Magnetic Resonance Imaging in Sudden Sensorineural Hearing Loss. Time to Talk.

Abstract

Sudden sensorineural hearing loss (SSHL) is defined as acute hearing loss of the sensorineural type of at least 30 decibels over three contiguous frequencies that occurs within a 72 hour period. Although many different causative factors have been proposed, SSHL is still considered "idiopathic" in 71-85% of cases, and treatments are empiric, not based on etiology. Magnetic Resonance Imaging (MRI) implemented with a 3D Fluid Attenuated Inversion Recovery (FLAIR) sequence has provided new insights into SSHL etiology. Herein, we review the current management trends for patients with SSHL, from the initial clinical diagnosis to therapeutic strategies and diagnostic work-up. We focus primarily on MRI assessment and discuss the relevance that MRI findings might have for patient management, pointing out different perspectives for future clinical research.

Abbreviations: ABR = Auditory Brainstem Response; BLB = Blood-Labyrinth Barrier; cVEMPs = Cervical Vestibular Evoked Myogenic Potentials; SSFP = steady state free procession; SSHL = Sudden Sensorineural Hearing Loss; SPIR = Spectral Presaturation with Inversion Recovery; TEOAE = Transiently Evoked Otoacoustics Emissions.

Introduction

According to the guidelines of the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology, sudden sensorineural hearing loss (SSHL) is defined as acute hearing loss of the sensorineural type of at least 30 decibels (dB) over three contiguous frequencies occurring within a 72-hour period [1]. SSHL occurs most often in the fourth decade of life, with an annual incidence that varies between 5 and 20 cases among 100,000 individuals. The severity of hearing loss is variable and is usually unilateral, although bilateral involvement has been reported in up to 4% of patients [2].

Although SSHL has been attributed to many different causative factors [3], there are still no audiological means of accurately assessing its etiology [4], and so it is still considered "idiopathic" in 71-85% of cases [5]. Consequently, there is not one treatment that targets the etiology, which partly explains the inconsistent results of the various empiric drug protocols proposed, which include systemic steroids, antiviral medications, vasodilators, carbogen or hyperbaric oxygen alone or in combination, none of which has been proved to be superior to the others [6-8].

Over the only last decade, MRI has provided new insights about SSHL etiology due to high-resolution sequences that are able to detect subtle changes in the inner ear [9-15]. However, for many reasons, the use of this imaging technique for SSHL is still not widespread. On one side, many radiologists are not yet aware of the possibilities offered by advanced MRI studies of the inner ear and/or of the possible MRI findings in SSHL. Secondly, it is still unclear and is not yet addressed in the literature how MRI could change therapeutic strategies.

In this paper, we review the current management trends of patients with SSHL, from the initial clinical diagnosis to the therapeutic strategies, through the diagnostic work-up. Our work focuses particularly on MRI assessment. We discuss how MRI should be performed and the relevance that the possible MRI findings in the inner ear might have. Finally, we address the perspectives of possible changes in the therapeutic management of SSHL based on MRI findings, which could impact the patient's prognosis.

Current diagnostic work-up of SSHL

SSHL is suspected in patients with a sudden onset of generally unilateral decrease or loss of hearing, occurring instantaneously or rapidly developing over a period of hours or days [3]. In other instances, primary care or emergency room physicians refer patients to the audiologist [16,17].

Evaluation usually begins with a careful history and physical examination to look for potential causes such as infections, systemic diseases and exposure to known ototoxic medications. Otomicroscopy is generally negative for external and middle ear pathologies. Pure-tone audiometry is the main diagnostic tool used to differentiate between conductive and sensorineural hearing loss, as they have very different management strategies. The physician can

differentiate SSHL (symptoms occur within 3 days) from progressive or fluctuating SSHL. The presence of bilateral sudden hearing loss, recurrent episodes of sudden hearing loss, or focal neurological findings suggest systemic disorders, autoimmune or metabolic disorders, bilateral Ménière's disease, or primary neurological disorders.

If SSHL is diagnosed, an empiric treatment is started while the diagnostic work-up continues. A complete audiovestibular evaluation, including speech audiometry, speech in noise test, tympanometry, acoustic reflexes, and otoacoustics emissions, is performed in an attempt to investigate whether the SSHL is cochlear or retrocochlear [8].

A brain MRI with and without gadolinium is often used to exclude a vestibular schwannoma (reported in up to 10-20% of patients with SSHL) as well as rarer causes of retrocochlear hearing loss such as other cerebellopontine tumours, brainstem infarctions and demyelinating disease [3,8,18-21]. A number of studies have advocated the use of MRI without gadolinium as the more appropriate means of screening patients with asymmetric SSHL suspected to be retrocochlear [22].

In cases of suspected cochlear SSHL, different studies have proposed a tailored temporal bone MRI **with 3D-FLAIR** sequence to exclude abnormalities in the inner ear structures [9-15]. MRI of the temporal bone can be negative, and the SSHL is therefore defined as idiopathic, or the MRI can show abnormalities in the inner ear structures, suggesting a specific etiopathogenesis (vascular or inflammatory) [14]. In both cases, as no trial has yet investigated different medical protocols guided by MRI findings, physicians continue with empiric therapy and so temporal bone MRI with **3D-FLAIR sequence** has not had any effect on the therapeutic management of cochlear SSHL in clinical practice. **Nevertheless, in our opinion, it is important that radiologists perform temporal bone MRI with 3D-FLAIR sequence in order to clarify the cochlear origin of SSHL, suggesting the probable pathogenesis, and provide prognostic information to physicians.**

How we perform MRI in SSHL patients

MRI of the temporal bone is challenging because of the complexity and small dimensions of the anatomic structures. Thus, MRI should preferably be performed on a 3 Tesla (T) scanner, which provides high-resolution images with a higher signal-to-noise ratio compared to a 1.5 T scanner. The basic MRI protocol should include an axial pre-contrast 3D Steady-State Free Precession (SSFP) sequence, a pre- and post-contrast T1-weighted FSE sequence, and a pre- and post-contrast 3D-FLAIR sequence [4-15]. Table 1, in supplementary material, summarizes the MRI scan parameters used in our department. It is recommended to use both post-contrast 3D-FLAIR and post-contrast T1-weighted FSE sequences because the former are more sensitive than T1-weighted sequence in detecting intralabyrinthine contrast enhancement [11,14,23], and the latter can clarify the presence of a small schwannoma.

Post-contrast sequences should be acquired approximately 10 minutes after contrast agent administration [13,15]. To cover part of the 10 minutes needed before the acquisition of post-contrast sequences, it is advantageous to inject a contrast agent before the acquisition of the 3D-SSFP sequence. **Although the contrast-enhancement of some structures can be seen on 3D-SSFP sequence [24], the assessment of the inner ear anatomic structures and internal auditory canal on this sequence is usually not hindered by the presence of the contrast agent.**

What can MRI detect?

Neuroradiologists should review MRIs and look for asymmetry of the signal between the affected and unaffected sides. In the literature, abnormalities on MRI are reported in 27% to 53% of SSHL cases [10,14].

Two patterns can be recognized based on the MRI signal of the inner ear on pre-contrast T1-weighted and 3D-FLAIR images:

1) The vascular pattern shows hyperintensity on pre-contrast T1-weighted and 3D-FLAIR images due to the presence of methemoglobin in the inner ear (Figure 1)

2) The inflammatory pattern shows hyperintensity only on 3D-FLAIR images due to the presence of proteinaceous exudate in the inner ear (Figure 2).

Regarding the vascular pattern, radiologists should be aware that pseudohyperintensity of the intralabirinthine fluid can normally be detected on fat-suppressed T1-weighted images, which could hamper the diagnosis. This pseudohyperintensity has previously been described as an artifact [**25**] and can be differentiated from methemoglobin because it is symmetrical and less evident on post-contrast fat-suppressed T1-weighted images (see Figure 3 for a detailed explanation).

Recent studies have investigated the relationship between pre-contrast 3D-FLAIR signals and clinical findings regardless of the hyperintensity on pre-contrast T1-weighted sequences [9-11,23]. The rate of abnormalities on pre-contrast 3D-FLAIR images correlated with the level of hearing loss at onset, resulting in fewer abnormalities in patients with mild to moderate hearing loss compared to patients with profound hearing loss [10,14,15]. However, Yoshida et al. did not observe similar results [11]. Inflammatory diseases can also affect the vestibule, the semicircular canals or the VIII cranial nerve, and pre-contrast 3D-FLAIR images can detect signal abnormalities in these structures (Figure 3) [10-14,23]. Hyperintensity on pre-contrast 3D-FLAIR images of the vestibule or the semicircular canals has been associated with vertigo [11,14], which is reported in approximately 30% of SSHL cases [9].

The two MRI patterns are not always associated with inner ear enhancement on post-contrast 3D-FLAIR, which is consistent with Blood-Labyrinth Barrier (BLB) breakdown [14]. The advantage of post-contrast 3D-FLAIR images is that they identify patients with more severe BLB breakdown when signal abnormalities are subtle on pre-contrast 3D-

FLAIR images [23]. However, the significance of the inner ear enhancement is still unclear. Viral infection, immune-mediated inner ear disease and perilymphatic fistulas have been suggested as possible causes of cochlear enhancement [**26,27**].

Usually, the 3D T2-weighted Steady State Free Procession (SSFP) sequence does not show any pathological findings [23]; nevertheless, it is essential to investigate the morphology of the inner ear structures, VIII cranial nerve, internal auditory canal and cerebello-pontine angle.

Prognostic value of MRI

To the best of our knowledge, no published studies have investigated the difference in prognosis and outcome between the two MRI patterns mentioned above, although a vascular pattern has been associated with a poor prognosis [**28**]. On the contrary, the published studies have concentrated on the prognostic value of 3D-FLAIR abnormalities and reported conflicting results**.** Two studies showed that a high signal in the affected inner ear on pre-contrast 3D-FLAIR was associated with a poor prognosis [11,15]. In two other studies, the hearing outcome was worse in patients with multiple-location hyperintensities on pre-contrast 3D-FLAIR (cochlea plus vestibule) than in patients with a single-subsite hyperintensity (cochlea only) [12,**29**]. **A recent study by Liao *et al*. showed that the more asymmetric FLAIR signal between the affected ear and the normal one and presence of high signal beyond the cochlea indicated a poorer prognosis [23].** Lee et al. demonstrated that pre-contrast 3D-FLAIR abnormalities do not affect the prognosis when the initial hearing loss is mild to moderate, while such abnormalities represent a negative prognostic factor in patients with initial profound hearing loss [10]. However, Berrettini et al. failed to find a correlation between the severity of pre-contrast 3D-FLAIR abnormalities and hearing improvement [14]. Lee et al. reported that high signal on pre-contrast 3D-FLAIR did not significantly affect the final hearing ability [9].

These inconsistent results are partially explained by methodological differences, including time span between SSHL onset and MRI, MRI assessment method, pharmacological protocols of drug administration during follow-up, length of follow-up, and assessment criteria used to determine hearing improvement [**30**]. A meta-analysis by Gao et al. that included studies of patients without primary treatment before temporal bone MRI concluded that pre-contrast 3D-FLAIR hyperintensity in the inner ear is associated with more severe initial hearing loss and a lower chance of recovery [**30**].

Current management of SSHL and perspectives in MRI research

The results of SSHL treatment are still largely unpredictable; a very large variability in responses has been reported, ranging from no response to total recovery [7,**31**]. Moreover, the high rate of spontaneous recovery, which varies from 45% to 65% [21], should be considered.

Although cochlear vascular micro-thrombosis has been hypothesized as the main pathogenic mechanism [**32,33,34**], since there is no objective test that can detect the occlusion of micro-vessels, the pharmacological treatment remains highly empirical and its overall efficacy is controversial owing to the absence of prospective double-blind studies [3]. Pharmacological treatment comprises many drugs that, without a certain etiology of SSHL, are often prescribed in combination: oral and/or intratympanic corticosteroids, hyperbaric oxygen therapy, antivirals drugs, vasodilators and vasoactive substances.

For decades, the gold standard treatment for SSHL has been the oral administration of corticosteroids [**35,36**]. The exact mechanism by means of which steroids improve hearing is still unknown, although some of the major hypotheses are that they modulate cochlear function, decrease inflammation and edema, improve cochlear blood flow, and protect against cochlear ischemia. Although the evidence concerning the use of oral corticosteroids remains contradictory [**37**], it seems reasonable to offer it because of the potentially devastating disability caused by SSHL and the relatively low morbidity of the treatment [3]. Intratympanic steroid treatment allows high steroid concentrations to be reached in the perilymph while avoiding the common side effects of systemic steroids. A recent meta-analysis has shown the benefit of intratympanic steroid treatment in combination with oral corticosteroids as first-line therapy, and salvage monotherapy in idiopathic SSHL [**38**]. Hyperbaric oxygen treatment is also used as primary, adjuvant or salvage therapy because it is thought to protect hair cells from ischemic damage by increasing oxygenation [**39,40**]. However, cost/benefit issues, limited availability, and the absence of strong evidence because of the difficulties of organizing controlled randomized studies have halted its widespread application. Although there is no clear evidence supporting their use, antiviral drugs are often used in pharmacological protocols for the treatment of SSHL because various viruses have been implicated in its etiology [**41**]. Vasodilators and vasoactive substances are sometimes used because obstructed vascular flow to the cochlea is a theoretical cause of SSHL, but there is currently insufficient evidence to support their routine use [3].

Future research should investigate the role of MRI with 3D-FLAIR sequence in the therapeutic management of SSHL (Supp. Fig. 4). Before future trials can investigate different medical protocols guided by MRI findings, longitudinal studies should investigate the effect of early steroid treatment and the time interval between disease onset and scanning on the sensitivity of MRI. In our opinion, its optimal timing would be upon admission to the emergency ward or on the same day as a clinical assessment by an audiologist or otolaryngologist. We hypothesize that pre-contrast and contrast-enhanced MRI abnormalities are more easily detected before steroid

treatment is started, and so MRI should be performed as soon as possible, and preferably before the masking effect of steroids becomes apparent [28]. In line with this suggestion, Berettini *et al*. have reported that the time interval between SSHL onset and MRI tended to be shorter in patients with 3D-FLAIR abnormalities than in those without 3D-FLAIR abnormalities (p = 0.06) [14]. However, this has not been confirmed by other authors [10,11].

Conclusion

MRI with 3D-FLAIR sequences provide new insights into SSHL etiology and may change current clinical and therapeutic practices. Radiologists should therefore be trained to perform tailored temporal bone MRI in the case of SSHL, and recognize the common findings and pitfalls of the technique in order to provide clinicians with information useful for patient management.

REFERENCES

1. Rauch SD. Idiopathic sudden sensoneurinal hearing loss. N Eng J Med 2008;359(8):833–40
2. Byl FM Jr. Sudden hearing loss: eight years' experience and suggested prognostic table. Laryngoscope. 1984;94(5 Pt 1):647-61.
3. Lawrence R, Thevasagayam R. Controversies in the management of sudden sensorineural hearing loss: an evidence-based review. Clin Otolaryngol. 2015;40(3):176-82.
4. Kuhn M, Heman-Ackah SE, Shaikh JA, Roehm PC. Sudden sensorineural hearing loss: a review of diagnosis, treatment, and prognosis. Trends Amplif 2011;15:91-105
5. Chau JK, Lin JR, Atashband S, Irvine RA, Westerberg BD. Systematic review of the evidence for the etiology of adult sudden sensorineural hearing loss. Laryngoscope. 2010; 120:1011-21.
6. Schreiber BE, Agrup C, Haskard DO, Luxon LM. Sudden sensorineural hearing loss. Lancet. 2010;375(9721):1203-11
7. Conlin AE, Parnes LS. Treatment of sudden sensorineural hearing loss: I. A systematic review. Arch Otolaryngol Head Neck Surg. 2007;133:573–81.
8. Stachler RJ, Chandrasekhar SS, Archer SM, et al. Clinical practice guideline: sudden hearing loss. Otolaryngol Head Neck Surg. 2012;146(3 Suppl):S1-35.
9. Lee HY, Jung SY, Park MS, Yeo SG, Lee SY, Lee SK. Feasibility of three-dimensional fluid-attenuated inversion recovery magnetic resonance imaging as a prognostic factor in patients with sudden hearing loss. Eur Arch Otorhinolaryngol. 2012;269(8):1885-1891
10. Lee JI, Yoon RG, Lee JH, et al. Prognostic Value of Labyrinthine 3D-FLAIR Abnormalities in Idiopathic Sudden Sensorineural Hearing Loss. AJNR Am J Neuroradiol. 2016. [Epub ahead of print].
11. Yoshida T, Sugiura M, Naganawa S, et al. Three-dimensional fluid-attenuated inversion recovery magnetic resonance imaging find-ings and prognosis in sudden sensorineural hearing loss. Laryngoscope. 2008;118:1433–37.
12. 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): 1205-1209.
13. Kim TY, Park DW, Lee YJ, et al. Comparison of Inner Ear Contrast Enhancement among Patients with Unilateral Inner Ear Symptoms in MR Images Obtained 10 Minutes and 4 Hours after Gadolinium Injection. AJNR Am J Neuroradiol. 2015;36(12):2367-72.

14. Berrettini S, Seccia V, Fortunato S, et al. Analysis of the 3-dimensional fluid-attenuated inversion-recovery (3D-FLAIR) sequence in idiopathic sudden sensorineural hearing loss. JAMA Otolaryngol Head Neck Surg. 2013;139(5):456-64.

15. Zhu H, Ou Y, Fu J, Zhang Y, Xiong H, Xu Y. A comparison of inner ear imaging features at different time points of sudden sensorineural hearing loss with three-dimensional fluid-attenuated inversion recovery magnetic resonance imaging. Eur Arch Otorhinolaryngol. 2015;272(10):2659-65.

16. Witsell DL, Khoury T, Schulz KA, Stachler R, Tucci DL, Wojdyla D. Evaluation of Compliance for Treatment of Sudden Hearing Loss: A CHEER Network Study. Otolaryngol Head Neck Surg. 2016;155(1):48-55.

17. Coelho DH, Thacker LR, Hsu DW. Variability in the management of idiopathic sudden sensorineural hearing loss. Otolaryngol Head Neck Surg. 2011;145(5):813-7.

18. Portmann M, Dauman R, Duriez F, Portmann D, Dhillon R. Modern diagnostic strategy for acoustic neuromas. Arch Otorhinolaryngol. 1989;246(5):286-91.

19. Biavati MJ, Gross JD, Wilson WR, Dina TS. Magnetic resonance imaging evidence of a focal pontine ischemia in sudden hearing loss and seventh nerve paralysis. Am J Otol. 1994;15(2):250-3.

20. Franklin DJ, Coker NJ, Jenkins HA. Sudden sensorineural hearing loss as a presentation of multiple sclerosis. Arch Otolaryngol Head Neck Surg. 1989;115(1):41-5.

21. Hagiwara M, Roland Jr JT, Wu X, et al, Identification of endolymphatichydrops in Meniere's disease utilizing delayed postcontrast 3D FLAIR andfused 3D FLAIR and CISS color maps. Otol. Neurotol. 2014;35:e337–e342.

22. Ryan M, Weissman JL, Kaylie D. Is Gadolinium contrast enhancement necessary in screening MRI for asymmetric sensorineural hearing loss? Laryngoscope 2015;125(4):783-4.

23. Liao WH, Wu HM, Wu HY, et al. Revisiting the relationship of three-dimensional fluid attenuation inversion recovery imaging and hearing outcomes in adults with idiopathic unilateral sudden sensorineural hearing loss. Eur J Radiol. 2016;85(12):2188-2194.

24. Yagi A, Sato N, Takahashi A, et al. Added value of contrast-enhanced CISS imaging in relation to conventional MR images for the evaluation of intracavernous cranial nerve lesions. Neuroradiology 2010;52(12):1101-9.

25. Huynh TN, Johnson T, Poder L, Joe BN, Webb EM, Coakley FV. T1 pseudohyperintensity on fat-suppressed magnetic resonance imaging: a potential diagnostic pitfall. J Comput Assist Tomogr 2011;35(4):459-61.

26. Mark A, Fitzgerald D. Segmental enhancement of the cochlea on contrast-enhanced MR: correlation with the frequency of hearing loss and possible sign of perilymphatic fistula and autoimmune labyrinthitis. AJNR Am J

Neuroradiol. 1993;14:991–996 11.

27. Fitzgerald DC, Mark AS. Sudden hearing loss: frequency of abnormal findings on contrast-enhanced MR studies. AJNR Am J Neuroradiol. 1998;19(8):1433-6.

28. 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 Apr;20(1):31-5.

29. Ramos HV, Barros FA, Yamashita H, Penido Nde O, Souza AC, Yamaoka WY. Magnetic resonance imaging in sudden deafness. Braz J Otorhinolaryngol. 2005; 71(4):422-426.

30. Gao Z, Chi FL. The clinical value of three-dimensional fluid-attenuated inversion recovery magnetic resonance imaging in patients with idiopathic sudden sensorineural hearing loss: a meta-analysis. Otol Neurotol. 2014;35(10):1730-5.

31. Conlin AE, Parnes LS. Treatment of sudden sensorineural hearing loss: II. A Meta-analysis Arch Otolaryngol Head Neck Surg. 2007;133(6):582-6.

32. Passamonti SM, Di Berardino F, Bucciarelli P, et al. Risk factors for idiopathic sudden sensorineural hearing loss and their association with clinical outcome. Thromb Res. 2015; 135(3):508-12.

33. Capaccio P, Ottaviani F, Cuccarini V, et al. Genetic and acquired prothrombotic risk factors and sudden hearing loss. Laryngoscope 2007;117(3):547-51.

34. Quaranta N, De Ceglie V, D'Elia A. Endothelial Dysfunction in Idiopathic Sudden Sensorineural Hearing Loss: A Review. Audiology Research. 2016;6(1):151.

35. Moskowitz D, Lee KJ, Smith HW. Steroid use in idiopathic sudden sensorineural hearing loss. The Laryngoscope 1984;94:664-6.

36. 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;106:772-6.

37. Crane RA, Camilon M, Nguyen S, Meyer TA. Steroids for treatment of sudden sensorineural hearing loss: a meta-analysis of randomized controlled trials. Laryngoscope 2015;125(1):209-17.

38. Lavigne P, Lavigne F, Saliba I. Intratympanic corticosteroids injections: a systematic review of literature. Eur Arch Otorhinolaryngol. 2016;273(9):2271-8.

39. Fattori B, Berrettini S, Casani A, Nacci A, De Vito A, De Iaco G. Sudden hypoacusis treated with hyperbaric oxygen therapy: a controlled study. Ear Nose Throat J. 2001;80(9):655-60.

40. Narozny W, Sicko Z, Przewozny T, Stankiewicz C, Kot J, Kuczkowski J. Usefulness of high doses of glucocorticoids and hyperbaric oxygen therapy in sudden sensorineural hearing loss treatment. Otol Neurotol. 2004 Nov;25(6):916-23.

41. Awad Z, Huins C, Pothier DD. Antivirals for idiopathic sudden sensorineural hearing loss. Cochrane Database Syst Rev. 2012;(8):CD006987.

FIGURE LEGENDS

Figure 1: Vascular pattern in a 20-year-old woman with left SSHL. Pre-contrast T1-weighted (a) and pre-contrast 3D-FLAIR (b) sequences show a high signal in the middle and upper turns of the left cochlea without enhancement on post-contrast T1-weighted (c) and 3D-FLAIR images (d).

Figure 2: Inflammatory pattern in a 35-year-old man with right SSHL. The pre-contrast T1-weighted sequence (a) shows no signal abnormalities. The pre-contrast 3D-FLAIR sequence **(b)** shows a high signal in the right VII and VIII cranial nerves and in the middle and upper turns of the left cochlea. A post-contrast T1-weighted sequence (c) does not show significant enhancement, whereas post-contrast 3D-FLAIR sequence (d) show the cochlea and the VII/VIII cranial nerves as markedly enhanced on the right side.

Figure 3: Normal MRI findings in a 59-year-old man with right SSHL. The pre-contrast T1-weighted sequence (a) shows spontaneous hyperintense intralabyrinthine fluid in both inner ear structures (arrows), which is symmetrical and less evident on the same sequence after contrast injection (b). This hyperintensity is an artefact due to the altered dynamic range when the fat signal is subtracted, but also reflects an alteration in the visual appearance of signal intensity as the ambient contrast is changed (checker-shadow illusion) [24].

Supplementary Figure 4: **Clinical management algorithm for SSHL. Future research should investigate the role of temporal bone MRI in choosing a targeted treatment for patients with cochlear SSHL**. Abbreviations: ABR = Auditory Brainstem esponse; cVEMPs = Cervical Vestibular Evoked Myogenic Potentials. TEOAE = Transiently Evoked Otoacoustics Emissions.