



Research Paper

Improvement in word recognition following treatment failure for sudden sensorineural hearing loss



Taha A. Jan, Elliott D. Kozin, Vivek V. Kanumuri, Rosh K. Sethi, David H. Jung*

Department of Otolaryngology, Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston, MA 02114, USA

Received 9 May 2016; received in revised form 20 June 2016; accepted 29 June 2016
Available online 24 November 2016

KEYWORDS

Sensorineural hearing loss;
Sudden sensorineural hearing loss;
Word recognition score;
Pure tone average

Abstract *Objectives:* Patients with sudden sensorineural hearing loss (SSNHL) may have word recognition scores (WRS) that correlate with pure tone average (PTA). We hypothesize that there is a subset of patients with SSNHL who have improved WRS despite stable PTA. *Methods:* Retrospective case review at a tertiary otolaryngology practice. *Results:* We identified 13 of 113 patients with SSNHL whose WRS increased despite overall stable pure tone averages. There was an observed average improvement in WRS by 23.8 points in this patient cohort at follow-up, with mean initial PTA in the affected ear at 48.7 dB. *Conclusions:* We identify a novel cohort of SSNHL patients that have failed treatment as measured by PTA, but who have increased WRS over time. These data have implications for patient counseling and lend insight into the pathophysiology of SSNHL.
Copyright © 2016 Chinese Medical Association. Production and hosting by Elsevier B.V. on behalf of KeAi Communications Co., Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

* Corresponding author. Department of Otolaryngology, Massachusetts Eye and Ear Infirmary, Harvard Medical School, 243 Charles Street, Boston, MA 02114, USA. Fax: +1 617 573 3914.

E-mail address: David_Jung@meei.harvard.edu (D.H. Jung).

Peer review under responsibility of Chinese Medical Association.



Production and Hosting by Elsevier on behalf of KeAi

Introduction

Sudden sensorineural hearing loss (SSNHL) occurs with an incidence of 5–20 cases per 100,000 persons per year.¹ Classically, it is defined as new onset unilateral hearing loss that occurs within a time period of three consecutive days.^{2,3} In addition to sensorineural hearing loss (SNHL), patients can report aural fullness, tinnitus, and vestibular symptoms.^{4–6} One of the leading theories regarding the etiology of SSNHL includes an inflammatory insult related to a viral infection.^{7–14}

Numerous studies have investigated treatment options for SSNHL. Steroid therapy, via either oral corticosteroids or intratympanic injection, is the current standard of care for treatment of SSNHL.^{3,15–19} Formally speaking, however, the efficacy of steroid treatment for SSNHL has never been proven with a randomized, placebo-controlled trial.²⁰ Further confounding the picture, it has also been reported that 32%–65% of cases recover without any treatment.^{3,5,16}

Prior studies have principally utilized audiometric data, including pure tone averages (PTA) and word recognition scores (WRS) to quantify audiometric outcomes following SSNHL. In brief, the PTA is considered as the hearing sensitivity averaged over four standard frequencies: 500, 1000, 2000, and 4000 Hz.^{21,22} WRS is a measure of intelligibility of a standard list of monosyllables, measured as a percentage of correctly recognized words.^{3,23} In some studies, PTA and WRS have been found to track one another; WRS typically improves with decreasing PTA thresholds.¹⁵ Few studies, however, have addressed whether the converse clinical scenario may be true: can WRS significantly improve in the absence of PTA improvement?

Herein, we examine the hypothesis that there may be a subset of patients with SSNHL who have improved WRS despite stable PTA. Identification of this novel patient population has implications both for patient counseling and for the pathophysiology of SSNHL.

Materials and methods

Basic inclusion and exclusion criteria

We retrospectively analyzed patients with an ICD-9 code diagnosis of SSNHL (388.2) treated at our tertiary care center between 2011 and 2014. Inclusion criteria included at least 10 dB or greater difference in sensorineural hearing loss as measured by PTA in affected versus unaffected ears and follow-up of at least 60 days later with an audiogram. Exclusion criteria included first audiogram greater than 30 days post symptoms, Meniere's disease, otosclerosis, immune mediated SNHL, perilymph fistula, non-SNHL, ototoxicity, bilateral severe SNHL, congenital SNHL, mixed SNHL with conductive components, fluctuating SNHL, ipsilateral vestibular schwannoma, or patients without available audiograms from our institution. This study was approved by our Institutional Review Board (IRB), protocol #14-116H.

Out of the overall sample of patients with SSNHL, we aimed to identify patients who had stable PTA and improving WRS. To identify this cohort of patients, only

patients with at least 10 dB difference between affected versus non-affected ears' PTAs was examined. Additionally, we required patients to have follow-up in a 2–18 month period.

Audiometric data

Audiologists from our institution conducted all audiograms analyzed in this study. PTA was calculated in the standard method with averaging of thresholds at 500, 1000, 2000, and 4000 Hz.²⁴ WRS here were calculated based on four sets of 50 word monosyllable sets (CID W-22),^{3,24} which at our institution is presented in a pre-recorded format. An abbreviated version of these lists was utilized at our institution with a list of 10 words out of the 50 from each of the four sets that have been found to be highly predictive of a subject's WRS. If all 10 words were answered correctly, then a score of "pass" is registered. However, if one out of ten answers is incorrect, then the full 50-word list is tested as a default. This method involving an abbreviated list has been internally validated by our audiologists. Patients with WRS recorded as "pass" were therefore designated a value of 100% while those recorded as below threshold or no words recognized as 0%. All words used in the lists are open-set male English speakers tested usually at a threshold of 70 dB. PTAs recorded as above threshold were assigned a value of 100 dB.

Statistical analysis and reporting of audiometric data

We utilized a standardized method for reporting collective audiometric data as agreed upon by the Hearing Committee of the American Academy of Otolaryngology-Head and Neck Surgery.²⁵ Statistical analyses were carried out using Microsoft Excel (Redmond, Washington). Two tailed, paired student's *t*-test was utilized to generate *P*-values. We consider *P*-value less than 0.05 to be statistically significant. All \pm refer to standard deviation (SD) unless otherwise specified.

Results

Baseline sample of patients with SSNHL

We queried all electronic medical records at the study institution from 2011 to 2014 for patients with a primary or secondary ICD-9 diagnosis of 388.2, which generated a list of 569 patients. Fig. 1 is a flow chart that demonstrates inclusion and exclusion of study patients. Patients without study institution audiograms were excluded ($n = 74$). Any patient with an audiogram greater than 30 days from onset of symptoms and/or treatment was excluded ($n = 112$). Additionally, patients with Meniere's disease (or "cochlear hydrops") ($n = 16$), immune mediated SNHL ($n = 1$), ototoxicity ($n = 1$), vestibular schwannomas ($n = 2$), perilymph fistula ($n = 2$), otosclerosis ($n = 6$), bilateral severe SNHL ($n = 6$), congenital SNHL ($n = 2$), mixed SNHL and conductive hearing loss ($n = 8$), and fluctuating SNHL ($n = 10$) were all excluded (Fig. 1).

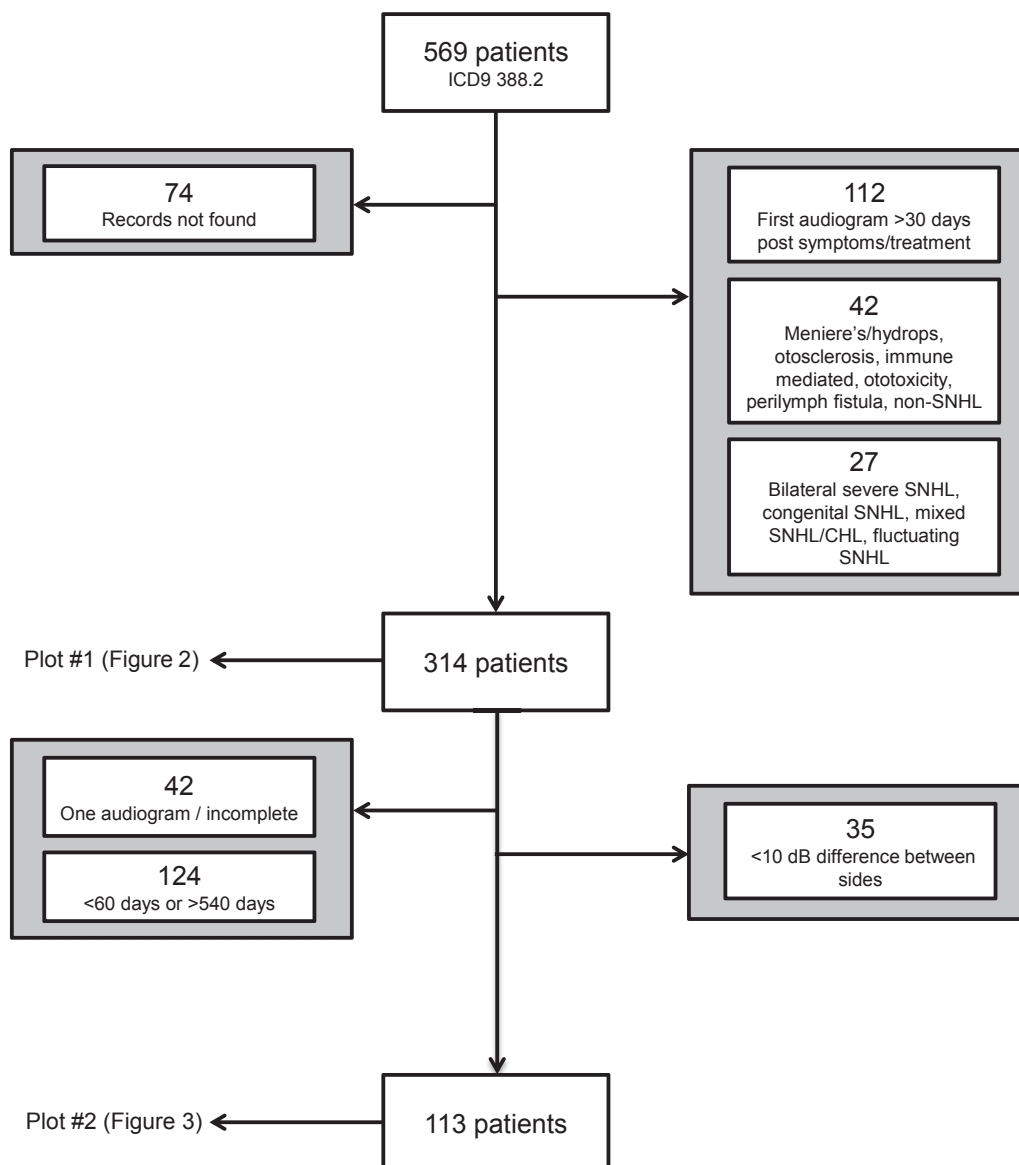


Fig. 1 Diagram of patients included and excluded in this study. All audiograms considered were obtained at our institution.

We identified a range of PTAs and WRS, generally in agreement with prior description of patients with SSNHL.¹⁵ Results of this first audiogram are plotted in Fig. 2. The average age of these patients was 56.1 ± 14.6 years old with a range from 8 to 90 years old. There was a male preponderance at 54.69% ($n = 175$), and 45.31% females ($n = 145$). The average initial WRS in the affected ear was $53.4 \pm 38.3\%$ ranging from 0% to 100% while the average PTA was 49.1 ± 29.3 dB ranging from 0 dB to 118 dB. The affected ear was found to have a significantly different PTA and WRS from the unaffected ear ($P < 0.001$).

Subgroup analysis of patients with improved WRS and stable PTA

We observed that the majority of patients improved both their WRS and their PTAs from initial audiogram (WRS $46.1 \pm 38.0\%$ and PTA 56.6 ± 26.9 dB) to final follow up audiograms (WRS $70.25 \pm 34.4\%$ and PTA 36.0 ± 27.5 dB)

(Fig. 3). This improvement in WRS and PTAs between initial audiogram to final follow up audiograms is significant ($P < 0.001$). While these patients improved their WRS and PTAs, we do not set a specific threshold for recovery being mindful of the floor effect observed when such thresholds for improvement are set.²⁶

Out of the original cohort of SSNHL patients, using the aforementioned inclusion and exclusion criteria we identified 13 patients with improved WRS despite stable PTA (Fig. 4). These patients had an average age of 60.6 ± 11.8 years with a range of 43–86 years old. There were 8 male and 5 female patients. The WRS of these patients increased on average by $23.8 \pm 9.8\%$ ($P = 0.031$), with a range of 12–42%. Although the starting WRS were across a range, these improvements are not likely to be due to chance.²⁶ For these patients, however, the PTAs remained essentially stable at an average of -0.8 ± 6.6 dB ($P = 0.92$) with a range of -13 to $+10$ dB (Fig. 5). We defined stable or worsening PTA here from -20 dB to $+10$ dB. As a

Word Recognition Score (%)

	100 - 90	89 - 80	79 - 70	69 - 60	59 - 50	49 - 40	39 - 30	29 - 20	19 - 10	9 - 0
0 - 10	24	1	2				1	1		
11 - 20	22	7	1	2					1	
21 - 30	28	9	4	4				2		
31 - 40	9	12	7	9	3	2		1	1	
41 - 50	1	4	5	2	8	2		4	3	
51 - 60	1		2	3	1	2	4	2	3	2
61 - 70		2	2	3	2	1	1	6	4	6
71 - 80				1	1		3	3	6	15
81 - 90			1				1	1	3	16
>91		1							2	31

Fig. 2 Word recognition scores and pure tone averages of affected side from patients with SSNHL on first audiogram presented in standard plot diagram as proposed by American Academy of Otolaryngology-Head and Neck Surgery Hearing Loss Scale.²⁵ Each box represents the number of patients that have a result within its designated box. *n* = 314.

Word Recognition Score (%)

	Improved					Worse					
	≥50	40	30	20	10	No Change	10	20	30	40	≥50
Improved ↑											
≥50	16	1	2								
40	1	3	3	2							
30	4	2	1	4	1	2	3				
20	4	1	3	6	8	3	1				
10	1		3	3	7		4	1	1		
No Change		1	1			3					
10		2		1	3	4	1	2		1	
20				1			1				
30							1				
40											
Worse ↓											
≥50											

Fig. 3 Changes in word recognition scores and pure tone averages comparing the initial audiogram at our institution and the last available audiogram (follow-up: 2–18 months). Presented in standard format as proposed by American Academy of Otolaryngology-Head and Neck Surgery Hearing Loss Scale.²⁵ *n* = 113.

		Word Recognition Score (%)										
		Improved					No Change	Worse				
		≥50	40	30	20	10	No Change	10	20	30	40	≥50
Pure Tone Average (dB)	Improved	≥50	16	1	2							
	40	1	3	3	2							
	30	4	2	1	4	1	2	3				
	20	4	1	3	6	8	3	1				
	10	1		3	3	7		4	1	1		
	No Change		1	1			3					
	10		2		1	3	4	1	2		1	
	20				1			1				
	30							1				
	Worse	≥50										

Fig. 4 Highlighted are two groups of patients for further analysis. In solid gray are patients whose WRS improved with overall stable PTAs ($n = 13$). The complementary group, hatched, is used as a comparison with worsening WRS and stable PTA ($n = 11$). Only 5 patients from this group were considered for further analysis as the remainder had starting WRS $\geq 90\%$.

comparison, there is a subset of patients (Fig. 4) with stable PTAs and worsening WRS in the same range as the previous group. From this cohort, we similarly excluded patients with initial WRS greater than or equal to 90% thereby identifying five patients in this category (Fig. 5).

This subgroup of patients have an average age of 65.6 ± 17.2 years with a range of 37–78 years old that include 2 male and 3 female patients. On average, these patients had a worsening WRS of $22.0 \pm 11.6\%$ ($P = 0.15$) ranging from 10% to 40% and PTA remaining stable at 0.2 ± 7.82 dB ($P = 0.99$) ranging from -10 to $+8$ dB. While these two patient groups are very limited in number, the average initial WRS and PTA between these two cohorts are not statistically different (WRS $P = 0.984$, PTA $P = 0.347$).

The subgroup of patients with improved WRS despite stable PTA presented with varying degrees of classic symptoms associated with SSNHL, including ear blockage, tinnitus, dizziness, and vertigo (Fig. 5). These patients presented to our institution up to four weeks from the onset of symptoms. With respect to treatments received, six patients received oral prednisone taper and either concurrent or salvage intratympanic steroid injections, while the remaining seven patients were treated with oral steroids only. The comparison group similarly presented with tinnitus, ear blockage, and dizziness. One patient was found to have an incidental small vestibular schwannoma on the non-SSNHL side. Two of these patients were treated with oral steroids alone, one with oral steroids followed by intratympanic steroid injection, and one with oral steroid followed by intravenous methylprednisolone. One patient declined treatment. Review of medications taken by patients in either group did not reveal any class of medication

that was unique to each in a significant way (data not shown). One patient from the first cohort did have a history of eustachian tube dysfunction requiring tympanostomy tubes. Additionally, we evaluated cardiovascular risk factors between the two groups, including hypertension, hyperlipidemia, diabetes mellitus, history of myocardial infarction, transient ischemic attacks, congestive heart failure, or smoking status. There was no clear difference between the two groups (data not shown).

Discussion

We have identified a small but distinct group of patients whose WRS increases despite stable or worsening PTAs. We found no obvious distinct feature of this patient population in comparison to a complementary group of patients without improved WRS (Figs. 4 and 5). Of note, we observed that six out of 13 patients who had improved WRS did receive both oral and intratympanic steroids (Fig. 5), although this number is too small to draw firm conclusions relative to the group without improved WRS. While the starting WRS for the patient population of interests were across a range, these improvements are unlikely to be due to chance.²⁶ Specifically, when analyzing this group of 13 patients, 11 of them met criteria for significant change in word recognition score as defined by a binomial model of variance based on the starting value of the initial WRS, as first described by Thornton and Raffin.²⁷

The identification of this patient cohort raises the question as to how the WRS might improve despite stable PTAs. Temporal bone histopathological studies may shed

	Age	Sex	Treatment	Dizziness	Initial WRS	Initial PTA	Last WRS	Last PTA	
WRS – improved PTA – Stable/Worsened	1	49	F	PO, IT	-	54	50	86	52
	2	53	M	PO	-	6	75	18	88
	3	55	F	PO (stopped)	-	12	80	24	88
	4	57	F	PO	+	22	48	56	52
	5	64	F	PO	-	48	52	80	52
	6	72	M	PO, IT	-	52	45	76	45
	7	75	M	PO	+	54	40	80	30
	8	54	M	PO, IT	+	86	30	98	20
	9	53	F	PO	-	86	15	100	8
	10	43	M	PO+IT	-	80	35	98	32
	11	61	M	PO, IT	-	68	28	90	25
	12	86	M	PO+IT	-	32	75	62	72
	13	66	M	PO	+	36	60	78	58
Average	60.6				48.9*	48.7	72.8*	47.8	
± S.D.	±11.8				±26.7	±19.8	±26.5	±24.9	
WRS – Worsened PTA – Stable/Worsened	1	62	M	PO, IV	-	20	70	10	60
	2	74	F	PO, IT	-	38	88	12	82
	3	37	F	PO	+	60	52	20	55
	4	77	F	PO	-	78	42	62	50
	5	78	M	Declined	+	50	42	32	48
Average	65.6				49.2	58.8	27.2	59.0	
± S.D.	±17.2				±21.9	±19.9	±21.3	±13.7	

Fig. 5 Selected group of patients from Fig. 4 with specific patient characteristics. The starting/initial audiogram WRS and PTA are listed along with the final WRS and PTA on last available follow up. “PO, IT” = PO steroids followed by salvage intratympanic steroid injection. “PO + IT” = PO and concurrent intratympanic injection. Averages for each column are listed. The WRS for the first cohort of patients was significantly improved from initial to last audiograms as indicated by asterisk ($P < 0.05$).

some light in this regard, as they have demonstrated that the primary site of damage in SSNHL is in hair cells and supporting cells of the organ of Corti.^{28,29} There are at least two potential mechanisms by which patients might experience improvement in WRS without improvement in PTA. One explanation is that these patients simply experience broader recovery within the cochlea. That is, although PTA does not significantly improve because the density of functional hair cells remains decreased on average around each of the tested frequencies, there is a greater density of hair cell recovery overall across the cochlea to result in improved WRS. Another explanation is that these patients might demonstrate a superior ability to decode the new, distorted signal presented to the brain from the affected ear. Such adaptation could be analogous to the process by which patients learn to interpret new signals presented to the brain following cochlear implantation,³⁰ and future treatments for SSNHL might therefore focus on similarly “re-training” the affected ear. Such approaches have been previously attempted for patients with hearing loss,³¹ although definitive studies remain to be performed.

There are several limitations of our study. First, we are unable to draw any conclusions from a statistical perspective about the two patient populations we identified. Second, this study does not define specific criteria for the diagnosis of SSNHL, instead relying on clinician’s reporting

based on a standardized coding system, ICD-9, that is tied to compensation for all providers. Third, in terms of WRS protocol, if the subject misses one of the initial 10 words, then the full 50-word list is tested. While this method has been internally validated by our audiologists, we recognize that fewer words may increase intersubject variability, and this is a limitation of this technique.²⁶ Finally, there may be more patients with increased WRS despite stable PTA that are lost to follow-up. Indeed, many patients did not have serial audiograms. It is possible that patients partially or completely recovered.

In summary, this study identifies a novel patient population that merits further investigation. It is clear that a small subset of patients have improved word recognition despite unchanged hearing thresholds. A large-scale study would lend more power to such an investigation. Such a study might allow for finer dissection of potential confounders or associated symptoms that could impact intervention.

Conclusion

We identify a novel cohort of SSNHL patients that have failed treatment as measured by PTA, but who have increased WRS. This patient cohort has important implications for patient counseling and understanding the etiology and natural history of SSNHL.

Financial disclosures

None.

Conflict of interest

None.

References

- Byl Jr FM. Sudden hearing loss: eight years' experience and suggested prognostic table. *Laryngoscope*. 1984;94:647–661.
- DeKleyn A. Sudden complete or partial loss of function of the octavus-system in apparently normal persons. *Acta Otolaryngol*. 1944;32:407–429.
- Chen CY, Halpin C, Rauch SD. Oral steroid treatment of sudden sensorineural hearing loss: a ten year retrospective analysis. *Otol Neurotol*. 2003;24:728–733.
- Shaia FT, Sheehy JL. Sudden sensori-neural hearing impairment: a report of 1,220 cases. *Laryngoscope*. 1976;86:389–398.
- Mattox DE, Simmons FB. Natural history of sudden sensorineural hearing loss. *Ann Otol Rhinol Laryngol*. 1977;86:463–480.
- Kallinen J, Laurikainen E, Bergroth L, Grenman R. A follow-up study of patients suffering from sudden sensorineural hearing loss. *Acta Otolaryngol*. 2001;121:818–822.
- Van Dishoeck HA, Bierman TA. Sudden perceptive deafness and viral infection; report of the first one hundred patients. *Ann Otol Rhinol Laryngol*. 1957;66:963–980.
- Jaffe BF. Viral causes of sudden inner ear deafness. *Otolaryngol Clin North Am*. 1978;11:63–69.
- Rowson KE, Hinchcliffe R. A virological and epidemiological study of patients with acute hearing loss. *Lancet*. 1975;1:471–473.
- Veltri RW, Wilson WR, Sprinkle PM, Rodman SM, Kavesh DA. The implication of viruses in idiopathic sudden hearing loss: primary infection or reactivation of latent viruses? *Otolaryngol Head Neck Surg*. 1981;89:137–141.
- Wilson WR, Veltri RW, Laird N, Sprinkle PM. Viral and epidemiologic studies of idiopathic sudden hearing loss. *Otolaryngol Head Neck Surg*. 1983;91:653–658.
- Woolf NK, Harris JP, Ryan AF, Butler DM, Richman DD. Hearing loss in experimental cytomegalovirus infection of the guinea pig inner ear: prevention by systemic immunity. *Ann Otol Rhinol Laryngol*. 1985;94:350–356.
- Adams JC. Clinical implications of inflammatory cytokines in the cochlea: a technical note. *Otol Neurotol*. 2002;23:316–322.
- Wilson WR. The relationship of the herpesvirus family to sudden hearing loss: a prospective clinical study and literature review. *Laryngoscope*. 1986;96:870–877.
- Rauch SD, Halpin CF, Antonelli PJ, et al. Oral vs intratympanic corticosteroid therapy for idiopathic sudden sensorineural hearing loss: a randomized trial. *JAMA*. 2011;305:2071–2079.
- Wilson WR, Byl FM, Laird N. The efficacy of steroids in the treatment of idiopathic sudden hearing loss. A double-blind clinical study. *Arch Otolaryngol*. 1980;106:772–776.
- Kopke RD, Hoffer ME, Wester D, O'Leary MJ, Jackson RL. Targeted topical steroid therapy in sudden sensorineural hearing loss. *Otol Neurotol*. 2001;22:475–479.
- Parnes LS, Sun AH, Freeman DJ. Corticosteroid pharmacokinetics in the inner ear fluids: an animal study followed by clinical application. *Laryngoscope*. 1999;109:1–17.
- Gianoli GJ, Li JC. Transtympanic steroids for treatment of sudden hearing loss. *Otolaryngol Head Neck Surg*. 2001;125:142–146.
- Wei BP, Stathopoulos D, O'Leary S. Steroids for idiopathic sudden sensorineural hearing loss. *Cochrane Database Syst Rev*. 2013;(7). CD003998.
- American National Standards Institute. *Methods for manual pure-tone threshold audiometry. ANSI S3.21-1978 R-1986 [report]*. New York: American National Standards Institute; 1978.
- Carhart R, Jerger J. Preferred method for clinical determination of pure tone thresholds. *J Speech Hear Dis*. 1959;24:330–345.
- Hirsh IJ, Davis H, Silverman SR, Reynolds EG, Eldert E, Benson RW. Development of materials for speech audiometry. *J Speech Hear Disord*. 1952;17:321–337.
- Halpin C, Shi H, Reda D, et al. Audiology in the sudden hearing loss clinical trial. *Otol Neurotol*. 2012;33:907–911.
- Gurgel RK, Jackler RK, Dobie RA, Popelka GR. A new standardized format for reporting hearing outcome in clinical trials. *Otolaryngol Head Neck Surg*. 2012;147:803–807.
- Halpin C, Rauch SD. Using audiometric thresholds and word recognition in a treatment study. *Otol Neurotol*. 2005;27:110–116.
- Thornton AR, Raffin MJ. Speech-discrimination scores modeled as a binomial variable. *J Speech Hear Res*. 1978;21:507–518.
- Merchant SN, Adams JC, Nadol Jr JB. Pathology and pathophysiology of idiopathic sudden sensorineural hearing loss. *Otol Neurotol*. 2005;26:151–160.
- Linthicum Jr FH, Doherty J, Berliner KI. Idiopathic sudden sensorineural hearing loss: vascular or viral? *Otolaryngol Head Neck Surg*. 2013;149:914–917.
- Moore DR, Shannon RV. Beyond cochlear implants: awakening the deafened brain. *Nat Neurosci*. 2009;12:686–691.
- Henshaw H, Ferguson MA. Efficacy of individual computer-based auditory training for people with hearing loss: a systematic review of the evidence. *PLoS One*. 2013;8:e62836.

Edited by Xin Jin