Purpose of review
Sudden sensorineural hearing loss (SNHL) is an otologic emergency and should be managed quickly and effectively. This review focuses on the management of sudden SNHL, primarily idiopathic sudden SNHL as it is the most common cause.

Recent findings
Management options include observation, oral steroids, intratympanic steroids, or combined oral/intratympanic steroids. One-third to two-thirds of patients will achieve spontaneous recovery, most likely within the first 2 weeks. Despite the lack of randomized controlled trials on steroid therapy efficacy, all patients should be offered steroid treatment given low risk and possible significant benefits. All patients should undergo MRI with gadolinium to rule out retrocochlear disorder. Bilateral sudden hearing loss should alert the clinician to possible systemic disease.

Summary
Sudden hearing loss is an otologic emergency. Appropriate counseling of patients is necessary to allow physician and patient to make a joint, educated decision. It is paramount to rule out retrocochlear disorder and to follow patients closely for improvement or need for future auditory rehabilitation.

Keywords
idiopathic, intratympanic steroids, sudden sensorineural hearing loss

INTRODUCTION
The evaluation of sudden hearing loss begins with a thorough history and physical exam. History is focused on symptom onset, progression, prior history of hearing loss as well as other symptoms, including dizziness, aural fullness, tinnitus, cardiovascular disease, and neurologic deficits. Medications should be reviewed to rule out ototoxic sources. Examination should rule out possible conductive hearing loss as well as confirm a sensorineural hearing loss (SNHL). Routine pure-tone audiometry, including speech reception thresholds (SRT) and word recognition scores (WRS), confirms the diagnosis of SNHL. When premorbid audiology is not available, the contralateral hearing is used for comparison in unilateral loss. The most common audiometric criterion for sudden SNHL is a decrease in hearing more than 30 dB affecting at least three consecutive frequencies within a 3-day period [1]. Unfortunately, not all patients who present with sudden SNHL fit these criteria and should not be excluded from treatment. For bilateral sudden SNHL, the audiometric criterion is less defined when premorbid audiology is not available. Treating physicians must rely on the patient’s subjective sense of decline and confirm a bilateral SNHL with pure-tone audiology.

Often, history will lead to a probable diagnosis, including idiopathic sudden SNHL (ISSNHL). At our institution, if ISSNHL is suspected, MRI with gadolinium is obtained to rule out retrocochlear disorder. For patients who are unable to undergo a MRI scan, a fine-cut computed tomography with contrast is ordered and auditory brainstem response is considered. Laboratory testing is not routinely ordered unless history suggests a need. A myriad of laboratory testing may be indicated and more common tests include complete blood count with differential, thyroid-stimulating hormone, syphilis serology, erythrocyte sedimentation rate, antinuclear antibodies, anticardiolipin antibodies, lupus anticoagulant, antineutrophil cytoplasmic antibodies, Lyme titers, and clotting factors. Despite ordering imaging and possible laboratory tests, treatment is not deferred.
Vasodilators and thrombolytics have minimal prescribed because of the lack of supporting evidence. Antivirals are not precluded because oral or intratympanic steroids are recommended. Common contraindications to oral steroids include diabetes, smoking, and a history of cardiovascular disease or hypertension. Despite the degree of hearing loss, patients will have varying treatment goals. For patients with serviceable hearing, treatment aims to maintain the current level and possibly restore hearing to baseline, defined by contralateral hearing or patient perceived premorbid baseline. Patients with nonserviceable hearing are counseled on the goal of reaching serviceable hearing and the hope of returning to premorbid hearing. Immediate treatment options include observation with close follow-up versus steroid therapy. We firmly believe in the practice of patient involvement in making treatment plan decisions, and therefore, patients are given options to help determine a plan. Patients are educated that one-third to two-thirds of patients will achieve spontaneous recovery, the majority within the first 2 weeks [2,3]. Despite the lack of randomized controlled trials on steroid therapy efficacy, it is offered to all patients given low risk and possible significant benefit [4]. The risks, benefits, and efficacy of oral steroids, intratympanic steroids as well as combination therapy (concomitant oral and intratympanic steroids) are discussed. Prior literature has demonstrated that combination therapy has advantages over oral or intratympanic therapy alone, and we agree based on our experience [5,6*,7,8]. If contraindications or patient preference either preclude oral or intratympanic steroids, then oral or intratympanic therapy is recommended quoting similar efficacy between the two [9]. Common contraindications to oral steroids include diabetes, hypertension, and gastritis. Antivirals are not prescribed because of the lack of supporting evidence. Vasodilators and thrombolytics have minimal evidence supporting efficacy and carry a high risk of adverse events, and therefore, are not routinely offered at our institute. Hyperbaric oxygen (HBO) is an encouraging adjuvant therapy and is discussed; however, limited access to HBO, high cost, and time-consuming appointments usually preclude therapy [4]. For patients who elect oral steroids or combination therapy, common side-effects of oral steroids, including insomnia, dizziness, weight gain, sweating, mood changes, gastric acid secretion, and hyperglycemia are discussed. Patients are prescribed a 7-day course of oral prednisonone, 1 mg/kg/day up to 60 mg/day, followed by a 3-day taper. A total of 10 days and maximum of 60 mg of steroids is selected as this has limited side-effects and minimal suppression of the hypothalamic–pituitary–adrenal axis with maximum dose delivery to inner ear. Steroids are recommended as a single dose in the morning, and all patients receive an oral steroid risk sheet. Patients are also instructed to begin a proton-pump inhibitor. If patients develop intolerable side-effects, the oral steroids are discontinued and intratympanic steroids are recommended.

Patients with ISSNHL who elect intratympanic or combination therapy undergo in-office injections. Prior to the injection, adverse side-effects, including pain, transient dizziness, infection, possible vasovagal response, and persistent tympanic membrane perforation are discussed. Concentration of intratympanic dexamethasone is 10 mg/ml; ideally, a higher concentration up to 24 mg/ml would be chosen. However, compounding pharmacies in our area have difficulty in providing a higher concentration. Once consent is obtained, patients are placed in the supine position and topical phenol is applied to the posterior tympanic membrane, above the round window membrane. One milliliter of dexamethasone (10 mg/ml) is drawn into a syringe and a 27-gauge needle is attached. The syringe and contents are warmed with hand massage prior to delivery to reduce caloric stimulation. On average, 0.4–0.5 ml of dexamethasone are injected transtympanically with single injection site into the middle ear space immediately lateral to the round window. Prior to injection, patients are educated on the possibility of transient caloric dizziness as well as bitter taste. After injection, patients remain in the supine position with affected ear up for 15–30 min and are instructed not to swallow. Patients are then discharged from clinic with a steroid injection information sheet, which includes procedure description, risks, and benefits. All patients, including observation patients, return in approximately 2 weeks with MRI with gadolinium, laboratory testing if ordered, and repeat pure tone audiogram, including SRT and WRS. Two
weeks is the selected follow-up as the greatest improvement in hearing occurs during the first 2 weeks. Recovery is defined as complete, partial, or no recovery based on SRT and WRS. For patients experiencing unilateral ISSNHL, complete recovery is defined as a return in SRT within 10 dB and WRS within 5–10% of unaffected ear. Partial recovery is more challenging to define and may be a progressive process. Partial recovery is considered if patients improve at least 10 dB on SRT and/or at least 10% WRS relative to audiogram on presentation. No recovery is defined as less than 10 dB improvement on SRT and less than 10% improvement on WRS compared with audiogram on presentation.

If patients have complete recovery, they are scheduled for repeat audiogram in 3 months and are educated on the possibility of hearing loss recurring. If hearing loss abruptly returns, patients are instructed to contact the clinic immediately for evaluation. Salvage intratympanic steroid therapy is offered to patients who have partial or no recovery. Patients are quoted up to 33% chance of improvement with salvage intratympanic therapy [10,11]. For partial recovery, salvage intratympanic steroid injections are offered with repeat audiograms every 2 weeks. Treatment is terminated when patients reach complete recovery or do not demonstrate continued partial recovery. For patients who do not recover after initial treatment, salvage intratympanic therapy is offered followed by repeat audiogram in 2 weeks. Again, treatment is aborted if no improvement occurs after one salvage injection, otherwise patients are treated via partial recovery pathway. Anecdotally, we have noted most salvage intratympanic steroid patients receive two injections.

Bilateral sudden SNHL is a rare disease and is defined similarly to unilateral sudden SNHL. Focus is mainly on cause as it may represent a systemic disorder. The most prominent causes include toxicity, neoplastic, vascular, and autoimmune. Acute SNHL treatment options include observation with close follow-up or a 7-day course of high-dose oral steroids followed by 3-day taper. Patients are educated on the limited evidence supporting steroid treatment of bilateral SNHL as well as that the prognosis is worse for bilateral loss compared with unilateral sudden SNHL [12]. Patients return for a repeat audiogram in 2 weeks. Complete recovery in patients with premorbid SNHL is difficult to determine, and physicians must rely upon patients’ subjective assessment if no premorbid audiogram is available. If hearing returns within limits of normal hearing, then complete recovery should be assumed. Partial improvement is defined as at least 10 dB gain in SRT and at least 10% WRS in one or both ears. Patients who have complete recovery return to clinic as previously described for unilateral ISSNHL complete recovery. Treatment dilemmas arise in patients who have partial recovery (unilateral or bilateral) or no recovery. For partial unilateral recovery, combination therapy with repeat oral steroids and salvage intratympanic steroids for the worse hearing ear are offered. For partial symmetric bilateral recovery, a second course of high-dose oral steroids is offered. Salvage intratympanic steroid therapy may be continued to be offered for the worse hearing ear with termination points described as above. Oral steroid therapy is not continued after two courses given the risk of chronic steroid use as well as the limited efficacy of steroid therapy in hearing loss recovery after 4–6 weeks [13–18].

As a tertiary neurology practice, we are often faced with considering treatment for patients outside the ideal treatment window of 4–6 weeks. After ensuring appropriate imaging and laboratory testing has been performed, patients who are within 3 months of ISSNHL onset are offered only salvage intratympanic dexamethasone with guarded prognosis [16]. They return with audiogram in 2 weeks and are managed in aforementioned algorithms for complete, partial, and no recovery. If patients are outside this time period, the lack of evidence supporting steroid therapy is discussed, and hearing rehabilitation is offered. Inevitably, scenarios of ISSNHL not mentioned will arise and create treatment dilemmas for practicing physicians. Proper patient education about the risks, benefits, and limitations of treatments form a cornerstone for management. The importance of patient education leading to involved decision-making cannot be underestimated given the lack of ‘best treatment’ for ISSNHL.

CONCLUSION

Sudden SNHL is an otologic emergency and requires prompt evaluation. The most common cause of sudden SNHL is idiopathic sudden loss. All patients should undergo MRI with gadolinium to rule out retrocochlear disorder and laboratory testing obtained when indicated. Bilateral sudden SNHL should alert physicians to a possible systemic disorder and require diligent evaluation. The cornerstone of therapy is steroid therapy, oral, intratympanic, or combination. HBO is a promising therapy when instituted early in treatment. All treatment modalities, including observation, should be discussed with patients including efficacy for each modality. Close audiometric follow-up is mandatory to determine need for salvage therapy.
intratympanic steroids versus need for future auditory rehabilitation.

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REFERENCES AND RECOMMENDED READING
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& of special interest
&& of outstanding interest