspected site of lesion. There are some data to suggest that standard ABR may assist in prognosticating hearing outcome after SSNHL, but this is not established.[139]

Normal ABR does not rule out retrocochlear pathology. Patients electing this method must be monitored closely.[85] Given no subjective change (for which prompt evaluation is indicated), a follow-up hearing test should be performed in 6 months.[85] A significant decrease in pure tones or speech recognition threshold of >10 dB (HL) in ≥ 2 frequencies and/or a drop in $>10\%$ in WRS should trigger another retrocochlear evaluation.[85] If the patient opts for a second ABR, any abnormality—particularly, a prolongation of wave V, which often indicates retrocochlear compression of the cochlear nerve—should spur the clinician to offer imaging. MRI is preferred, and CT is offered if MRI is precluded.[85]

ABR and audiometric follow-up may be appropriate for older patients in whom aggressive treatment of a small- or medium-sized retrocochlear lesion is less likely, in patients unable to tolerate MRI, or for patients with financial or other concerns, leading them to select a less definitive evaluation strategy. It is important to keep in mind the added cost of repetitive audiograms and ABRs and compare them with the single cost of MRI when counseling the patient. The role for patient and family involvement in shared decision making is high in these cases, as they must understand that this paradigm could lead to a delay in diagnosis. As with all shared decision making and, in particular, due to the complex nature of this counseling, appropriate documentation in the patient's chart is of great importance.

STATEMENT 7. PATIENT EDUCATION: Clinicians should educate patients with SSNHL 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: 7

- Quality improvement opportunity: Improve awareness of the natural history of SHL and the myriad treatment options to improve patient involvement in shared decision making (National Quality Strategy: Health and Well-being of Communities; Effective Communication and Care Coordination)
- Aggregate evidence quality: Grade B, based on systematic reviews
- Level of confidence in the evidence: High
- Benefits: Facilitate shared decision making, increase patient adherence to proposed therapy, empower patients, informed consent, link evidence to clinical decisions
- Risks, harms, costs: Time spent, miscommunication, patients get overwhelmed, patient anxiety
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: Based on the unclear benefit of primary treatments for SHL, patients should be informed regarding the uncertainty in treatment effectiveness to make an informed treatment decision
- Intentional vagueness: None
- Role of patient preferences: Large
- Exceptions: None
- Policy level: Strong recommendation
- Differences of opinion: None

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 SSNHL. With a favorable natural history and inconclusive or modest benefit of the myriad treatment options, patients need to be engaged in the decision for what, if any, treatment to undertake. Clinicians are encouraged to provide patients with the information necessary to participate fully in shared decision making.

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. 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.[140] This discussion should be documented in the patient chart in addition to the patient's final decision. It is very important for patients to share their values, goals, and the relative importance of the potential benefit or harm associated with the various options. This process allows patients and their families/caregivers the autonomy to make difficult decisions.[141] There are 3 key elements to true shared decision making:

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

A basic protocol for management would include a discussion of

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 and goals or desired outcomes. The clinician should focus on QOL and functional health status in addition to objective treatment outcomes. Shared decision making may be limited by obstacles related to patient factors, clinician factors, and system factors. A reengineered model proposes that the clinician elicit and prioritize the patient's goals for care and then translate those goals into treatment options. Preliminary evidence suggests better patient confidence in the decision made and compliance with the treatment plan when there is sufficient time for this collaboration to take place.[140] Successful shared decision making can be accomplished with the use of various decision aids, such as pamphlets or videos, to provide information that can make health care decisions less difficult. A recent review found that the use of decision aids improved patient/caregiver 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.[142] A patient information pamphlet with frequently asked questions is available on the American Academy of Otolaryngology–Head and Neck Surgery (AAO-HNS) website (www.entnet.org). **Table 9** also presents a summary of important discussion points for the patient and clinician to consider in making treatment decisions.

STATEMENT 8. INITIAL CORTICOSTEROIDS: Clinicians may offer corticosteroids as initial therapy to patients with SSNHL within 2 weeks of symptom onset. *Option based on systematic reviews of RCTs and new RCTs and a balance of benefit and harm.*

Action Statement Profile: 8

- Quality improvement opportunity: More selective and appropriate use of a treatment option with modest potential benefit but only when used appropriately (National quality strategy: Prevention and Treatment of Leading Causes of Morbidity and Mortality; Effective Communication and Care Coordination)
- Aggregate evidence quality: Grade C, based on RCTs and systematic reviews of randomized trials downgraded for methodological limitations and again for inconsistent results
- Level of confidence in the evidence: Medium
- Benefits: Hearing improvement
- Risks, harms, costs:
  - Systemic steroids: Suppression of hypothalamic-pituitary-adrenal axis and Cushing's-like syndrome (minimal with 10- to 14-day treatment); aseptic necrosis of the hip; hyperglycemia; low cost
  - IT corticosteroids: minimal systemic effect; local reactions of pain, tympanic membrane perforation, transient dizziness; high cost and multiple office visits
- Benefits-harm assessment: Balance of benefit and harm
- Value judgments: Even a small possibility of hearing improvement makes this a reasonable treatment to offer patients, considering the profound impact on QOL that hearing improvement may offer
- Intentional vagueness: None
- Role of patient preferences: Large role for shared decision making with patients
- Exceptions: Systemic steroids: medical conditions affected by corticosteroids, such as insulin-dependent or poorly controlled diabetes, tuberculosis, peptic ulcer disease, among others
- Policy level: Option
- Differences of opinion: While all members of the GUG favored having steroids as an option as early as possible, several group members were reluctant to endorse the 2-week time frame due to concerns that patients presenting later may be denied therapy. We ultimately agreed to leave the time frame of 2 weeks to encourage patients and clinicians to seek care early if they choose to be treated.

Supporting Text

The purpose of this statement is to clarify the role of corticosteroids, a commonly employed treatment modality, in the initial treatment of SSNHL within the first 2 weeks of symptom onset. There is laboratory evidence of an inflammatory cell-death cascade in SSNHL that is modified by steroid therapy. The term ''corticosteroid'' refers to common synthetic glucocorticoids delivered via systemic (oral, IV, or intramuscular) and/or IT routes. IT steroids given as salvage therapy are covered in a different section of the guideline. The steroids discussed in this section include prednisone, prednisolone, methylprednisolone, and dexamethasone. Corticosteroids are known to have sites of action in the inner ear in viral, vascular, syphilitic, autoimmune, endolymphatic hydrops (Ménière's disease), and other etiologies of hearing loss.[8,143] Since the initial guideline publication in 2012, there have been multiple RCTs as well as systematic reviews of steroid use for SSNHL with a larger emphasis on IT steroids. The majority of these trials use a 1- to 2-week window from symptom onset as the

Table 9. Frequently Asked Questions/Patient Education.

Question	Answer
What is causing the problem?	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 ringing (tinnitus), dizziness (vertigo), and fullness in the ear.
How is sudden hearing loss diagnosed?	The sudden change in hearing is obvious to you. It may be accompanied by loud ringing, vertigo (spinning sensation or balance problems), and/or pressure in the ear and should be evaluated as quickly as possible. Your health care provider will take a comprehensive history and complete a physical exam. Routine labs and x-rays are not recommended, but a hearing test (audiogram) should be done.
Will my hearing come back?	Approximately one-third to two-thirds of patients with SSNHL may recover some percentage of their hearing within 2 weeks. Those who recover half of their hearing in the first 2 weeks have a better prognosis. Patients with minimal change within the first 2 weeks are unlikely to show significant recovery. Additionally, patients with dizziness at the time of onset of SSNHL have a poorer prognosis.
Is there additional testing needed with SSNHL?	SSNHL can rarely be associated with benign tumors of the vestibular nerve. These tumors are called vestibular schwannomas and can lead to progressive hearing loss, balance problems, and in some cases compression of the brainstem with severe neurologic symptoms. Your provider may order magnetic resonance imaging (MRI) to screen for these tumors. While MRI of the brain and internal auditory canals is the most sensitive test, some patients opt for an ABR (auditory brainstem response). This is a less sensitive screening tool but is less expensive and does not require being in the confined space of the MRI machine. If the ABR is abnormal, good practice requires an MRI.
How is sudden hearing loss treated?	Many treatments have been proposed for SSNHL. 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. Based on current research, clinicians may offer corticosteroids as initial therapy. This is most commonly given in pill form but can be done with an injection through the eardrum (intratympanic) for those patients that oral steroids are contraindicated. Although antivirals are commonly prescribed, there is insufficient evidence to support their effectiveness in treating sudden hearing loss. Hyperbaric oxygen may also be offered within 2 weeks of the initial diagnosis of SSNHL or up to 1 month in conjunction with steroids. Clinicians should offer salvage therapy (usually intratympanic steroids) for incomplete recovery after initial therapy. The benefits of therapy may include more prompt and complete recovery of hearing, but there are also side effects that must be considered when choosing among the available options.
What are the side effects of each treatment?	Side effects vary with each treatment modality but may include increased anxiety, pain, dizziness, elevated blood sugar, elevated blood pressure, depression, or insomnia. You should have a conversation with your provider regarding the specific side effects associated with your treatment.
What else can I expect?	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 SSNHL. 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. Clinicians should obtain follow up audiometry within 6 months of initial diagnosis of SSNHL.

enrollment criteria to determine steroid effectiveness as initial treatment.[144-151] However, the protocols for these trials differed greatly, making interpretation of outcomes difficult.

Systemic Steroids vs Placebo. A Cochrane review, first published in 2006 and most recently updated in 2013, found only 3 trials that met their inclusion criteria of RCTs of steroids versus placebo or no treatment.[152] All looked at oral steroids versus placebo, and all were found to have high risk of bias. Two of the included trials demonstrated no significant benefit between steroids and placebo, while 1 trial showed improvement of hearing in 61% of patients in the steroid group as compared with 32% in the control group.[10,148,153] Because of the contradictory outcomes and small number of patients in the 3 trials reviewed, the role of steroids ''[remained] unclear.''[152] Another systematic review also noted that there was no statistically significant treatment effect with these same trials.[154]

In 2016 another group compared the addition of IV methylprednisolone to oral steroids and noted no difference in hearing outcomes with the addition of IV steroids.[144]

IT Steroids vs Placebo. Only 1 RCT to date has focused on the efficacy of IT steroid versus placebo as initial therapy for

patients with SHL. Filipo and colleagues looked at 50 patients with moderate hearing loss and randomized them into 2 groups, with the first receiving 3 days of daily IT prednisolone and a control group receiving daily IT saline. Both groups were given oral prednisone if there was incomplete recovery at 7 days.[145] In this study, 76% of patients receiving IT steroids demonstrated complete recovery, as opposed to 20% of the control group (P = .0002). Giving additional oral steroids to those in the IT group who failed to improve did not change their long-term outcome when compared with those in the placebo group, who eventually reached 72% complete recovery after a course of oral steroids. While IT steroids conferred early benefit of hearing recovery versus placebo, the study could not prove long-term superiority over systemic steroids.[145]

Systemic vs IT Steroids as Initial Therapy. The majority of the RCTs that studied the use of steroids as initial therapy for SHL compared systemic steroids with IT steroids alone or in combination with the systemic steroids. In their systematic review, Crane and colleagues also looked at the efficacy of systemic steroids against IT steroids as initial therapy and showed no overall advantage of IT steroids over systemic steroids.[154] In the 6 trials included, only Battaglia and colleagues demonstrated a significant treatment effect of the use of IT steroids, either alone or combined with systemic steroids, when compared with systemic steroids alone.[147,150,154-159] The remainder showed that systemic steroids versus IT steroids, often in combination with systemic steroids, led to similar hearing outcomes. Garavello and colleagues, using similar studies, came to the same conclusions in their meta-analysis.[10,150,155,156,159,160] The largest RCT to date comparing systemic and IT steroids was a 16-center study that enrolled 250 patients and showed that hearing at 2 months did not differ between patients who received prednisone (60 mg/d) for 14 days and those who received 4 doses of IT methylprednisolone (40 mg/mL) over 14 days.[150] Swachia and colleagues also found no difference between systemic steroids and IT steroids.[151]

A systematic review published in 2017 by Han and colleagues[161] suggests that combination therapy may offer improved hearing, with an odds ratio of 2.5 (95% CI, 1.95-2.1) with a mean difference of 13 dB (95% CI, 9.24-16.77). Their review included some more recent RCTs not included in prior systematic reviews. The use of early IT steroids in the Battaglia et al study suggests that in combination with oral steroids, IT steroid use within 7 days of onset of sudden hearing was associated with a 20-dB PTA and 30% speech discrimination score improvement relative to those treated with IT steroids in combination with systemic steroids after 7 days.[162] However, a meta-analysis with mathematical simulations of the various IT protocols used in these studies as well as others showed that the trend of early treatment having a positive effect on hearing is likely a ''sham effect'' due to spontaneous recovery.[163] The mathematical model accounted for the variability in drug used, concentration, application time, number of injections, frequency of injections, and duration of treatment in addition to patient's pre- and posttreatment PTAs. The authors concluded that the final hearing did not depend on the drug given, dose used, dosing frequency, or duration of treatment but rather on the severity of the hearing loss.[163]

Benefits, Risks, and Dosing of Systemic Corticosteroid Therapy for Individual Patients. Given the studies cited here, the clinician might choose not to prescribe corticosteroids for SSNHL. However, when 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 any data showing efficacy, although even those data are somewhat equivocal.[3,10,13,148,153,164-168] While the greatest spontaneous improvement in hearing occurs during the first 2 weeks,[4] 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.[3,12,39,99,168-172] More recent studies encourage treatment within 7 days of onset.[150,162]

For optimal treatment outcomes, the recommended dose of oral prednisone is 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.[150,168] Data comparing treatment protocols are limited. Representative regimens include (1) the maximum daily dose for 4 days, followed by a 10-mg taper every 2 days; (2) the more commonly employed maximum daily dose for 7 to 10 days, followed by a taper over the next week; and (3) the maximum daily dose for 4 weeks followed by a taper. The equivalent dose of prednisone (60 mg) is 48 mg for methylprednisolone and 10 mg for dexamethasone. Underdosage, by the aforementioned standards, is a possibility if attention is not given to these ratios. As noted, early treatment is important, so the clinician should ensure that the patient is initially adequately dosed, whether the steroid is given orally or intravenously. The methylprednisolone and prednisone dose packs do not provide adequate doses of steroids for treatment and should be avoided, both for underdosing and for delaying potentially effective intervention. General guidelines for systemic corticosteroid treatment are summarized in **Table 10**.

Potential side effects of systemic corticosteroid therapy are reported to affect many organ systems. Common side effects of oral steroids include acne, blurred vision, cataracts or glaucoma, easy bruising, insomnia, hypertension, increased appetite, weight gain, increased growth of body hair, lower resistance to infection, muscle weakness, nervousness/restlessness, osteoporosis, gastric irritation, mood swings, facial swelling. fluid retention, and worsening of diabetes.[173]

Alexander et al reviewed the safety of high-dose steroids taken for up to 22 weeks for autoimmune inner ear disease and found that the majority of patients completed the course and that the most frequent adverse events were hyperglycemia and weight gain.[10] 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.[174]

Table 10. General Guidelines for Corticosteroid Therapy for SSNHL.[a]

	Systemic Corticosteroids	Intratympanic Corticosteroids
Timing of treatment	Immediate, ideally within first 14 days. Benefit has been reported up to 6 weeks postonset of SSNHL.	1. Immediate 2. Salvage (rescue) after initial treatment fails or after 2 weeks of symptom onset
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 (compounded) or 10 mg/mL (stock) if compounded concentration unavailable 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 up to 4 injections over a 2-week period
Technique	Do not divide doses	1. Fill the middle ear with steroid solution 2. Keep head in otologic position (one side down, affected ear up) for 15-30 minutes
Monitoring	Audiogram at completion of treatment course and at delayed intervals	Audiogram at completion of treatment course and at delayed intervals. Interval audiograms between injections may help direct early termination of therapy if hearing loss resolves. Inspect tympanic membrane 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 tympanic membrane perforation.

Abbreviation: SSNHL, sudden sensorineural hearing loss.

[a]This table is designed to provide guidance for systemic and intratympanic steroid treatment for SSNHL and is based on the literature as well as expert input from the members of the guideline update group. Treatment is routinely individualized by provider and per patient. The most important principles pertain to early institution of high-enough dosages of treatment.

Patients with certain systemic medical conditions, such as insulin-dependent or poorly controlled diabetes, labile hypertension, glaucoma, tuberculosis, peptic ulcer disease, and prior psychiatric reactions to corticosteroids, among others, may not be able to receive systemic corticosteroids. However, the data are clear that if they receive IT steroid injections, their treatment will not be inferior in terms of efficacy.

The lack of clear evidence supporting systemic steroids for SHL and the existence of potential adverse treatment effects support a large role for shared decision making with patients.[175] Most serious side effects, however, occur with chronic use, and adverse events are rare and manageable for the short 10- to 14-day course of steroids recommended for SSNHL.

Benefits, Risks, and Dosing of IT Corticosteroid Therapy for Individual Patients. Since the publication of the initial recommendations in 2012, IT steroid use has gained popularity, especially in those who cannot receive systemic steroids.[10,176,177] Parnes et al 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.[172] Subsequent laboratory data have substantiated the claim of higher perilymph steroid concentrations after IT steroid application when compared with systemic steroid use, in addition to demonstrating the added utility of round window membrane transport facilitators histamine and hyaluronic acid.[178] However, similar to those for systemic steroids, the data supporting the use of IT steroids as initial therapy for SHL are equivocal, as discussed before.

The IT steroids administered are either dexamethasone or methylprednisolone.[172] Corticosteroid concentrations vary widely among studies; the majority of papers on IT corticosteroids refer to dexamethasone (4-24 mg/mL) and methylprednisolone ($\geq$30 mg/mL).[13,179] Higher concentrations appear to have better outcomes, although only 1 study comparing concentration outcomes has been published to date. A small retrospective study of 37 patients compared outcomes between IT dexamethasone concentrations of 24 mg/mL and 10 mg/mL and noted that a larger percentage of patients (53% vs 17%) had >30-dB improvement in PTA (P = .0382) with a trend toward improved WRS outcome for the higher steroid concentration.[180] 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, and possible vasovagal or syncopal episode during injection. In addition, cost and the need for multiple office visits must be taken into consideration.

Frequency of IT steroid administration also varies widely among studies, from self-administration by the patient across a pressure-equalizing tube several times per day to physician administration from once daily to once weekly or less.[13,179] Due to heterogeneity in concentration, frequency of dosing, and combination with systemic steroids, studies on IT steroids as initial therapy are difficult to assess. Nevertheless, all studies were in agreement that the IT steroids should be instilled in the affected ear for 15 to 30 minutes per injection.[145-147,149-151] However, the data suggest that they may be a reasonable alternative to systemic therapy. General guidelines for IT steroid treatment are summarized in **Table 10**.

Harm vs Benefit of Corticosteroid Therapy. Despite the uncertain balance of benefit versus harm for steroid therapy based on existing RCTs, there is also insufficient evidence to conclude the treatment is ineffective. Moreover, there is a large volume of observational studies and RCTs that suggest a treatment benefit, although to what degree this exceeds spontaneous resolution is not known.[13,179,181] Considering the devastation of SSNHL and the profound impact on QOL that a hearing improvement may offer, the GUG concludes that even a small possibility of hearing improvement makes this a reasonable treatment to offer to patients.

STATEMENT 9a. INITIAL THERAPY WITH HYPERBARIC OXYGEN THERAPY: Clinicians may offer, or refer to a clinician who can offer, hyperbaric oxygen therapy (HBOT) combined with steroid therapy within 2 weeks of onset of SSNHL. *Option based on systematic reviews of RCTs with a balance between benefit and harm.*

STATEMENT 9b. SALVAGE THERAPY WITH HYPERBARIC OXYGEN THERAPY: Clinicians may offer, or refer to a clinician who can offer, hyperbaric oxygen therapy (HBOT) combined with steroid therapy as salvage within 1 month of onset of SSNHL. *Option based on systematic reviews of RCTs and new RCTs with a balance of benefit and harm.*

Action Statement Profile: 9

- Quality improvement opportunity: To allow the use of HBOT, which may have some limited benefit early after SHL as a potential option for primary or salvage therapy (National quality strategy: Prevention and Treatment of Leading Causes of Morbidity and Mortality; Effective Communication and Care Coordination; Patient Safety)
- Aggregate evidence quality: Grade B, based on systematic review of RCTs with methodological limitations and new RCTs with limitations
- Level of confidence in the evidence: Medium
- Benefits: Hearing improvement
- Risks, harms, costs: Costs, patient time/effort, patient anxiety and stress, hyperbaric-associated complications such as barotrauma, oxygen toxicity, worsening of cataracts, fatigue, seizure, and death
- Benefits-harm assessment: Balance of benefit and harm
- Value judgments: Although HBOT may not be readily available in all regions, the panel felt that the level of evidence for hearing improvement, albeit modest and imprecise, was sufficient to include HBOT as an option for patients with SSNHL
- Intentional vagueness: None
- Role of patient preferences: Large role for shared decision making
- Exceptions: None
- Policy level: Option
- Differences of opinion: None

Supporting Text

The purpose of these statements is to increase awareness of the potential role of HBOT to treat sudden SNHL. While HBOT is not commonly used as therapy for SSNHL in the United States and is not currently FDA approved for this indication, there have been multiple RCTs and a Cochrane review performed on this topic that indicate some potential benefit. Vascular compromise, and associated cochlear ischemia, is a potential etiology of SSNHL in some cases and may be part of the final common pathway to hearing loss. HBOT exposes a patient to 100% oxygen at a pressure level >1 atmosphere absolute in a specially designed chamber. Effective therapeutic levels are typically between 1.5 to 2.0 atmosphere absolute. The increased partial pressure of oxygen allows for more delivery of oxygen to the tissues—in this case, the cochlea, which is very sensitive to ischemia. Furthermore, HBOT is thought to have complex positive effects on immunity, oxygen transport, and hemodynamics, reducing hypoxia and edema and potentiating normal host responses to infection and ischemia.[182]

The Underseas and Hyperbaric Medical Society approved HBOT for the treatment of idiopathic SNHL on October 8, 2011.[183] Their recommendation was to use HBOT within 14 days of symptom onset. HBOT was also recommended during the Tenth European Conference on Hyperbaric Medicine.[184]

Their consensus recommendation was for use of HBOT combined with medical therapy in patients with acute SSHNL who present within 2 weeks of disease onset (type 1 recommendation, level B evidence). Beyond 6 months, there was no role for HBOT (type 1 recommendation, level

C evidence). Between 2 and 4 weeks, HBOT was a potential adjunct to corticosteroids in patients with SSNHL, particularly in patients with severe and profound hearing loss (type 3 recommendation, level C evidence).

HBOT has been used, typically as an adjunctive treatment, for SSNHL. The most recent Cochrane review on this topic[185] reports that HBOT was first used to treat SHL in the late 1960s in France and Germany. 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. Most studies lacked a control group, and few studies employed HBOT without concurrent or prior medical therapy.

HBOT as Primary Therapy. The Cochrane review, updated in 2012, included 7 RCTs published between 1985 and 2004.[10,185,186] No newer trials were included in its review. It reported no significant benefit of HBOT, using a 50% improvement in hearing as the primary outcome. It did find a significant benefit when the primary outcome was 25% improvement in hearing. The number needed to treat for this outcome was 5. This is an atypical outcome measure, and the clinical significance of this difference is not well established.

The small total number of subjects in the pooled group (n = 392) precluded extensive subgroup analysis; however, there was an apparent association between response to HBOT and the severity of hearing loss on presentation. Patients with moderate to severe hearing losses improved more than those with mild losses. Results were better if HBOT was performed within 2 weeks of acute onset. The ultimate conclusion, however, was that larger randomized trials of high methodological rigor were needed to define the true benefit from HBOT.

A more recent retrospective study of 59 patients treated with HBOT showed that there was no difference in hearing recovery in patients whose treatment was initiated within the first week of symptom onset (hearing gain, 23.55 dB) versus the second (hearing gain, 22.92 dB), but there was a statistically significant decrease in recovery if HBOT was initiated between 2 and 4 weeks of symptom onset (hearing gain, 5 dB).[187]

Most prospective trials compare HBOT with medical therapy and medical therapy alone. One RCT, not included in the Cochrane review, treated all patients with prednisolone and randomized them to receive additional HBOT (n = 36) or not (n = 21).[10] There was no significant difference between groups (79% HBOT vs 71% controls) based on the outcome of complete recovery (>50-dB improvement) or moderate recovery (10- to 50-dB improvement).

The systematic review for this guideline identified 2 other prospective trials of HBOT for initial treatment of SHL within 15 days of onset. In those trials, all patients got HBOT, and only the steroid treatments were varied. The first study compared IT and IV steroids, with all 48 patients getting the same HBOT.[188] Patients were also assessed according to degree of hearing loss (severe, >70 to 90 dB; profound, >90 dB). Despite a large difference in recovery rates between IT and IV steroids in patients with severe loss (83% vs 53%), there was no statistically significant difference (P = .202), due to a small sample size. Similarly, there was no difference in patients with profound SSNHL (60% IT vs 53% IV).

The other prospective randomized trial included 58 patients. All received HBOT. The first arm (n = 20) was also treated with systemic steroids. The other arm (n = 38) was treated with IV and IT steroids. Both treatment arms showed significant improvements in hearing, but there was no significant difference between the treatment arms (55% in group 1 vs 63% in group 2).[189]

HBOT as Salvage Therapy. HBOT has also been studied as salvage therapy for those not responding to other primary treatments. Several older trials found significant improvement in hearing when HBOT was combined with steroids in patients who did not respond to primary therapy (IV steroid or antiviral medications).[10,190,191]

More recent trials compared HBOT and IT therapy alone as salvage. Alimoglu et al had 4 treatment protocols for SSNHL: oral steroids, IT steroids, HBOT only, and HBOT combined with oral steroids.[192] Full hearing recovery occurred in 42.6% of the patients treated with combined HBOT and steroids, as opposed to 19.0% (oral steroids), 17.5% (HBOT), and 11.6% (IT steroids). Mathur observed that 50% of their patients improved in the 2- to 6-week time frame by 20 dB. Delay in treatment of >3 months resulted in minimal improvement of <5 dB.[193] Cvorovic et al prospectively randomized 50 patients who failed primary treatment (<10-dB hearing gain) to receive either IT steroids or HBOT.[194] Both treatments were associated with significant hearing improvements, but there were no significant differences between treatment arms. Overall, patients with lesser degrees of hearing loss (<81-dB PTA) and age <60 years improved more than those with profound deafness and older age. Early treatment (<4 weeks from symptom onset) was also associated with better outcomes. Slightly less positive findings were noted in a retrospective study of 57 patients with <20-dB improvement after primary therapy who were treated with IT steroids (n = 30) or HBOT (n = 27).[195] While there were trends toward improvement, there were no significant differences in hearing changes from baseline in both treatment arms.

Risks and Costs of HBOT. 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, as well as 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 difficultly equalizing pressure in the middle ear, which occurred in 17% of patients.[196] Another study found that 45% of patients undergoing HBOT for a variety of indications had eustachian tube

dysfunction.[197] In a study of 80 patients undergoing HBOT for SSNHL, these complications were less common, with only 5 patients (6.25%) experiencing ear or sinus barotrauma.[198] In addition, patients may suffer from some degree of confinement anxiety while undergoing HBOT.[185,196,198]

Finally, HBOT is a costly and time-consuming intervention. Therapy typically involves 10 to 20 one- to 2-hour sessions over days to weeks. While costs may vary considerably among facilities, queries by the GUG showed that typical fees in academic institutions are approximately $600 to $700 per session, including both technical and professional fees. Insurance coverage of HBOT for SSNHL indication is sporadic at best.

Given the small number of patients in the trials reviewed, methodological shortcomings, and poor reporting, there remains uncertainty regarding the real benefit of HBOT for SSNHL. There is substantial cost, potential adverse effects, uncertainty regarding the clinical significance of the hearing improvement in treated patients, and the confounding effect of concurrent steroid therapy. With this balance of benefits and potential harms and cost, the GUG could not recommend HBOT but reserves it as an option when combined with steroid therapy in SSNHL as primary therapy within 2 weeks of onset of symptoms and as salvage therapy when used within 4 weeks of onset, with potentially more benefit noted in cases of severe to profound loss.

STATEMENT 10. IT STEROIDS FOR SALVAGE THERAPY: Clinicians should offer, or refer to a clinician who can offer, IT steroid therapy when patients have incomplete recovery from SSNHL 2 to 6 weeks after onset of symptoms. *Recommendation based on systematic reviews of RCTs with a preponderance of benefit over harm.*

Action Statement Profile: 10

- Quality improvement opportunity: Encouraging the use of IT steroids, which may be effective to provide additional hearing recovery in patients with an incomplete response to initial therapy (National quality strategy: Prevention and Treatment of Leading Causes of Morbidity and Mortality)
- Aggregate evidence quality: Grade B, based on RCTs with limitations and systematic reviews of RCTs with limitations
- Level of confidence in the evidence: High
- Benefits: Hearing recovery
- Risks, harms, costs: Perforation, discomfort, cost, patient anxiety
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: Patients qualifying for salvage therapy have had an incomplete recovery of hearing after 2 weeks from onset regardless of initial therapy. Incomplete recovery 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 GUG 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: Large role for shared decision making
- Exceptions: None
- Policy level: Recommendation
- Differences of opinion: None

Supporting Text

The purpose of this statement is to encourage the use of IT steroids, either alone or in combination with oral steroids or HBOT, as salvage therapy for patients with incomplete hearing recovery following any initial management for SSNHL. As in the previous CPG, ''salvage'' refers to failure of any initial ''treatment,'' which includes systemic or topical steroids, HBOT, and observation. This recommendation is distinct from the earlier KAS (KAS 8) regarding IT steroid therapy in the context of initial management only.

Although a clear definition for failure of initial therapy does not exist, a number of patients with SSNHL fail to respond, either completely or partially, to initial treatment. For patients who fail to recover spontaneously or after initial systemic therapy, the data do not support systemic steroid therapy (oral or IV) as salvage therapy. However, IT delivery of steroids has been proposed as an option to obtain additional hearing recovery. Since publication of the original CPGs, additional research investigating the use of IT steroids as salvage treatment for SSNHL has been published, including 5 RCTs,[149,199-202] 5 meta-analyses,[154,160,203-205] and 1 systematic review.[181] While these studies suffer from considerable design flaws and differences in experimental methods, the majority do show improved hearing outcomes after IT steroid therapy.

Similar to the concept of systemic steroids for SSNHL, IT steroid therapy aims to reduce inflammation in the inner ear that may be contributing to or preventing recovery from hearing loss. An alternative theory proposes that steroids may help inhibit or reverse the apoptotic pathway of the injured cochlear hair cells.[206] 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 an IT route, alone or with a round window membrane facilitator, as compared with systemic administration.[172,178]

While there are no data to support an absolute time window after which initiation of salvage therapy with IT steroids loses effectiveness for the treatment of SSNHL, 4 of the 5 RCTs (summarized in **Table 11**) began administering IT steroids within 7 days of completion with systemic treatment.[149,199,201,202] Li et al did not specify an exact time

Table 11. Summary of the Randomized Controlled Trials Evaluating IT Steroids Therapy as Salvage for SSNHL.

Study (Subjects)	Initiation of IT Salvage Therapy	Dose/Method of Injection	Definition of Improvement	% Improvement
Lee et al 2011 (n = 46)[199]	Within 2 days after systemic treatment	5 mg/mL of dexamethasone; 4 injections over 2 weeks	≥10-dB improvement in PTA	47.6% IT group vs 16% control group
Li et al 2011 (n = 65)[200]	Timing after systemic treatment not defined	40 mg of methylprednisolone in 1 mL of sodium bicarbonate; injection every 3 days for 4 injections	≥10-dB improvement in PTA	37.5% IT group vs 0% control group
Park et al 2011 (n = 88)[149]	Within 7 days after systemic treatment	5 mg/mL of dexamethasone; 6 injections over 2 weeks	Siegel's criteria (complete, partial, slight, no recovery)[a]	No significant differences between simultaneous and salvage groups for any measure
Wu et al 2011 (n = 60)[201]	Within 7 days after systemic treatment	4 mg/mL of dexamethasone; 4 injections over 2 weeks	≥10-dB improvement in PTA	44.4% IT group vs 10.7% control group
Zhou et al 2011 (n = 76)[202]	Within 7 days after systemic treatment	40 mg of methylprednisolone in 1 mL of sodium bicarbonate; injection every other day for 4 injections	≥15-dB improvement in PTA and/or ≥15% improvement in WRS	45.9% ≥15-dB PTA improvement (20.5% control); 43.2% ≥15% WRS improvement (17.9% control)

Abbreviations: IT, intratympanic; PTA, pure tone average; WRS, word recognition score.
[a]Siegel's criteria: complete recovery (PTA <25 dB), partial recovery (>15-dB PTA improvement and PTA between 25 and 45 dB), slight recovery (>15-dB PTA improvement and PTA <45 dB), no recovery (<15-dB PTA improvement and PTA >75 dB).

between completion of systemic treatment to initiation of IT salvage.[200] Several older studies report longer durations (up to 3 months,[207] <40 days[13]) before initiation of IT steroid salvage therapy.

IT steroids are delivered to the middle ear and then absorbed via diffusion through the round window membrane into the inner ear. However, existing studies show considerable variability in the concentration of IT steroid (4 mg/mL, 10 mg/mL, 24 mg/mL for dexamethasone and 30 mg/mL to 40 mg/mL or more of methylprednisolone) as well as the timing, frequency, total number of injections, and drug selection (dexamethasone vs methylprednisolone).[10] Only 1 meta-analysis has shown a significant difference in outcomes between dexamethasone and methylprednisolone, with dexamethasone yielding significantly better outcomes.[205] There is no consensus as to how IT steroids should be delivered into the middle ear. Steroids may be delivered via needle perforation or via myringotomy (incision in the ear drum) with or without placement of a tympanostomy tube. There are other drug carrier systems that have been described for IT steroid application, including microcatheters,[10,208] MicroWick,[209] hydrogel applications,[210] and nanoparticles. Transtympanic needle perforation and myringotomy with tympanostomy tube are the most frequently utilized.[143] Only 1 meta-analysis has shown improvement in outcomes with IT injection as compared with round window infusion catheter, and that study included 5 very heterogeneous IT injection studies and only 1 round window catheter study.[205]

While no direct comparisons exist evaluating hearing outcomes as related to the number and timing of IT injections performed for IT salvage therapy, all 5 RCTs referenced herein used a paradigm of at least 4 injections over 2 weeks (1 study had 6 injections over 2 weeks), and all 5 used a needle perforation technique with IT injections. The concentration of these steroids for these RCTs is low. As previously noted, Haynes et al[13] demonstrated noninferiority of a single 24-mg/mL dexamethasone injection as compared with multiple dexamethasone injections at multiple intervals. Of note, the experts on IT injection in the GUG indicated that they perform injections once per week for a maximum of 3 or 4 injections, stopping either when hearing recovers or after a fourth injection. Four of the 5 RCTs evaluating IT steroids as salvage therapy found that IT steroids improved hearing outcomes significantly more than control. Hearing improvement occurred in 37% to 48% of patients receiving salvage IT steroids.[199,200] The other RCT compared simultaneous systemic steroids plus IT steroid therapy and IT salvage therapy alone.[149] Using Siegel's criteria to quantify hearing improvement (complete recovery: final PTA better than 25 dB; partial recovery: >15-dB gain and final PTA of 25-45 dB; slight recovery: >15-dB gain and final PTA worse than 45 dB), this study showed no significant difference between groups for any subcategory of hearing improvement. Additionally, when complete and partial recovery groups were combined, there was again no significant difference between groups. All 6 of the meta-analyses and systemic reviews demonstrated a significant effect of IT steroid therapy as salvage treatment for SSNHL. Despite the paucity of well-executed trials, the majority of studies of IT steroids as salvage treatment demonstrate a significant benefit of therapy. A limited

meta-analysis of the higher-quality studies revealed a mean difference in improvement of 13.3 dB in the IT salvage group versus placebo (95% CI, 7.7-18.9; $P < .0001$).[181]

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 95%.[10,13,176,211-215] One critical problem in these trials is that the definition of hearing improvement following IT steroid therapy varies among studies. Some studies use PTA improvements, while others use percentage change in WRS. Furthermore, depending on the initial degree of hearing loss, statistically significant improvements in PTA or WRSs may not equate to a clinically significant improvement in functional hearing.

There is only 1 study in the literature specifically comparing IT steroids alone as salvage therapy and combination salvage therapy with HBOT and IT steroids. Yang et al, in a historical cohort study, demonstrated a mean PTA improvement of 22.5 dB in the IT + HBOT group as compared with 18.9 dB in the IT-only group.[216] Additionally, 68.4% of patients in the combination salvage group had ≥15-dB PTA improvement, as opposed to 48.6% in the IT-only group. These differences were not statistically significant.

There is also 1 study comparing salvage therapy with systemic steroids and salvage therapy with IT steroids. Moon et al, in a retrospective study, demonstrated an overall rate of hearing improvement based on Siegel's criteria in 48.5% of subjects receiving salvage IT steroids.[217] This was significantly higher than improvement seen in the systemic steroid salvage group (15.4%) and placebo group (16.9%). If Siegel's criteria type I and II are used to categorize hearing recovery as ''favorable,'' 24.2% of subjects receiving IT salvage therapy showed favorable improvement, as opposed to 11.9% in the systemic steroid salvage group and 3.8% in the placebo group. There are no studies specifically comparing combined salvage therapy with systemic and IT steroids versus IT steroid therapy alone. Studies specifically comparing salvage therapy with HBOT alone and IT steroids are summarized in KAS 9.

Despite the limitations of the existing research, the majority of studies evaluating IT steroids as salvage therapy for SSNHL, including nonrandomized prospective trials, retrospective trials, and RCTs, demonstrate additional hearing improvements beyond those resulting from initial treatment with systemic steroids. Since salvage IT steroid therapy has been found to be beneficial, treatment is recommended for those who have persistent hearing loss despite conventional treatment with systemic steroids or steroids + HBOT or observation. The decision to perform salvage IT steroid therapy should be made in a shared fashion between the clinician and patient and based on the amount of persistent hearing loss following initial therapy, patient preference, as well as the risks and benefits of the treatment itself.

STATEMENT 11. OTHER PHARMACOLOGIC THERAPY: Clinicians should not routinely prescribe antivirals, thrombolytics, vasodilators, or vasoactive substances to patients with SSNHL. *Strong recommendation against based on systematic reviews of RCTs with a preponderance of harm over benefit.*

Action Statement Profile: 11

- Quality improvement opportunity: Avoidance of ineffective treatment(s) and associated risks, complications, side effects, costs, and potential adverse interactions with effective therapies (National quality strategy: Patient Safety; Prevention and Treatment of Leading Causes of Morbidity and Mortality)
- Aggregate evidence quality: Grade B, based on systematic reviews of RCTs
- Level of confidence in the evidence: High
- Benefits: Avoidance of unnecessary treatment, avoid adverse events of unnecessary treatment, cost saving
- Risks, harms, costs: None
- Benefits-harm assessment: Preponderance of harm over benefit
- Value judgments: None
- Intentional vagueness: The word ''routine'' is used to avoid setting a legal standard recognizing that there may be patient-specific indications for ≥1 of these therapies that may be reasonable to try on an individualized basis, with shared decision making
- Role of patient preferences: None
- Exceptions: None
- Policy level: Strong recommendation
- Differences of opinion: None

Supporting Text

The purpose of this statement is to discourage clinicians from routinely using pharmacologic agents that have potential side effects and no documented efficacy in SSNHL. This does not preclude the use of pharmacotherapy for known conditions that cause SHL, such as for Ménière's disease or Ramsay Hunt syndrome. 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.[218] Theoretically, initiation of antiviral agents may be valuable for aiding in the recovery of hearing. Since 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. In 2007, Conlin and Parnes published both a systematic review and a meta-analysis of treatments for SSNHL and found 4 RCTs comparing antiviral therapy and steroid therapy versus placebo and steroid therapy.[10] None of the studies reported

statistically significant benefit from antiviral therapy. 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. Blood supply to the inner ear is tenuous as there is no collateral circulation. As with most vascular disorders, hemorrhage, embolism, and vasospasm may affect the inner ear negatively and cause damage resulting in SSNHL. Fisch et al 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%.[219] Hypercoagulability, which is associated with a number of medical conditions, has also been seen in blood samples of patients with SSNHL. There remains contradictory histopathological and clinical evidence against the vascular theory of SSNHL.[10,220-222]

Despite this, vasoactive and rheologic agents have been tried in the management of SSNHL. These include prostaglandin E_1, naftidrofuryl, calcium antagonists, *Ginkgo biloba*, pentoxifylline, dextran, defibrinogenation therapy, and aspirin. The use of vasodilators and vasoactive substances for SSNHL was reviewed by the Cochrane Collaborative in 2009.[10] Only 3 RCT studies met inclusion criteria. 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. Due to the degree of heterogeneity in methodology and outcomes assessment, the results could not be combined to reach a conclusion of efficacy. Others found no clinically significant benefit of rheologic agents or defibrinogenation therapy.[10]

A systematic review of vasodilators for SSNHL showed no benefit of vasodilators alone but a possible benefit of vasodilators when administered with steroids. A meta-analysis of interventions in the management of blood viscosity in the management of SSNHL that included 49 papers of uncertain quality, the majority of which were published in non–English language journals, found there to be evidence that fibrinolytic therapies offer benefit, while there was a suggestion that other therapies, including hemodilution, anticoagulation, and rheophoresis, may be helpful but need further study.[223] Vasoactive therapies for SSNHL, in addition to being unproven in efficacy, may pose meaningful side effects, including allergic reactions, bleeding, hypotension, arrhythmias, seizures, circulatory collapse, and drug interactions.

Research looking for other treatments for SSNHL has shown potential promise. One of the agents is AM-111, which is not commercially available as of this writing. It is a 31–amino acid cell-permeable peptide formulated in a hyaluronic acid gel for IT injection, which blocks the JNK protein kinase. In small phase 1 trials, AM-111 has been found to be otoprotective in various models of cochlear insult, including acute noise trauma, acute labyrinthitis, aminoglycoside ototoxicity, bacterial infection, cochlear ischemia, and cochlear implantation trauma. A double-blind randomized placebo-controlled phase 2 study of 210 patients documented a statistically significant and clinically relevant effect of AM-111 in the treatment of profound autoimmune SNHL: improvement in PTA and speech discrimination score was more rapid and more profound than for placebo, and complete hearing recovery and tinnitus remission were more frequent than placebo.[224] These results compare favorably with recent well-designed trials for the treatment of SSNHL with prednisolone or methylprednisolone, while AM-111 appears to have none of the potential risks, complications, limitations, and side effects of oral and/or IT corticosteroids.[224]

In addition to the therapies discussed here, there is a host of other therapies that have been proposed in the treatment of SSNHL, such as vitamins (eg, high-dose vitamin C),[225] minerals (eg, zinc),[226] supplements (eg, N-acetyl-cysteine, alpha lipoic acid, CoQ10), alternative medications (eg, Chinese herbal medications, *Ginkgo biloba*),[227,228] and complementary therapies (eg, acupuncture).[229] There is insufficient evidence to make recommendations at this time; therefore, no comment is made on their use. In addition, there is the potential for harm from any treatment, and potential drug interactions are possible (including potentially negative interactions with treatments, for which there is good evidence).

STATEMENT 12. OUTCOMES ASSESSMENT: Clinicians should obtain follow-up audiometric evaluation for patients with SSNHL at the conclusion of treatment and within 6 months of completion of treatment. *Recommendation based on observational studies with a preponderance of benefit over harm.*

Action Statement Profile: 12

- Quality improvement opportunity: Following patients with SHL may allow for identification of underlying causes not evident at presentation and will allow for appropriate rehabilitation of hearing loss in those that fail to recover hearing (National quality strategy: Effective Communication and Care Coordination)
- Aggregate evidence quality: Grade C, based on observational studies
- Level of confidence in the evidence: High
- Benefits: Assess outcome of intervention, identify patients who may benefit from audiologic rehabilitation, identify cause of hearing loss, identify progressive hearing loss, improve counseling
- Risks, harms, costs: Procedural cost
- Benefits-harm assessment: Preponderance of benefit over harm
- 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: Small
- Exceptions: None
- Policy level: Recommendation
- Differences of opinion: While the entire group agreed that a hearing test at the conclusion of therapy is warranted, there was some disagreement about when a longer-term follow-up audiogram should be obtained.

Supporting Text

The purpose of this statement is to highlight the importance of audiometric follow-up in patients with SSNHL for completeness of care, to identify patients who might benefit from rehabilitation options for residual hearing loss and/or tinnitus, and to assess for possible etiologies that may manifest much later after the initial hearing loss episode. It is inadequate to rely on patient self-report, as there are no studies confirming that patients can accurately differentiate degree of hearing loss and type or etiology of hearing loss or identify those hearing losses that require audiologic or medical intervention. Patients and managing care teams are best served by a comprehensive audiologic evaluation to guide treatment as well as additional rehabilitative 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.

Long-term follow-up was reported on 156 patients diagnosed with SSNHL.[230] Of the 121 patients (76%) who showed recovery after 10 days of combination therapy, 54.5% (66 patients) recovered within that 10-day period. Of the other 55 patients (45.5%), 78.2% recovered within 1 month, but 21.8% (12 patients) had delayed hearing recovery later than 1 month after discharge. Delayed recovery was seen within 1 to 2 months in 3 patients, 2 to 3 months in 7 patients, and after 3 months in 2 patients. Those 2 showed significantly delayed recovery at 6 and 8 months postdischarge. While the majority of patients did not recover completely, of those demonstrating recovery, final hearing levels were reached by 1 month in 90% of patients and by 3 months in 98.3% of patients.

Additional studies have noted hearing recovery at 3 months,[144,224] while others have noted hearing stability from months 2 to 4,[231] months 2 to 6,[232] and months 1 to 12.[233] In a noninferiority RCT comparing oral versus IT steroid therapy in 250 patients with unilateral SSNHL, improvement was seen in the majority (193 of 250, 77%) of cases at 2 months posttreatment and was found to be stable at 6 months for both groups.[232] There was a single outlier patient who showed recovery after 6 months.

If there is residual or permanent hearing loss and/or tinnitus, this may require auditory rehabilitation. For management of persistent, bothersome tinnitus in patients with SSNHL or any other cause, see the CPG on tinnitus.[11] 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 (see KAS 13). 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. It is beneficial to discuss the need for ongoing monitoring as well. The exact frequency of monitoring evaluations may vary depending on other suspected etiologies as well as the patient's rehabilitative needs. Periodic evaluations of hearing to monitor stability as well as evaluation of amplification/assistive listening devices to monitor device function, settings, and benefit are recommended at least annually for adults and children aged >5 years.[234] More frequent monitoring may be warranted in individuals with fluctuating hearing loss.[234] State licensure and several policy documents (refer to American Speech-Language-Hearing Association website; http://www.asha.org/Advocacy/state/) as well as clinical convention recommend a hearing evaluation within 6 months of fitting amplification but not audiologic monitoring per se. US FDA statements allude to this as well.[235] These regulatory recommendations and requirements are felt to bolster the argument for a 6-month follow-up audiogram after SSNHL.

Follow-up Audiometric Measures to Assess the Effectiveness of Treatments for SSNHL. The most accurate and cost-efficient method to monitor the effectiveness of medical interventions to treat SSNHL is to compare initial audiometric evaluation (ie, frequency-specific hearing thresholds and speech audiometry) with follow-up audiometric evaluations. This is also the method utilized in the literature, as shown in a meta-analysis of 20 studies with placebos, steroids, antiviral agents, other active therapies, and IT dexamethasone injections to treat SSNHL.[10] Although the treatments were diverse, all the studies used pure tone hearing threshold assessments, PTA, and/or WRSs to determine the effectiveness of treatment leading to recovery of hearing.

As most patients with SSNHL do not have premorbid audiograms, after verification from the patient that the ears were subjectively the same in terms of hearing pre-SSNHL, it is reasonable to use the opposite, uninvolved, ear as the ''baseline'' for the involved ear. This stems from the work used to define SSNHL.[5,10] There have been many definitions of ''recovery'' to define improvement in hearing attributable to treatment. Two of the commonly used definitions are summarized in **Table 12**.

A review of 25 studies (summarized in **Table 12**) on IT steroid therapy for SSNHL showed that the definition of ''recovery'' was varied and ranged from any improvement to full recovery to normal hearing.[10]

The suboptimal nature of many of these outcome measures is as follows. A 10–dB HL change in PTA is not

Table 12. Definition of Recovery across 25 Papers of Intratympanic Steroids in SSNHL.[a]

Paper	Recovery	Comparator
Battaglia et al (2008), Ahn et al (2008), Xenellis et al (2006), Ho et al (2004), Kilic et al (2007), Van Wijck et al (2007), Kakehata et al (2006), Lautermann et al (2005), Plontke et al (2005), Dallan et al (2006), Lefebvre et al (2002)	10– to 30–dB HL improvement in PTA	Pretreatment PTA
Plaza et al (2007), Roebuck et al (2006), Choung et al (2006), Haynes (2007), Banerjee and Parnes (2005), Herr and Marzo (2005), Gianoli and Li (2001), Kopke et al (2001), Silverstein et al (1996)	10– to 30–dB HL improvement in PTA AND 10%-20% improvement in WRS	Pretreatment PTA and WRS
Battista (2005), Gouveris et al (2005)	Individual PTA recovery with Wilson et al (1980) criteria	Presudden HL PTA
Slattery et al (2005)	Improvement to 50%	Opposite ear
Chandrasekhar (2001)	Improvement in WRS and PTA	Opposite ear
Parnes et al (1999)	Hearing is within normal limits and serviceable	None

Abbreviations: PTA, pure tone average; SSNHL, sudden sensorineural hearing loss; WRS, word recognition score.
[a]For all 25 papers, see Hu and Parnes (2009).[273]

much above the test-retest reliability of measuring pure tone hearing thresholds.[10,80] Using a fixed 10% to 20% WRS criterion is also problematic. First, for profound hearing loss with very low speech understanding, a 10% or even 20% improvement might still leave a patient with nonserviceable hearing (below 50% understanding). Additionally, speech audiometry can be confounded by type of speech stimulus (recorded voice vs live voice; within live voice, accent issues), level of presentation, as well as word list length. It is established that recorded-voice WRS testing is consistently more accurate than monitored live voice. Monitored live voice generally takes a shorter time than recorded testing but can be variable, probably dependent on the accents of the audiologist and the patient. WRS is a suprathreshold test and is ideally measured at 20 to 30 dB above detection levels. A monosyllabic word list of only 10 or 25 words may result in statistically incorrect assessments of WRS outcome, especially if compared with a list of 50 words, which is considered the standard.[85]

Additionally, as part of the hearing health care team, the clinician should document the patient's comments concerning hearing, tinnitus, sensation of fullness, dizziness/unsteadiness, or vertigo following treatment.

Recommendations for Outcomes Assessment in Future Studies.

Current Limitations. Two primary limitations exist with these strategies. First, while an absolute improvement in measures may be statistically significant, this may not be clinically significant. Second, in the majority of patients, the pre-SSNHL hearing levels in the affected ear are not known and must therefore be estimated according to available information (eg, patient report).

Recommendations. To address these issues, the GUG 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 WRSs to within 5% to 10% of the unaffected ear
3. Partial recovery should be defined in 2 ways based on whether the degree of initial hearing loss after the event of SSNHL rendered the ear nonserviceable (per AAO-HNSF definition) or not.
4. Anything less than a 10–dB HL improvement should be classified as no 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, an improvement of >10 dB in pure tone thresholds (accounting for test-retest variability in audiometry) 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.

For future studies of treatment outcomes, the panel urges the use of the standardized format for reporting hearing outcomes detailed in Gurgel et al.[236]

The GUG recognizes that these criteria still have limitations in that the impact of an absolute 15-dB improvement in pure tone sound detection or 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 SSNHL who have residual hearing loss and/or tinnitus about the possible benefits of audiologic rehabilitation and other supportive measures. *Strong recommendation based on systematic reviews and observational studies with a preponderance of benefit over harm.*

Action Statement Profile: 13

- Quality improvement opportunity: To inform patients about strategies to help manage residual hearing loss and tinnitus (National quality strategy: Effective Communication and Care Coordination; Prevention and Treatment of Leading Causes of Morbidity and Mortality; Health and Well-being of Communities)
- Aggregate evidence quality: Grade B, based on systematic reviews and observational studies
- Level of confidence in the evidence: High
- Benefits: Improved awareness of options that may improve QOL, functionality, hearing, and tinnitus and offer emotional support
- Risks, harms, costs: Time and cost of counseling
- Benefits-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: None
- Role of patient preferences: Large
- Exceptions: None
- Policy level: Strong recommendation
- Differences of opinion: None, but 2 panelists were recused from the discussion regarding cochlear implants as rehabilitation for tinnitus and single-sided deafness, as they are investigators on industry-funded studies of that technology

Supporting Text

The purpose of this statement is to increase awareness that counseling and education for patients on the options available to manage their existing hearing loss are beneficial. Counseling is a critical component of all aspects of patient care. While this action statement emphasizes its importance for patients with incomplete recovery from SSNHL, it should be noted that counseling is an integral focus throughout the assessment and treatment process for SSNHL.

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 that 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 or other assistive listening technologies. While these questions cannot be answered during the initial treatment period, a continuous dialogue to share information and listening will assist the patient's adjustment to the changes that have occurred and, in some cases, may be permanent.[237] Carlsson et al recommends a multidisciplinary approach to rehabilitation for these patients to contend with the multifaceted problems associated with SSNHL.[14] Dallan et al found that detailed and honest counseling should be considered a vital element in patient management and can be beneficial even after failure of audiologic management.[1] **Table 13** highlights common issues that may need to be addressed when counseling your patient through the process of managing SSNHL.

While the majority of hearing loss associated with SSNHL is unilateral, this does not diminish the handicapping effect that 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.[23,238] For those who reported the presence of tinnitus, 56% demonstrated handicap as measured by the Tinnitus Handicap Inventory.[239]

Self-assessment measurement tools, such as the Hearing Handicap Inventory for the Elderly,[240] and the modified version for use with adults, the Hearing Handicap Inventory for Adults, 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 aids or hearing assistive technology 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 feasible. 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.[10]

There are a variety of amplification options available for the management of unilateral impairment.[241] Traditional recommendations are the contralateral routing of signal 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. For individuals who may have a preexisting hearing loss in the better-hearing ear, bilateral contralateral routing of signals hearing aids are recommended that will allow both contralateral routing of signal 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. The development of further technology has been

Table 13. Common Issues Raised by Individuals with SSNHL.

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	Advise that the risk of SSNHL in the other ear is very low (see Introduction)
Is there anything I can do to help my hearing now that medical therapies are done and I still have hearing loss?	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 America
Do I have to wear a hearing aid?	Personal adjustment/ informational counseling	Discuss types of hearing aids and CROS and BiCROS options if appropriate
Is there any surgery I can have to get my hearing back?	Informational/personal adjustment	Discuss surgical options (ie, cochlear implant, osseointegrated implant) if a candidate

Abbreviation: BiCROS, bilateral contralateral routing of sound; CROS, contralateral routing of signal; SSNHL, sudden sensorineural hearing loss.

investigated for amelioration of single-sided deafness. Osseointegrated bone conductive devices use bone conduction as a means of transferring sound from the affected side to the better-hearing cochlea. Although this is a surgical option, head band placement is available for those individuals who may not be surgical candidates. Deep, snug intracanal devices provide nonsurgical bone conduction sound transmission for the treatment of single-sided deafness.

Cochlear implantation is an option in the rehabilitation of patients with unrecovered severe to profound SSNHL, especially in those with associated tinnitus. In a systematic review, 96% of patients with preoperative tinnitus reported an improvement of tinnitus following cochlear implantation, and, overall, patients experienced improvement in sound localization and speech discrimination.[242] Cochlear implantation for unilateral SNHL has been shown to provide both improved hearing and a significant improvement in QOL.[243]

Hearing assistive technology 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 through either hardwire or wireless technologies, such as infrared and frequency modulated. Other considerations for assistive technology include auditory, visual, and tactile alerting systems. For additional information regarding the rehabilitative options for adults with SNHL, the reader is referred to the CPGs of the American Academy of Audiology.[244]

Coping with the issues resulting from the SSNHL may require more than professional intervention. Consumer-based organizations may be a valuable resource for support and information. The Hearing Loss Association of America is the largest, but by no means the only, consumer-driven organization for adults with hearing loss. Many patients rely on the information that 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.[10] 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. A recent RCT shows evidence that the internet can be used to deliver intervention of rehabilitation to the hearing impaired.[245] There are various online resources that can offer assistance on auditory rehabilitation, including online/DVD self-study programs.

Counseling and rehabilitative services are effective ways to allow the patient with SSNHL to cope with the loss of hearing and manage independently to the best of their 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 at http://www.entnet.org, the AAO-HNSF website, and will include decision tools and patient aids. The updated guideline was presented as a panel presentation to AAO-HNS members and attendees at the AAO-HNSF 2018 Annual Meeting & OTO Experience. Existing AAO-HNSF printed and online patient information will be updated to reflect the updated guideline recommendations.

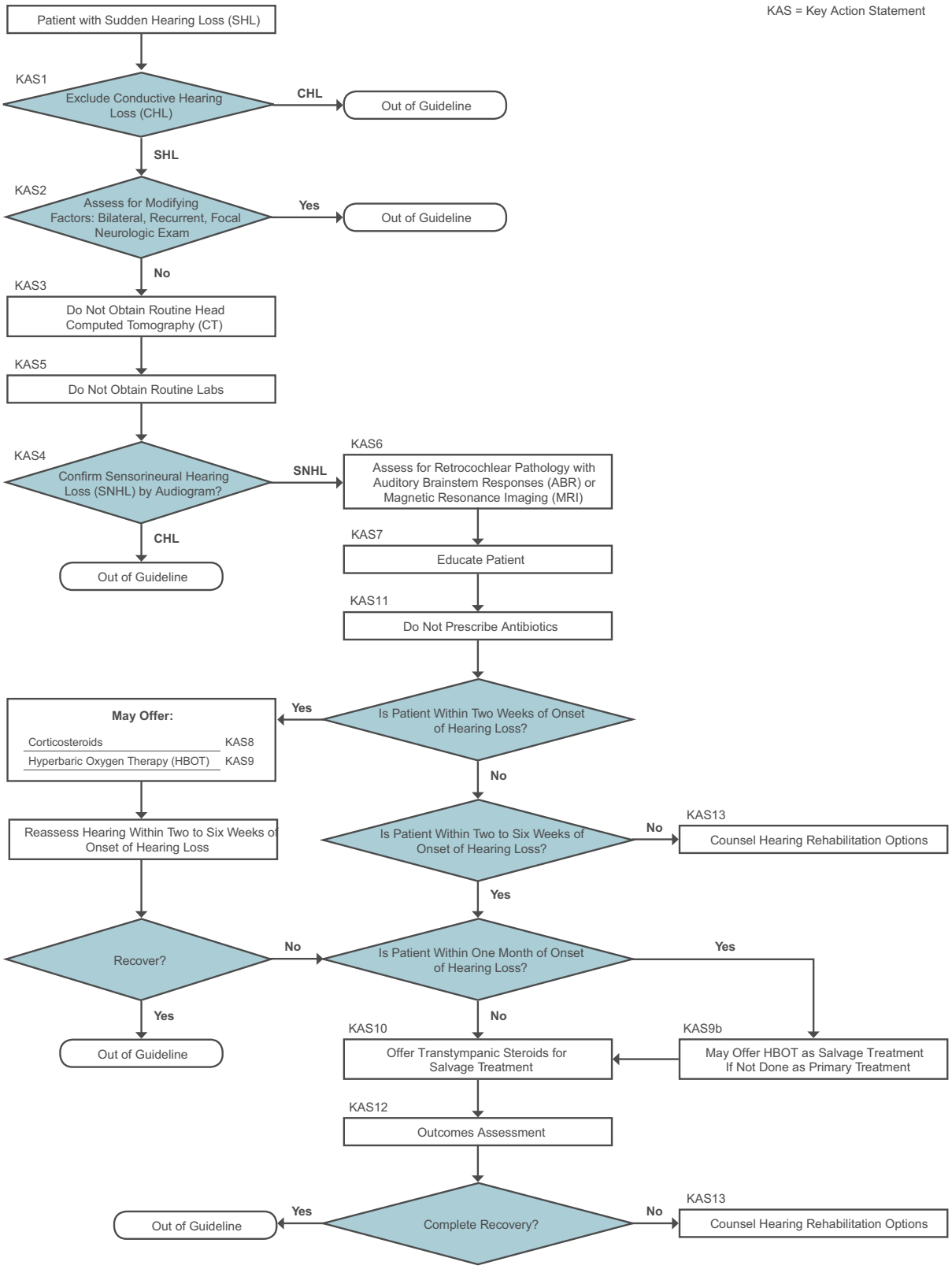

Figure 1. Sudden hearing loss clinical practice guideline algorithm.

An algorithm of the guideline's KASs has been provided (**Figure 1**). The algorithm allows for a more rapid understanding of the guideline's logic and the sequence of the KASs. The GUG hopes that the algorithm can be adopted as a quick reference guide to support the implementation of the guideline's recommendations.

To distinguish SNHL from CHL, the GUG recommends a combination of history, physical examination, tuning fork tests, and audiometry. To aid clinician's implementation of this recommendation, a description of the Weber and Rhine 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 education 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 SSNHL. As such, it is important for both clinicians and patients to be aware of the possible etiology of the 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 SSHNL, with a list of discussion points. The panel believes that these resources can be incorporated into patient information 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, HBOT, 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 SSNHL.

To aid patients in managing their SSNHL, **Tables 9 and 13** (counseling issues raised by patients with SSNHL) will be adapted into patient handouts. The AAO-HNSF will seek the assistance of the consumer groups represented on the GUG 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 GUG'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. Determine the actual incidence of SSNHL in the United States.
3. Investigate the impact of ethnicity and socioeconomic status on timeliness of diagnosis and treatment, treatment outcome, and rehabilitation.
4. Investigate the effectiveness of systemic corticosteroid treatment versus a placebo. The panel believes that such a clinical trial should be conducted due to the equipoise of existing data.
5. Investigate the benefit of HBOT. Current evidence regarding this treatment option is equivocal. Additionally, there is a bias among US physicians and payers not to offer this therapy. Standardized treatment protocols and outcomes assessments are needed for HBOT for ISSNHL.
6. Development of standardized outcome criteria to aid the comparison of clinical studies.
7. The use of IT steroids, as primary and salvage therapy, needs to be further studied. Particularly, the optimal medications, dosage, concentrations, timing, and administration schedules for IT therapy need investigation. The panel believes that differing concentrations of steroid in injected solutions, ranging from 4 mg/mL to 24 mg/mL of dexamethasone, for example, contribute to the inhomogeneity of study outcomes.
8. Develop criteria to determine at what level of hearing-recovery IT steroids would be offered as salvage.
9. Determine the percentage of patients who gain serviceable hearing as a result of treatment. Here we emphasize the importance of WRS percentages, acknowledging that even a severe pure tone loss but with good or better word recognition ability is a good outcome.
10. Investigate the use of ''combined therapy'' (ie, oral and IT steroids) in patients with SSNHL.
11. Develop long-term follow-up protocols for patients with SSNHL.
12. Evaluate therapies with standardized definitions and treatment protocols across studies.
13. Develop a protocol with stacked ABR to better detect small retrocochlear lesions, which is usable in the routine clinical audiologic setting.
14. Investigate the effectiveness of targeted laboratory assessment in determining the etiology of SSNHL.
15. Investigate association of SHL with other risk factors, such as stroke and coagulopathies.
16. Investigate new agents for the treatment of SSNHL.

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Disclaimer

This CPG 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. As medical knowledge expands and technology advances, clinical indicators and guidelines are promoted as conditional and

provisional proposals of what is recommended under specific conditions but are not absolute. Guidelines are not mandates. These do not and should not purport to be a legal standard of care. The responsible physician, in light of all circumstances presented by the individual patient, must determine the appropriate treatment. Adherence to these guidelines will not ensure successful patient outcomes in every situation. The AAO-HNSF emphasizes that these clinical guidelines should not be deemed to include all proper treatment decisions or methods of care or to exclude other treatment decisions or methods of care reasonably directed to obtaining the same results.

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