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Audiological outcomes in sudden sensorineural hearing loss with presumed inner ear hemorrhage

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ABSTRACT

Objective: To explore the hearing outcomes and prognostic factors in patients with sudden sensorineural hearing loss resulting from inner ear hemorrhage.

Methods: 42 patients (22 male and 20 female) were recruited from January 2016 to December 2017. Intravenous methylprednisolone and/or intratympanic corticosteroid were used as salvage therapy. The main measures included systemic risk factors and audiometric outcomes as proposed by American Academy of Otolaryngology-Head and Neck Surgery Hearing Loss Scale. All individuals were assessed at baseline, discharge (2 weeks post-treatment) and at 1, 3 and 6 months.

Results: The mean ages of patients were 39.3 ± 14.8 yrs. Cardiovascular disorders were seen in 19.0–33.3% of cases. Restoration of hearing and speech discrimination abilities were assessed at the first month post-treatment versus initial levels (95.5 ± 15.5 vs. 109.2 ± 9.6 dB, $p = 0.000$; and 17.6 ± 24.4 vs. 1.3 ± 4.0%, $p = 0.003$, respectively). Word recognition scores continued to recover at month 6 (38.7 ± 35.4%, $p = 0.000$), whereas puretone ceased to change (90.8 ± 16.2 dB, $p = 0.139$). The final percentages of complete, partial and no recovery were 0%, 57.1% and 42.9% respectively. The prognosis was independent of accompanying systemic risk factors as analyzed in this study. Intratympanic intervention was associated with improved word recognition scores, although intravenous corticosteroid was not.

Conclusions: Profound sudden sensorineural hearing loss caused by inner ear hemorrhage often has an unsatisfactory prognosis. However, this cohort did experience partial audiological recovery with delayed onset. Immediate and effective intratympanic corticosteroid may have therapeutic potential for this intractable disease.

1. Introduction

Inner ear hemorrhage (IEH) is increasingly recognized as a cochlear lesion that can cause sudden sensorineural hearing loss (SSNHL) [1–6]. Since it is often subclinical, SSNHL may be under-diagnosed without immediate magnetic resonance imaging (MRI) scanning. MRI results characteristic of IEH are hyperintensity within the intralabyrinthine portions on T1-weighted and 3D-fluid-attenuated inversion recovery (FLAIR) images [1–6]. Recent studies into the temporal bone have explored the pathologic process of this condition, and found that cochlear sections showed significant losses of outer hair cells (OHCs) and endolymphatic hydrops in cases of IEH [7,8].

Several studies stated the characteristic hearing results of IEH with limited numbers of cases [1,3–6]. A study of seven patients with IEH found their initial and final hearing levels to be 72 ± 26.8 dB and 82.2 ± 41 dB, respectively, and concluded that IEH was prognostic of poor recovery [1]. Another report, which evaluated 12 patients, found that both initial and final hearing levels were significantly greater in FLAIR-positive patients compared to FLAIR-negative patients (98.0 ± 31.1 dB vs. 64.1 ± 26.7 dB; 30 days after treatment 87.7 ± 33.1 dB vs. 30.2 ± 24.0 dB) [9]. Our previous study of seven patients suggested that vestibular dysfunction and sudden profound hearing loss may also indicate IEH [4].

Despite these findings, the chance of recovery of hearing and auditory perception through conventional interventions after IEH is largely unknown, and an effective treatment for this disease is yet to be established. In this study, we collected data from 42 patients with SSNHL following presumed IEH in our center, and followed up the hearing outcomes for six months. Prognostic factors and possible therapy regimes were investigated.

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2. Materials and methods

The study was approved by the institutional review board of the First Affiliated Hospital of Sun Yat-sen University. We retrospectively analyzed results in patients with SSNHL, and IEH suggested by 3D-FLAIR MRI findings, in our institution from January 2016 to December 2017. Patient inclusion criteria were based on the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) guidelines definition of SSNHL [10].

In this study, all patients received regular treatment once the diagnosis of SSNHL was confirmed. Intravenous (IV) Methylprednisolone (48–60 mg/d) in a single dose was prescribed for 7 days, then tapered over a similar time period during hospitalization. Oral steroid was not prescribed in this study as it has previously been associated with unsatisfactory results in IEH patients [4]. Intratympanic (IT) corticosteroids were used in patients who agreed to this as a salvage therapy: an IT injection of 0.4–0.8 ml Methylprednisolone (40 mg/ml) every 3 days for 3 consecutive sessions was arranged. Patients were followed up for at least 6 months.

Hearing outcomes were measured following the recommendations of the AAO-HNS guideline [10] and included scattergrams, word recognition scores (WRS) and pure-tone audiometry (PTA): PTA average thresholds were calculated in the conversational frequencies (0.5, 1, 2 and 4 kHz). Audiometric evaluations were performed at the time of discharge (2 weeks) and repeated at 1, 3 and 6 months post-treatment. Response to therapy was categorized as “Complete recovery”, “Partial recovery” or “No recovery” as set out in AAO-HNS guideline [10].

Statistical tests included the chi-square test, Fisher exact test and Student's *t*-test as appropriate. All statistical analyses were performed using SPSS version 23.0 (IBM Corp., Armonk, New York, USA); *p*-values < 0.05 were considered statistically significant.

3. Results

A total of 42 adult patients (42 ears) who met the criteria for SSNHL and IEH were included in the study (Fig.1, Table 1). The average age of patients was 39.3 ± 14.8 yrs. Cardiovascular risk morbidities (e.g. hypertension, hyperlipidemia or diabetes mellitus) were recorded in 19.0–33.3% of individuals. No other severe systemic diseases (renal, hepatic, hematologic, pulmonary or central nervous system disorders) or comorbidities (such as chemotherapy history, drug addiction) were present in this cohort. All the patients reported experiencing episodes of vertigo/dizziness.

At 6 months, the mean PTA and WRS were 90.8 ± 16.2 dB and 38.7 ± 35.5%, statistically better than at baseline (109.2 ± 9.6 dB, $p = 0.000$; and 1.3 ± 4.0%, $p = 0.000$, Fig. 2). In terms of PTA recovery, an average regain of 13.8 ± 11.3 dB was seen during the first month (95.5 ± 15.5 dB), which then remained stable during the third and sixth months. This observation was consistent with the change in the tested frequencies in PTA (250 Hz–8000 Hz). A difference was seen between baseline and 6 months for each frequency, but only the 125 Hz frequency improved at each follow up interval. WRS was 17.6 ± 24.4% at one month post treatment, and then showed a progressive recovery from month 3 to 6. At month 6 WRS was significantly better than at month 1 ($p = 0.000$). Half of these patients (21/42) achieved serviceable hearing that would be a candidate for traditional hearing amplification [10].

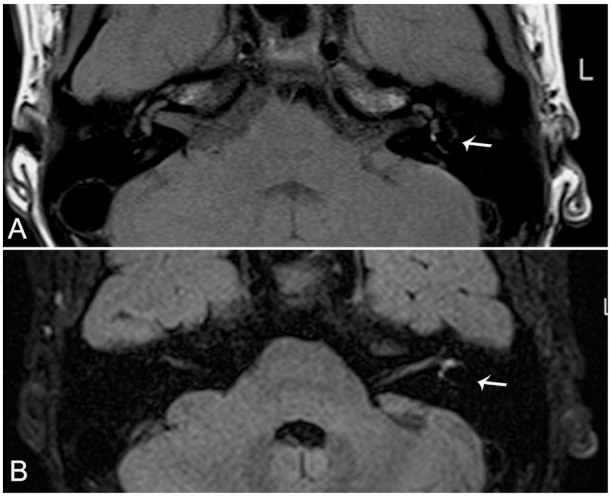

Fig. 1. Example of IEH as demonstrated by MRI. White arrows indicated hyperintensitites in T1WI (A) and T2WI-FLAIR (B).

The results were further analyzed via sub-categories. First, we investigated whether the addition of IT to IV steroid (IV + IT group, $n = 18$) affected the outcomes compared to IV treatment alone ($n = 24$). The onset of therapy was similar (7.3 ± 4.9 and 7.7 ± 5.2 days respectively, $p > 0.05$), as was initial hearing level (108.1 ± 9.9 and 110.1 ± 9.4 dB respectively, $p > 0.05$). The average post-treatment recovery at each frequency in either group was significant. Although IT did not seem to provide any additional therapeutic effect in PTA, it did improve WRS at 1, 3 and 6 months (Fig. 3). The percentages of serviceable hearing in both groups differed significantly (33.3% for the IV group and 72.2% for the IV + IT group, $p < 0.05$).

Finally, all patients were subdivided into categories based on recovery results proposed by the AAO-HNS hearing loss scale. The percentages of Complete, Partial and No recovery were 0%, 57.1% and 42.9%, respectively. The 24 patients with a partial recovery in PTA and/or WRS were assigned to Group A, while the remaining 18 individuals with no recovery were assigned to Group B (Fig. 2D and Table 1). A few possible risk factors were analyzed, but none seemed to affect the outcomes.

4. Discussion

A series of previous case studies suggested that IEH results in hearing loss with sudden onset. However, the sample sizes were small and the follow-up period was short [1,2,4–6,9]. Only 1 study has investigated long-term audiologic outcomes in IEH patients; of the 11 studied, most experienced profound irreversible sudden deafness which did not improve over the 13.3-month follow-up [3]. Our study monitored the largest cohort with IEH to date over a relatively long follow-up (at least 6 months).

The prognosis of IEH is different in adults and children [5]. A continued recovery of hearing and speech discrimination was noted in 6 children with presumed IEH at 6 months post-treatment (19.3 dB in PTA and 27.5% in WRS, respectively). In contrast, adult patients' improvement in hearing threshold 1 month post-treatment was worse than in children (13.8 ± 11.3 vs. 30.6 ± 11.3 dB). While PTA remained stable during the 5 months post-treatment in adults, in children this was found to recover. Therefore, there may be a “window period” for hearing recovery in adults, and watchful waiting for 1 month may be necessary.

Another finding from our study was the delayed improvement of speech recognition, although the audiometric threshold showed no further change at 6 months. Speech restoration after SSNHL onset has previously been addressed [3,5,11–13]. For example, Noguchi et al. retrospectively examined 20 patients aged 24 to 71 with idiopathic SSNHL from the period of 1–3 months to 11–13 months. Nine adults showed an increase of 10–19% in maximum speech discrimination score, in which an increase of > 20% was seen in four of them [12]. Likewise, in a study of 113 patients, Jan et al. identified 13 individuals with WRS recovery, despite no improvement in PTA in the affected ear (48.7%). During a 2–18 month follow up, an average increase of 23.8% in WRS was observed [11]. Our investigation in IEH was similar to

Table 1
Basic data and comparison within groups.

	Total	Subgroups		Reference value	Statistical comparison[a]
		Group A	Group B		
Gender (M:F)	22:20	9:15	13:5	–	NS
Age (years)	39.3 ± 14.8	39.3 ± 16.5	39.4 ± 12.7	–	NS
Onset of therapy (days)	7.5 ± 5.1	8.2 ± 4.8	6.7 ± 5.4	–	NS
Tinnitus rate (%)	90.5	91.7	94.4	–	NS
Ear fullness (%)	61.9	58.3	66.7	–	NS
Blood pressure (mmHg)	120.8/74.8	121.2/74.9	120.4/74.7	130/80	NS
Hypertension %	19.0	25	11.1	–	NS
Cholesterol (mmol/L)	5.3 ± 0.9	5.5 ± 1.0	5.1 ± 0.7	3.1–5.7	NS
Hypercholesterolemia %	19.0	20.8	16.7	–	NS
Triglyceride (mmol/L)	1.5 ± 1.2	1.7 ± 1.4	1.3 ± 0.9	0.33–1.70	NS
Hypertriglyceridemia %	21.4	29.2	11.1	–	NS
Blood glucose (mmol/L)	5.5 ± 1.3	5.4 ± 1.3	5.8 ± 1.3	2.9–6.0	NS
Diabetes mellitus %	33.3	29.2	38.9	–	NS
APTT (seconds)	28.1 ± 4.9	28.1 ± 4.8	28.1 ± 5.2	25–35	NS
PT (seconds)	11.6 ± 0.8	11.6 ± 0.9	11.5 ± 0.6	11–14	NS
Fbg (g/L)	2.5 ± 0.8	2.7 ± 1.0	2.2 ± 0.5	2–4	NS
Neutrophil ($X\,10^9$/L)	6.8 ± 3.6	6.7 ± 3.8	6.9 ± 3.4	1.8–6.4	NS
Lymphocyte ($X\,10^9$/L)	1.6 ± 0.8	1.6 ± 0.9	1.5 ± 0.7	1–3.3	NS
NLR	5.6 ± 3.4	5.8 ± 3.7	5.4 ± 3.1	–	NS

[a] Comparison among total and subgroups analysis; APTT: Activated partial thromboplastin time; NLR: Neutrophil/Lymphocyte ratio; NS: not significant; PT: Prothrombin time.

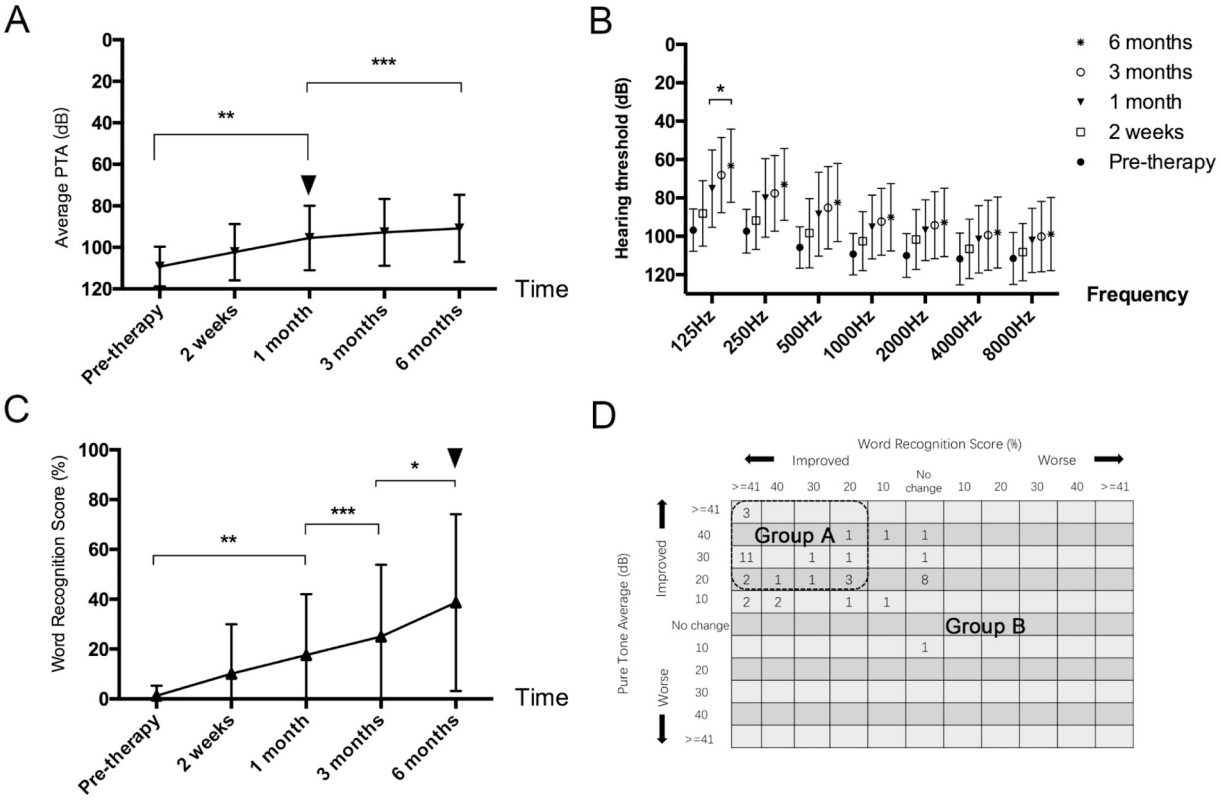

Fig. 2. Observed audiometric improvements. The arrowheads in (A) and (C) denote the periods when PTA and WRS were stable. (D) shows the post-treatment scattergram at month 6. Patients in the dotted box are those with partial recovery (Group A) and those with no recovery are Group B. The hearing thresholds are shown as mean ± standard deviation. $*p < 0.05$, $**p < 0.01$, $***p > 0.05$.

preceding studies in suggesting potential recovery, either spontaneously or via corticosteroid intervention.

IT corticosteroid is used widely but varies between studies. Regarding the lack of evidence or guidelines for IEH-related SSNHL [10], we adopted IT and/or IV for this cohort. Neither combination therapy nor IV corticosteroid alone was successful in ensuring a complete recovery. We did find that the low and middle frequencies were more likely to improve than the higher frequencies, particularly in those who received IT treatment. This indicated less impairment in the residual numbers/function of OHCs in the middle to apical turn. In terms of the initial profound hearing loss in IEH, there is still insufficient evidence to conclude that either treatment is ineffective. The present results implied that immediate and effective intratympanic corticosteroid may be a potential remedy for IEH. Although potential attributable risk factors had been taken into account, the mechanism of prognosis in IEH remains unclear.

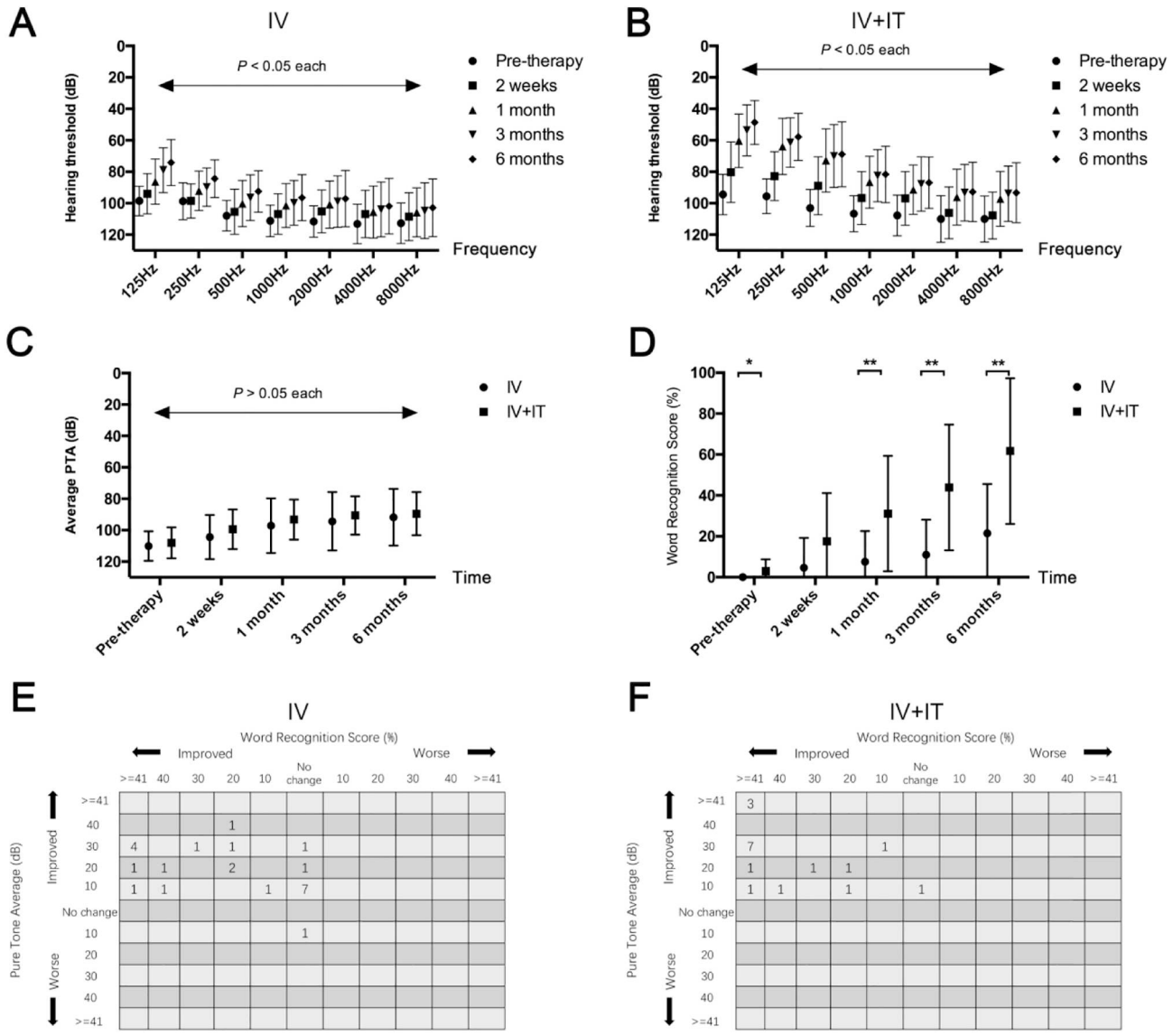

Fig. 3. PTA and WRS results comparison according to different treatments (IV or IV plus IT). The hearing thresholds are shown as mean ± standard deviation. $^{*}p < 0.05$, $^{**}p < 0.01$.

Two hypothetical mechanisms have been proposed to explain why patients can show audiometric improvement [11,12]. First, histopathological studies on temporal bone from patients with IEH showed endolymphatic hydrops and significant loss of OHCs, similar to that shown in Meniere's disease [7]. The absorption of accumulated hemoglobin and secondary excessive endolymph into the intralabyrinthine spaces may alleviate inner ear inflammation, and lead to subsequent hearing preservation and WRS increase. Another explanation is the compensational ability of cochlea function and/or neuroplasticity. Compensatory mechanisms may allow the patient to regain the ability to decode distorted signals sent from the affected ear to the brain; a process analogous to learning to interpret new signals following a cochlear implant [14].

The retrospective nature of the study does reduce its strength, and therefore further investigation of efficient therapy strategy is needed. A systematic, multi-center analysis would also help us to better understand the impact of IEH on hearing consequences.

5. Conclusion

Although IEH caused profound SSNHL with an unsatisfactory prognosis, partial audiological recovery with delayed onset occurred in this cohort. Immediate and effective intratympanic corticosteroid may have therapeutic potential for this intractable disease.

Conflict of interest

None.

Acknowledgement

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References

[1] Cho J, Cheon H, Park JH, et al. Sudden sensorineural hearing loss associated with inner ear lesions detected by magnetic resonance imaging. PLoS One 2017;12(10):e186038.

[2] Kim DS, Park DW, Kim TY, et al. Characteristic MR findings suggesting presumed labyrinthine hemorrhage. Acta Otolaryngol 2017;137(12):1226–32.

[3] Vivas EX, Panella NJ, Baugnon KL. Spontaneous Labyrinthine Hemorrhage: A Case Series[J]. 194599818785900 2018 Jun 1. https://doi.org/10.1177/0194599818785900.

[4] Wu X, Chen K, Sun L, et al. Magnetic resonance imaging-detected inner ear hemorrhage as a potential cause of sudden sensorineural hearing loss. Am J Otolaryngol 2014;35(3):318–23.

[5] Wu X, Jiang H, Wen L, et al. Delayed recovery in pediatric sudden sensorineural hearing loss predicted via magnetic resonance imaging. Ann Otol Rhinol Laryngol 2018;127(6):373–8.

[6] Lee JW, Park YA, Park SM, et al. Clinical features and prognosis of sudden sensorineural hearing loss secondary to Intralabyrinthine hemorrhage. J Audiol Otol 2016;20(1):31–5.

[7] Kaya S, Hizli Ö, Schachern PA, et al. Effects of intralabyrinthine hemorrhage on the cochlear elements. Otol Neurotol 2016;37(2):132–6.

[8] Kaya S, Paparella MM, Cureoglu S. Histopathologic changes of human vestibular epithelia in intralabyrinthine hemorrhage. Ann Otol Rhinol Laryngol 2017;126(6):445–50.

[9] Ryu IS, Yoon TH, Ahn JH, et al. Three-dimensional fluid-attenuated inversion recovery magnetic resonance imaging in sudden sensorineural hearing loss: correlations with audiologic and vestibular testing. Otol Neurotol 2011;32(8):1205–9.

[10] Stachler RJ, Chandrasekhar SS, Archer SM, et al. Clinical practice guideline: sudden hearing loss. Otolaryngol Head Neck Surg 2012;146(3 Suppl):S1–35.

[11] Jan TA, Kozin ED, Kanumuri VV, et al. Improvement in word recognition following treatment failure for sudden sensorineural hearing loss. World J Otorhinolaryngol Head Neck Surg 2016;2(3):168–74.

[12] Noguchi Y, Takahashi M, Ito T, et al. Delayed restoration of maximum speech discrimination scores in patients with idiopathic sudden sensorineural hearing loss. Auris Nasus Larynx 2016;43(5):495–500.

[13] Yeo SW, Lee DH, Jun BC, et al. Hearing outcome of sudden sensorineural hearing loss: long-term follow-up. Otolaryngol Head Neck Surg 2007;136(2):221–4.

[14] Moore DR, Shannon RV. Beyond cochlear implants: awakening the deafened brain. Nat Neurosci 2009;12(6):686–91.