

ORIGINAL RESEARCH

Outcomes of unilateral idiopathic sudden sensorineural hearing loss: Two decades of experience

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Abstract

Objectives: (a) Determine the demographic and medical risk factors for patients who presented with unilateral idiopathic sudden sensorineural hearing loss (ISSNHL); (b) identify treatments that patients underwent; (c) evaluate the adequacy of follow-up and compliance with long-term hearing rehabilitation.

Methods: Retrospective review of patients who presented with unilateral ISSNHL between January 1998 and December 2017 at a tertiary care academic medical center.

Results: Two hundred-four patients met inclusion criteria. Of these, 129 (63.2%) did not undergo treatment at an outside hospital prior to our evaluation. In this subgroup, the average pretreatment pure tone average (PTA) was 61.9 ± 2.5 dB (dB). The most common treatment was oral steroids and was recommended in 76 patients (59.9%). Patients also underwent intratympanic (IT) steroid injections (7.2%) or oral steroids followed by salvage IT injections (19.4%). Mean follow-up duration was 17.9 (± 29.2) months, and posttreatment PTA (45.6 ± 2.6 dB) was significantly better than baseline ($P < .001$). In this cohort, hearing amplification was infrequently recommended. Less than 20% of patients reported active hearing amplification use at their most recent visit. At follow-up, 90 patients (69.8%) reported subjective improvement in hearing after treatment. Only 55 patients (42.6%) showed improvement in PTA compared to their pretreatment audiograms.

Conclusion: Many patients with ISSNHL experienced audiometric improvement after treatments, but most had persistent hearing loss. The duration of follow-up was short. Most patients did not use long-term hearing amplification. Future studies are needed to identify factors that contribute to reduced follow-up and low compliance with hearing amplification use in ISSNHL.

Level of Evidence: 2c.

KEYWORDS

hearing amplification, idiopathic hearing loss, idiopathic sensorineural hearing loss, sudden hearing loss

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1 | INTRODUCTION

Idiopathic unilateral sudden sensorineural hearing loss (ISSNHL) is a rapid onset of hearing loss from cochlear or retrocochlear origins. Unilateral ISSNHL is characterized by subjective and objective hearing impairment with decrease in hearing thresholds of ≥ 30 dB (dB) in three or more consecutive frequencies on dedicated audiometric testing.¹ Global incidence of ISSNHL is estimated to be 5 to 20/100000 persons.²⁻⁴ In the United States, 4000 new cases are reported each year.³ Viral infections, cochlear ischemia, metabolic derangement, and autoimmune processes have all been proposed as potential etiologies.^{3,5}

Treatment for unilateral ISSNHL is aimed at the recovery of hearing thresholds.² Various treatment options, including oral and intratympanic (IT) steroids, diuretics, antiviral, and hyperbaric oxygen therapy, have been studied in case reports, retrospective reviews, and randomized controlled trials.^{1,2,5-7} In the American Academy of Otolaryngology—Head and Neck Surgery (AAO-HNS) clinical practice guideline (CPG) on sudden hearing loss, systemic steroids were proposed as a first-line agent for ISSNHL.¹ IT steroid injections were determined to be a useful adjunct for select patients.⁷ For individuals with partial or no hearing recovery, long-term hearing amplification is recommended. Options for hearing amplification include conventional hearing aids, contralateral routing of signal (CROS) devices, osseointegrated bone conduction devices, and cochlear implants (CI).⁸⁻¹² Prior studies suggest the need for ongoing otolaryngologic, audiologic, and psychosocial evaluations for patients with ISSNHL.¹

While the treatment efficacy for steroids has been well described, less is known about the adequacy of follow-up and patient compliance with long-term hearing amplification. With two decades of experience, we set out to systematically characterize long-term hearing outcomes in patients with unilateral ISSNHL. Beyond providing descriptive information on demographics and medical risk factors, our analysis sought to provide a summary of treatment modalities and duration of follow-up. We also sought to characterize patients' perception of hearing recovery and their audiometric outcomes. These analyses offer insight into how patients with unilateral ISSNHL recover, and data obtained in this study may be used to inform patient expectations or guide long-term treatment strategies for unilateral ISSNHL.

2 | MATERIALS AND METHODS

2.1 | Study population

This study was a retrospective review of clinical data. Individuals 18 years and older who presented to the University of Michigan between January 1998 and December 2017 were identified. Consecutive data were reviewed. Patients met the following inclusion criteria: a history of unilateral sudden hearing loss of ≥ 30 dB in three or more consecutive frequencies (confirmed on pure tone audiometry) without an identifiable cause (ie, normal imaging, no identified middle ear infection, or history of trauma). We excluded patients who presented with conductive hearing loss, gradual hearing loss, congenital

deafness, retrocochlear lesions (ie, vestibular schwannoma), endolymphatic hydrops, intracranial pathology, or those with other known causes of hearing loss. The study was approved by the University of Michigan Institutional Review Board (IRB-MED) and conforms to previously published standards of the Declaration of Helsinki.

Demographics, medical comorbidities, and otologic risk factors were collected. Medical comorbidities included a history of cardiovascular disease (ie, hypertension, coronary artery disease, cerebrovascular disease, peripheral vascular disease, and valvular heart disease), type II diabetes, migraine, and immunosuppression. In the present study, we defined immunosuppression as immune dysfunction that resulted from an identified autoimmune process (ie, lupus, human immunodeficiency virus/acquired immunodeficiency syndrome, systemic vasculitis, sarcoidosis, or multiple sclerosis) or active use of immune-suppressive medications at the time of sudden hearing loss (ie, chemotherapy, transplant-related medication, or other immune-suppressive medications). We collected information on the duration of hearing loss prior to evaluation at our institution. We categorized this duration as " ≤ 10 days," "11 to 21 days," "22 to 30 days," or longer than 1 month from the onset of auditory symptoms. Many patients presented to their primary care physicians or local otolaryngologists before coming to a tertiary care institution. Therefore, we made note of the treatments they received prior to our assessment by reviewing medical records from referring physicians. At our institution, brain imaging (MRI per the inner ear protocol) was routinely recommended for excluding inner ear or retrocochlear lesions (to include, but not be limited to, vestibular schwannomas). Patients whose diagnostic imaging revealed retrocochlear or intracranial pathologies were excluded from the study.

Patients underwent pure tone audiometry as part of their work-up. The pure tone average (PTA) is traditionally defined as the mean of air conduction thresholds at 500, 1000, and 2000 Hz. In calculating the PTA for this study, we included air conduction thresholds of 250, 500, 1000, 2000, 4000, and 8000 Hz to provide a more comprehensive dB average. Speech reception threshold (SRT, in dB) and word recognition score (WRS, %) were defined according to standard audiometric indications. Most patients had at least one audiometric testing after treatment. The pretreatment and posttreatment PTA, SRT, and WRS were compared. Patients' most recent audiograms were reviewed. We defined this as the most recent audiometric testing to date that was obtained at least 6 months from the time of treatment completion.

The severity of hearing loss was categorized based on the PTA. Normal hearing (< 26 dB), mild (≥ 26 -40 dB), moderate (41-70 dB), severe (71-90 dB), or profound hearing loss (≥ 91 dB) were defined accordingly. In addition, the shape of the pretreatment audiogram has been shown to prognosticate hearing recovery after ISSNHL.¹³ In a recent review by Kuhn et al in 2011, low- and mid-frequency sensorineural hearing loss was associated with better recovery compared to flat or a down-sloping audiogram.¹⁴ In our study, we described an audiogram as "up-sloping" (ie, hearing loss in the low- or mid-frequencies rising to normal hearing), "down-sloping" (ie, hearing loss in the high-frequencies), "flat" (ie, hearing loss across all frequencies),

or “normal hearing in low- and high-frequencies.” Audiogram shapes that did not fit with these descriptions were categorized as “other.”

The recommended treatment for unilateral ISSNHL was recorded. At our institution, patients who received systemic steroids were typically prescribed a course of 60 mg of oral steroid tablets (most commonly prednisone) for 7 to 14 days. This was followed by a 3 to 5-day out to a 2-week taper. No patients in our cohort received systemic steroids through an intravenous route. If an individual was not a candidate for systemic steroids (ie, severe diabetes, allergy or adverse reaction to steroids, chronic immunosuppression at baseline) they were deemed appropriate for IT steroid injections. The patient would receive at least one cycle of IT therapy in clinic (most commonly 0.5-1.0 mL of 10 mg/mL solution of dexamethasone). At our institution, there were variations among providers in their practice patterns regarding steroid dosing, duration, and frequency of medication administration. This variability was consistent with the current state of practice in otolaryngology.¹ Furthermore, we note that some patients deferred treatment for various reasons despite our recommendations. We have recorded this discrepancy as “recommended but did not pursue/use” vs “recommended, active user” at the time of their most recent clinic visit.

Subjective hearing improvement was defined as patient-reported recovery at the most recent clinic visit. This was further classified as patient reporting of “no improvement,” “partial improvement,” or “complete hearing recovery” as compared to the patient’s perceived baseline (pre-loss) hearing. To assess audiometric outcomes, we employed the Siegel criteria.¹⁵ The Siegel audiometric recovery criteria is an accepted classification system and has been used in the ISSNHL literature.^{15,16} We defined audiometric improvement as a PTA improvement of ≥ 15 dB in the first posttreatment audiogram. In this study we also included individuals who achieved normal hearing after treatment. Treatments are defined as “primary” if they were the first recommendations we provided. We defined subsequent therapy as “additional treatment.” The duration of follow-up was determined from the date of initial evaluation to the date of the most recent otolaryngology or audiology clinic visit at our institution.

2.2 | Analysis

Demographics, medical risk factors, treatment modality, and audiometric outcomes were analyzed using descriptive statistics. The STATA 15 statistical software was used for all analyses (StataCorp, College Station, Texas). An independent student’s *t* test was used to compare pretreatment and posttreatment audiometric variables. Two subgroup analyses were performed. First, we compared audiometric data in patients who underwent prior treatments at an outside hospital vs those who were never treated before being seen at our institution for evaluation. Next, we performed a subgroup analysis on treatment modalities in patients who had hearing improvement by the Siegel criteria vs those who did not show improvement. A *P* value $< .05$ was considered statistically significant for all analyses.

3 | RESULTS

3.1 | Demographic and audiometric data

At the University of Michigan, 2387 patients met initial search criteria and were evaluated for asymmetric/unilateral hearing loss between January 1998 and December 2017. Most patients were excluded due to progressive, non-acute hearing loss ($n = 1029$, 43.1%), non-audiologic complaints (ie, vestibular dysfunction, nasal congestion; $n = 859$, 36.0%), or conductive and mixed hearing loss ($n = 351$, 14.7%). Of note, 90 (3.8%) were excluded due to discovery of a middle ear or retrocochlear lesion that presented as unilateral SSNHL.

Two hundred-four patients (8.5% of 2387 patients) met criteria for unilateral ISSNHL (mean age: 55.4 years, 52.5% female, and 83.3% Caucasian). Demographics and medical comorbidities are summarized in Table 1. Most patients (82.8%) underwent imaging to formally rule out an inner ear or retrocochlear lesion. Approximately 16% of patients did not undergo imaging due to refusal or inability to tolerate MRI scans.

In the first subgroup analysis, patients were stratified into those who received initial treatment at an outside facility ($n = 75$, 36.8%) vs those who did not ($n = 129$, 63.2%). Most (60%) patients who underwent prior treatments at an outside facility had hearing loss for at least 31 days by the time they were seen in our institution. In contrast, 59.7% of patients who received no prior treatment presented to our institution with reported hearing loss onset for ≤ 10 days. Treatment with oral steroids was the most commonly prescribed modality in patients who were treated at local facilities. The follow-up duration for the overall cohort was 17.9 months. Follow-up duration between the two subgroups were similar ($P = .344$).

For subsequent analyses, we analyzed patients who were not treated prior to our evaluation ($n = 129$). We excluded patients who received previous treatments ($n = 75$) due to difficulty with extracting treatment details from outside clinical records. Our analysis demonstrated that the two subgroups were similar in terms of demographic and medical characteristics. In the subgroup of patients who received treatment after they were seen by our clinicians, we noted PTA improvement from 61.9 to 45.6 dB ($P < .001$; Table 2). Similarly, SRT improved from 44.6 to 27.5 dB ($P < .001$) while WRS increased from 64.5% to 81.8% ($P < .001$). Twenty-seven individuals (20.0%) had normal hearing and most (65.1%) had mild to moderate hearing loss posttreatment. Most patients had down-sloping (38.0%) or “flat” (30.2%) audiograms before and after treatment. The percentage of a more favorable up-sloping audiogram decreased from 14.7% to 3.1% after primary treatment.¹⁴ Interestingly, in patients’ audiograms at the most recent clinic follow up, we observed a slight increase in PTA and SRT dB level and a slight decrease in WRS percentage as compared to the first posttreatment audiograms.

3.2 | Treatment modality and follow-up

Treatment recommendations are summarized in Table 3 for the 129 patients who presented to our institution without prior treatment.

TABLE 1 Demographic, medical comorbidities, and symptoms of patients who presented with unilateral idiopathic sudden sensorineural hearing loss

Study characteristics	All patients (n = 204)	Subgroup analysis		P value
		No prior treatment (n = 129)	Received prior treatments (n = 75)	
Mean age in years (\pm SE ^a)	55.4 (\pm 15.9)	56.3 (\pm 1.4)	53.9 (\pm 1.8)	.287
Sex (%):				
Male	97 (47.5)	68 (52.7)	29 (38.7)	.053
Female	107 (52.5)	61 (47.3)	46 (61.3)	
Race/ethnicity (%):				
White	170 (83.3)	103 (79.8)	67 (89.3)	.339
African American	5 (2.5)	4 (3.1)	1 (1.3)	
Hispanic	1 (0.5)	1 (0.8)	0	
Other	28 (13.7)	21 (16.3)	7 (9.3)	
History of cardiovascular disease (%):				
Yes	79 (38.7)	59 (45.7)	20 (26.7)	.020*
No	125 (61.3)	70 (54.3)	55 (73.3)	
History of type II diabetes (%):				
Yes	35 (17.2)	26 (20.2)	9 (12.0)	.136
No	169 (82.8)	103 (81.7)	66 (88.0)	
History of migraine (%):				
Yes	21 (10.3)	12 (9.3)	9 (12.0)	.541
No	183 (89.7)	117 (90.7)	66 (88.0)	
History of loud noise exposure (%):				
Yes	20 (9.8)	17 (13.2)	3 (4.0)	.034*
No	184 (90.2)	112 (86.8)	72 (98.0)	
History of immunosuppression (%):				
None	188 (92.2)	115 (89.4)	73 (97.3)	.149
Lupus	5 (2.5)	3 (2.3)	2 (2.7)	
Systemic vasculitis	0	0	0	
Multiple sclerosis	1 (0.5)	1 (0.8)	0	
Medication-induced	1 (0.5)	1 (0.8)	0	
Other ^b	9 (4.4)	9 (7.0)	0	
Affected ear (%):				
Left	97 (47.5)	61 (47.3)	36 (48.0)	.922
Right	107 (52.5)	68 (52.7)	39 (52.0)	
Presenting symptoms (%):				
Tinnitus	52 (25.5)	34 (26.4)	18 (24.0)	.106
Ear fullness or pressure	15 (7.4)	8 (6.2)	7 (9.3)	
Vertigo or imbalance	41 (20.1)	20 (15.5)	21 (28.0)	
Other symptoms	5 (2.5)	5 (3.9)	0	
Multiple	50 (24.5)	32 (24.8)	18 (24.0)	
Hearing loss duration (%):				
\leq 10 days	96 (46.1)	77 (59.7)	19 (25.3)	<.001*
11-21 days	21 (10.3)	13 (10.1)	8 (10.7)	
22-30 days	7 (3.4)	4 (3.1)	3 (4.0%)	
\geq 31 days	80 (39.2)	35 (27.1)	45 (60.0)	
Diagnostic imaging (%):				
Normal	169 (82.8)	104 (85.2)	65 (86.7)	

(Continues)

TABLE 1 (Continued)

Study characteristics	All patients (n = 204)	Subgroup analysis		P value
		No prior treatment (n = 129)	Received prior treatments (n = 75)	
Abnormal: retrocochlear lesion	0	0	0	.305
No imaging	32 (15.7)	22 (17.1)	10 (13.3)	
Treatments prior to our evaluation (%):				
Oral steroid	37 (18.1)		37 (18.1)	
Intratympanic steroid	3 (1.5)	N/A	3 (1.5)	N/A
Both types of steroids	10 (4.9)		10 (4.9)	
Hearing aid	1 (0.5)		1 (0.5)	
Other ^c	16 (7.8)		16 (7.8)	
Multiple	8 (3.9)		8 (3.9)	
Follow up duration in months (\pm SE)	17.9 (\pm 29.2)	19.4 (\pm 2.7)	15.3 (\pm 3.0)	.344

Note: The sub-group analysis stratified patients by individuals who received no prior treatment vs those who were treated before our evaluation. An asterisk indicates statistically significant *P* values.

^aSE, SE of the mean.

^bThis includes Sjogren's syndrome, rheumatoid arthritis, psoriatic arthritis, autoimmune hepatitis, Hashimoto's thyroiditis, and type I diabetes.

^cThis includes treatment with oxymetazoline, anti-histamine medications, fluticasone nasal sprays, and oral antibiotics.

TABLE 2 Audiometric data in patients who presented without prior treatments (n = 129)

Study characteristic	Pre/Post intervention (n = 129)			Most recent audio, >6 months after treatments (n = 51)
	Pretreatment audiometry	Posttreatment audiometry	P value	
PTA, dB (\pm SE ^a)	61.9 (\pm 2.5)	45.6 (\pm 2.6)	<.001*	51.1 (\pm 3.2)
SRT, dB (\pm SE)	44.6 (\pm 2.4)	27.5 (\pm 1.9)	<.001*	39.6 (\pm 3.4)
WRS, % (\pm SE)	64.5 (\pm 3.8)	81.8 (\pm 3.1)	<.001*	73.4 (\pm 4.4)
Hearing loss severity based on PTA (%):				
Normal (<26 dB)	0	27 (20.9)		8 (15.7)
Mild (\geq 26-40 dB)	29 (22.5)	26 (20.2)		12 (23.5)
Moderate (41-70 dB)	59 (45.7)	31 (24.0)	<.001*	20 (39.2)
Severe (71-90 dB)	18 (14.0)	8 (6.2)		8 (15.7)
Profound (\geq 91 dB)	23 (17.8)	19 (14.7)		3 (5.9)
Audiogram shape (%):				
Up-sloping	19 (14.7)	4 (3.1)		1 (2.0)
Down-sloping	49 (38.0)	43 (33.3)		29 (56.9)
Flat	39 (30.2)	31 (24.0)	.761	12 (23.5)
Normal in low and high frequencies	6 (4.7)	8 (6.2)		2 (3.9)
Other	16 (12.4)	12 (9.3)		7 (13.7)

Note: Patients who received prior treatments (n = 75) were excluded. Posttreatment audiograms were obtained 49.5 days after pretreatment audiogram. An asterisk indicates statistically significant *P* values.

Abbreviations: PTA, pure tone average; SRT, speech reception threshold; WRS, word recognition score.

^aSE = SE of the mean.

Most patients (58.9%) underwent treatment with oral steroids. Conversely, 19.4% of patients underwent treatments with oral steroids followed by IT steroid injections. Only nine (7.2%) patients underwent IT steroid injections alone (due to medical contraindication to oral formulation). To assess audiometric response in patients who received steroid treatments, schematic diagrams of hearing results by treatment

modality are provided per published audiologic standards (Figures 1-4)¹⁷ and demonstrate that most patients achieved some hearing recovery after steroid treatment.

Hearing rehabilitation after steroids in the form of conventional hearing aids was recommended to 22 patients (17.1%) as a primary treatment modality. Only 4 patients received a recommendation to

TABLE 3 Treatment recommendations for patients who presented without prior therapy

Treatment modality	Primary treatment recommendation (s) (n = 129)	Additional treatment recommendation (s) (n = 104)
Oral steroids (%)	76 (58.9)	1 (1.0)
Intratympanic steroids (%)	8 (6.2)	1 (1.0)
Oral steroids followed by intratympanic steroid injections (%)	25 (19.4)	0
Conventional hearing aid (%):		
Not recommended	118 (91.5)	81 (62.8)
Recommended:	11 (8.5)	23 (17.8)
Did not pursue/use	7 (5.4)	12 (11.5)
Active user	4 (3.1)	11 (10.6)
CROS/BiCROS device (%):		
Not recommended	125 (96.9)	90 (86.5)
Recommended:	4 (3.1)	14 (13.5)
Did not pursue/use	3 (2.3)	6 (5.8)
Active user	1 (0.7)	8 (7.7)
Bone-anchored hearing aid (%):		
Not recommended	127 (98.4)	101 (97.1)
Recommended:	2 (1.6)	3 (2.9)
Did not pursue/use	0	1 (1.0)
Active user	2 (1.6)	2 (1.9)
Cochlear implant (%):		
Not recommended	129 (100)	102 (98.1)
Recommended but did not pursue	0	2 (1.9) ^a
More than one treatment recommended (%)	1 (0.7)	7 (6.7)
No further recommended treatment (%)	2 (1.6)	54 (49.0)

Note: Patients who received prior treatments at an outside hospital were excluded. Primary treatment is defined as the first set of recommended therapy at our institution. Additional treatments were offered to patients who did not respond to primary treatments. CROS: contralateral routing of signal (BiCROS: bilateral CROS).

^aIn our cohort, two patients were recommended to consider cochlear implants (CI). Both did not pursue CI. One patient was lost to follow-up. The other patient did not have insurance approval for unilateral idiopathic sudden sensorineural hearing loss.

use either a CROS or bilateral CROS (BiCROS) hearing device as a primary therapy. Similarly, bone-anchored hearing aids or cochlear implants (CIs) were not recommended as first-line treatments. Two patients (1.6%) received no further treatment recommendations. In both cases, hearing loss was deemed to be mild by the patient, and no further therapy was pursued.

One hundred four (81%) patients required additional recommendations after the initial therapy (Table 3). In contrast with primary

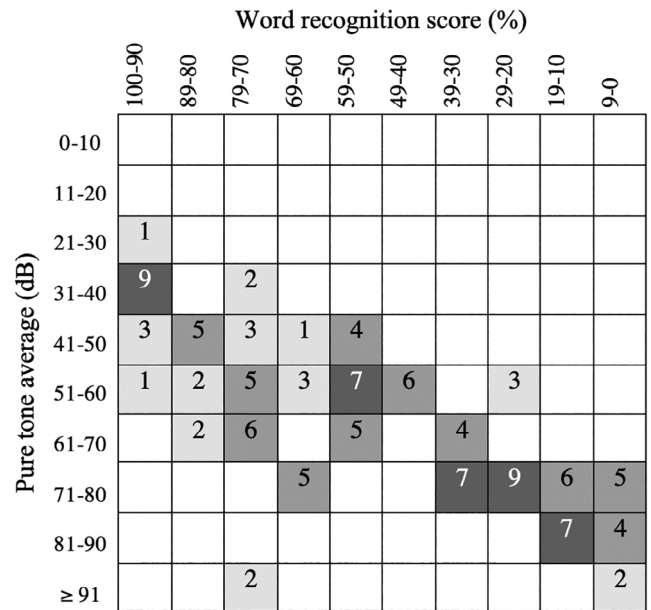


FIGURE 1 Scattergram of baseline audiometric characteristics in patients with idiopathic sudden sensorineural hearing loss (n = 129) who did not undergo prior treatment at outside facilities. The figure was constructed per recently published audiologic standards.¹⁷ Pure tone average (PTA) is traditionally defined as the mean of air conduction thresholds at 500, 1000, and 2000 Hz. In calculating the PTA for this study, we included air conduction thresholds of 250, 500, 1000, 2000, 4000, and 8000 Hz to provide a more comprehensive decibel average

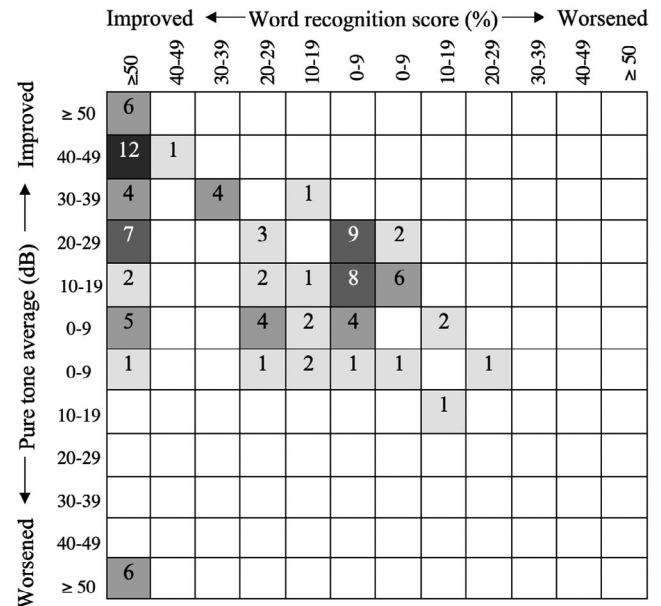


FIGURE 2 Scattergram of posttreatment audiometric characteristics in patients who underwent treatment with oral steroids for idiopathic sudden sensorineural hearing loss. At our institution, patients were typically prescribed a course of 60 mg of oral steroid tablets (most commonly prednisone) for 7 to 14 days. This was followed by a 3 to 5-day out to a 2-week taper. No patients received systemic steroids through an intravenous route

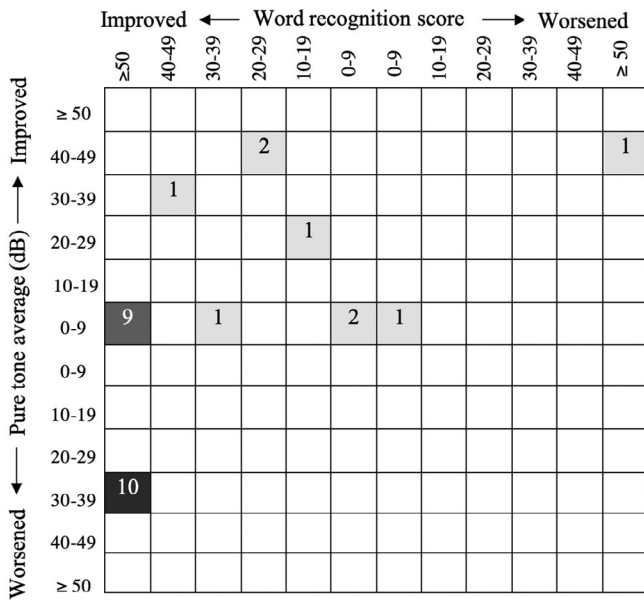


FIGURE 3 Scattergram of posttreatment audiometric characteristics in patients who underwent treatment with intratympanic (IT) steroid injections. IT injections were reserved for patients who were not candidates for systemic steroids (eg, severe diabetes, allergy or adverse reaction to steroids, chronic immunosuppression at baseline). Patient received one or more cycle of IT therapy in clinic (most commonly 0.5-1.0 mL of 10 mg/mL solution of dexamethasone)

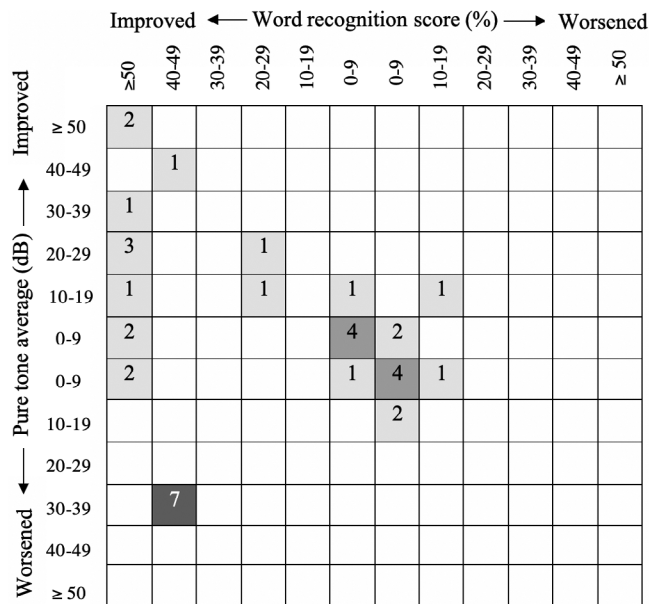


FIGURE 4 Scattergram of posttreatment audiometric characteristics in patients who were treated with dual therapy (ie, oral steroids followed by salvage intratympanic steroid injections). In accordance with the American Academy of Otolaryngology—Head and Neck Surgery Clinical Practice Guidelines,¹ intratympanic steroid injections were used as an adjunct therapy, when patients demonstrated poor or incomplete response to oral steroid. This scattergram displays audiometric outcomes of patients who received both forms of steroids

treatments, a larger proportion of individuals received a recommendation to use hearing aids (17.8%), CROS/BiCROS devices (13.5%), bone anchored hearing aids (2.9%), or consider undergoing a CI (1.9%). Steroids were recommended as a secondary therapy in only 2.0% of patients. Notably, nearly 50% of patients received no further secondary treatment recommendations. Although various modalities were recommended, a small proportion of patients reported active usage of hearing rehabilitation devices at their most recent clinic visit. In our

TABLE 4 Audiometric data in patients who had hearing improvement, defined by pure tone average increase of ≥15 dB between pretreatment and posttreatment audiograms or those who achieved normal hearing (<26 dB) after treatment

	Improved hearing (n = 55)	No improvement in hearing (n = 74)	P value
Study characteristic			
Baseline audio:			
PTA, dB (±SE ^a)	54.8 (±3.0)	67.2 (±2.9)	.004*
SRT, dB (±SE)	45.4 (±3.5)	49.1 (±2.9)	.419
WRS, % (±SE)	65.3 (±5.1)	60.8 (±4.7)	.510
Hearing loss duration (%):			
≤10 days	31 (56.4)	46 (62.2)	.507
10-21 days	4 (7.3)	9 (12.2)	.362
22-30 days	1 (1.8)	3 (4.1)	.469
≥31 days	19 (34.5)	16 (21.6)	.103
Posttreatment audio:			
PTA, dB (±SE)	20.5 (±2.9)	55.0 (±2.8)	<.001*
SRT, dB (±SE)	14.4 (±1.4)	39.3 (±2.8)	<.001*
WRS, % (±SE)	94.8 (±3.5)	67.6 (±4.5)	<.001*
Treatment(s) offered (%):			
Oral steroid	30 (54.5)	46 (62.2)	.385
Intratympanic steroid	3 (5.5)	5 (6.8)	.762
Oral followed by intratympanic steroids	3 (5.5)	22 (29.7)	.001*
Conventional hearing aid	10 (18.2)	1 (1.4)	.001*
CROS/BiCROS	4 (7.3)	0	.018*
Bone-anchored hearing aid	2 (3.6)	0	.098
Cochlear implant	0	0	N/A
Patient-reported hearing improvement:			
None	7 (12.7)	20 (27.0)	
Partial	15 (27.3)	37 (50.0)	.015*
Complete	21 (38.2)	17 (23.0)	
Follow up duration in months (±SE)	15.5 (±4.1)	22.3 (±3.6)	.217

Note: Patients who received prior treatments at an outside hospital were excluded. An asterisk indicates statistically significant P values. Abbreviations: CROS, contralateral routing of signal (BiCROS: bilateral CROS); PTA, pure tone average; SRT, speech reception threshold; WRS, word recognition score.
^aSE = SE of the mean.

study, a CI was recommended to two patients (1.9%) after they did not respond to primary treatment. In both cases, the patients did not seek further evaluation for a CI.

In our second subgroup analysis, we stratified the cohort into patients who had improved hearing (ie, defined by PTA improvement of ≥ 15 dB or normal hearing in the first posttreatment audiogram per Siegel criteria; $n = 55$) vs those who did not ($n = 74$, Table 4).¹⁵ Baseline audiometric characteristics were poorer among patients with no improved hearing. As expected, posttreatment audiograms were significantly better in patients with improved hearing ($P < .001$ for differences in PTA, SRT, and WRS). Both sub-groups presented with similar durations of unilateral ISSNHL. Interestingly, 29.7% of patients with no improved hearing underwent treatment with oral steroids followed by IT steroid injections, compared to only 5.5% in patients who had hearing improvement ($P = .001$). The use of hearing aids, CROS/BiCROS devices, and bone anchored hearing aids was much greater among patients with improved hearing. We note that despite objective evidence of recovery on pure tone audiometry, only 15 (27.3%) patients reported partial subjective hearing recovery and 21 (38.2%) reported a complete recovery of hearing. Patients' subjective perception of hearing recovery was statistically different ($P = .015$) between those with improved PTA vs those with no improved hearing. Patients without an improvement in hearing were followed up for a slightly longer duration (22.3 months) compared to those with improved hearing (15.5 months), although this difference was not statistically significant ($P = .217$).

4 | DISCUSSION

Unilateral ISSNHL is a debilitating condition that poses considerable diagnostic and treatment challenges for otolaryngologists and audiologists. Treatments for unilateral ISSNHL have been extensively reviewed in case reports, retrospective studies, and randomized controlled trials.¹⁻⁷ The AAO-HNS CPG supports early administration of systemic steroids followed by salvage IT steroid injections. Beyond treatments with steroids, the CPG advocates follow-up with otolaryngologists and audiologists and repeat audiometric assessment within 6 months of hearing loss diagnosis.¹

While the treatment efficacy for steroids has been well described, less is known about the adequacy of follow-up and patient compliance with hearing amplification. In reviewing two decades of data, we addressed these questions with a large cohort of patients with unilateral ISSNHL. We used the Siegel criteria for defining hearing recovery.¹⁵ The Siegel criteria was used in recent ISSNHL literature¹⁶ and served as a useful means to quantify treatment outcomes. In the present study, we also excluded patients ($n = 75$) who were treated at local facilities prior to our evaluation. Our rationale for this exclusion was to examine a uniform cohort comprised of patients who were evaluated and treated at a single institution with consistent medical documentation both pretreatment and posttreatment.

In our study, posttreatment audiograms were obtained ~50 days after the pretreatment audiograms. Within this period, we observed

audiometric improvement in 44.4% ($n = 55$) of patients. In reviewing their most recent audiograms, PTA was 51.5 dB and WRS improvement was modest (Table 2). About 60% of our cohort underwent treatment with oral steroids. Furthermore, 19.4% underwent dual therapy with oral followed by salvage IT steroid injections. Only nine patients (7.2%) underwent IT injections alone. These nine patients had underlying medical comorbidities (most commonly, uncontrolled diabetes) that precluded them from systemic steroid use. Among patients who achieved audiometric recovery, oral steroids followed by conventional hearing aids were the two most common recommendations.

Methods for hearing amplification were recommended to 47.3% of patients. Conventional hearing aids were the most popular recommendation, followed by recommendation of a CROS/BiCROS device and a bone anchored hearing aid. Surprisingly, despite low rates of audiometric recovery, few individuals with unilateral ISSNHL were seeking hearing amplification in the long-term. Only 8.4% of patients were reported as active hearing aid users, even though hearing aids were recommended in 20% of patients. Similarly, 0.7% were active users of the CROS/BiCROS device and only 1.6% were active users of a bone conduction hearing device. This is the first study to evaluate patient compliance with long-term hearing amplification use in the idiopathic sudden hearing loss population. The etiology of poor patient compliance with hearing rehabilitation is unclear. It is possible that patients achieved audiometric recovery beyond our follow-up period and were less inclined to use long-term amplification. It is also possible that patients encountered financial and/or insurance barriers that affected their ability to purchase a hearing device. Additionally, with a lack of follow-up beyond 18 months, patients with unilateral ISSNHL were no longer seeking medical advice from their clinicians, who might otherwise advocate for better compliance with hearing amplification use.

Cochlear implantation is an emerging option for sudden hearing loss and has been investigated in recent clinical trials.¹⁸ During the study period (January 1998 to December 2017), patients with unilateral ISSNHL did not meet the United States Food and Drug Administration criteria for a CI.¹⁹ It is thus not surprising that a CI was offered to only two patients in our cohort, after they had failed medical therapies. Both patients did not pursue a CI due to a lack of interest in a surgical option and difficulty with insurance approvals. Since the conclusion of this study period, ~20 patients were recommended to consider a CI for an "off label" indication at our institution. Additionally, the Food and Drug Administration recently approved the MED-EL cochlear implant systems for use in unilateral hearing loss and asymmetric hearing loss on July 22, 2019. Because these patients were recently treated, we do not have long-term data on their hearing recovery. In the future, CI for ISSNHL may represent a shift in treatment paradigm and is an area under active investigation. More studies will need to be performed to better understand the optimal timing for CI placement after unilateral ISSNHL.

Yeo et al reported audiometric outcomes in 156 patients with ISSNHL who were followed for 3 months after treatment.²⁰ This study demonstrated that 35% of patients showed delayed recovery within 1 month of treatment. However, the degree of hearing loss was stabilized in 97% of patients within 3 months. Beyond this, there

are no data to date to guide the timing of follow-up. In the present study, we observe that at 18 months after diagnosis, less than half of our cohort had audiometric improvement. Most patients had one repeat audiometric test within 50 days of presentation, but pure tone audiometry was not consistently obtained in the long term. We did observe a slightly longer follow-up duration of 22.3 months in patients who did not experience hearing improvement, compared with those with hearing improvement (15.5 months). This difference was not statistically significant ($P = .217$). Subjectively, 79 patients (61.2%) reported none to partial hearing improvement at their most recent visit. The discrepancy between patient-reported hearing recovery (61.2%) and audiometric recovery rate (44.4%) was noteworthy. It is possible that some patients were satisfied with even mild hearing improvement after treatment, given the devastating nature of their sudden hearing loss. We also speculate that many patients developed adaptive strategies to cope with their hearing loss, thereby leading to an exaggerated perception of recovery. In addition to follow-up with otolaryngology and audiology, prior studies demonstrated the importance of rehabilitative counseling, speech reading, and auditory training in helping patients cope with the handicapping effects of sudden hearing loss.²¹ Altogether, our data corroborate with existing literature and the AAO-HNS CPG in suggesting ongoing needs for long-term follow up and medical, and possibly surgical interventions, for hearing restoration.²²

The strengths and limitations of the study should be considered. First, all patients in this cohort were treated and followed up by a consistent group of clinical providers at a single tertiary care institution. Their clinical care was carefully documented in the electronic medical record system. However, although this is one of the larger series to examine long-term outcomes of unilateral ISSNHL, our study is retrospective and cannot support causal inferences. Second, there are variations in our treatment protocols by individual otolaryngologists and audiologists. For example, our oral steroid regimen ranged from a 7-day course to a 14-day course and was followed by various permutations of slow vs fast steroid taper and doses. The publication of the AAO-HNS CPG on sudden hearing loss calls for standardization in treatment paradigm and hopes to address these practice variations in the future. Third, despite our effort to examine a longitudinal cohort over 20 years, the follow-up duration in this study was short. To better evaluate long-term outcomes, longitudinal studies are required to ensure adequate length of follow-up. Along the same lines, future studies are needed to better understand potential patient factors and systemic barriers to reduced follow-up and poor patient compliance with hearing amplification.

5 | CONCLUSION

In our cohort of patients who presented to a tertiary care medical institution with unilateral ISSNHL, we observed a 44.4% audiometric recovery on pure tone audiometry and 61.2% of patient-reported hearing improvement. Most patients at our institution experienced some degree of hearing recovery, but many had persistent hearing

loss after the initial episode of ISSNHL. Over the course of 20 years, the average follow-up duration was only 18 months. Despite a 44% audiometric recovery rate, long-term hearing amplification was recommended in less than half of the cohort and patient compliance with hearing amplification devices was poor. Future studies are needed to better assess long-term audiometric outcomes and to determine factors that contribute to poor follow-up patterns and compliance with hearing rehabilitation strategies.

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CONFLICT OF INTERESTS

Dr Terry Zwolan is on the advisory board of Cochlear Americas and Envoy Medical Corporation. For the remaining authors, none were declared.

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