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RECURRENCE OF NON-HYDROPIC SUDDEN SENSORINEURAL HEARING LOSS (SSNHL): A LITERATURE REVIEW

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1. Introduction

Sudden Sensorineural Hearing Loss (SSNHL) is typically defined as the acute onset (less than 3 days) of a perceptible hearing loss of more than 30dB over at least three contiguous frequencies on pure tone audiometry [1]. The exact incidence of SSNHL is uncertain, since many patients have a rapid and spontaneous resolution of symptoms and therefore don't reach medical attention. Estimate of incidence ranges from 5 to 20 per 100.000 individuals, and bilateral involvement is very rare [2]; it increases in the older patients (>65 yo)(77 per 100.000) the in younger population (<18 yo)(11 per 100.000) [3]. The true incidence of paediatric SSNHL is not established in literature; 40% of examined child with SSNHL, showed anatomic abnormalities [4].

The left ear is more affected only in female patients; the cause is unclear, but this asymmetry might indicate a greater vulnerability only in female, due to hormonal factors in the genesis of sudden deafness [5].

The pathogenesis of SSNHL it has been widely studied but is not yet entirely clear: it is multifactorial, and various causes have been proposed, including viral infection of the labyrinth

or cochlear nerve, autoimmune ear disease, acoustic tumors, perilymphatic fistula, intracochlear membrane trauma or rupture, drug toxicity, vascular disorders (including hemorrhaging, thrombosis, embolism, vasospasm and hypercoagulability in the microcirculation of the cochlea)[6], Meniere's disease [7](**Figure 1**); in addition, major depression and anxiety can be the cause of sudden deafness [8].

However, in most patients the cause is idiopathic.

It is commonly presented as a sudden unilateral deafness on awakening; aural fullness, tinnitus, vertigo and dysequilibrium are present to a variable degree in approximately 40% of patients [9].

The clinician should exclude a conductive hearing loss, perform an audiometry test as soon as possible (within 14 days of symptoms onset) and evaluate the patient for retrocochlear pathology by optioning an MRI or auditory brainstem response (ABR); CT scan and routine laboratory tests are not recommended in the approach.

Most recovery occurs within the first 2 weeks after onset; without treatment of any kind, a significant proportion (30% to 65%) of patients experience complete or partial recovery. (Figure 2).

Figure 1. Cause of Sudden Sensorineural Hearing Loss (from “Hearing loss in adults: differential diagnosis and treatment”, Michels TC et al., Am Fam Physician 2019 Jul 15;100(2):98-108)

Type	Hearing recovery
Complete recovery	The final hearing level was better than 25 dB regardless of the size of the gain
Partial recovery	The improvement exceeded 15 dB, and the final hearing level was between 25 and 45 dB
Slight recovery	The improvement exceeded 15 dB, but the final hearing level was poorer than 45 dB
No recovery	The improvement was less than 15 dB, or the final hearing level was poorer than 75 dB

Figure 2. Siegel’s criteria: degree of hearing recovery from sudden hearing loss

Causes of Sudden Sensorineural Hearing Loss	
Type of hearing loss	Cause
Idiopathic	Unknown
Infectious	EBV, group A streptococcus, HSV, HZV, HIV, Lyme disease, meningitis, syphilis
Otologic	Autoimmune condition, Meniere disease
Trauma	Barotrauma, ear trauma or head trauma
Vascular	Cerebrovascular disease
Neoplastic	Angioma, hyperviscosity, meningioma, neurofibromatosis 2, schwannoma
Other	Genetic cause, mitochondrial disorder, ototoxins, pregnancy

Children and adults >40 years old have a poorer prognosis [2,10].

Treatment of SSNHL should be based on its etiology. Most cases are idiopathic; the best options is initial therapy with corticosteroids, which can be combined with hyperbaric oxygen therapy within 2 weeks of onset of SSNHL.

Hyperbaric oxygen therapy (HBO) can be offered or combined with steroid therapy within 2 weeks of onset of SSHL, or combined with steroid therapy as salvage within 1 month of onset of symptoms. Some

authors [11] recommend the routine application of HBO with intravenous steroid for all patients with idiopathic sudden sensorineural hearing loss within 14 days from the onset of symptoms.

Antivirals, trombolitics and a vasodilators are not recommended.

When patients have incomplete recovery 2 to 6 weeks after onset of symptoms, intratympanic steroids could be an option as salvage therapy [1]. Meniere syndrome can occur in 5% of cases with SSNHL; generally, other symptoms are present (vertigo, tinnitus)[12,13]. Schaaf et al. [14] reported that recurrent low-frequency sensory hearing loss

can be one characteristic of Meniere's disease; in this work, only 3.7% of the patients developed the typical signs of Meniere's disease, and 25.9% of these suffered from recurrent hearing loss. They conclude that although almost every patient with Meniere's disease suffers from recurrent hearing loss, only a few patients with recurrent hearing loss develop Meniere's disease.

Acute low-tone sensorineural hearing loss (ALSHL) is a deterioration in audiometric low frequencies (250-500 Hz) with preservation of high frequencies; it was initially considered as a variant of SSNHL, until it was described as an independent disease entity. Moreover, there are several differences between ALSHL and SSNHL; 1) predominance of female (72.9%), 2) more dizziness (36%) and tinnitus (42.8%), 3) ear fullness (20.8%), 4) better prognosis of complete recovery (from 67.7% to 77%), 5) absence of predictive factors, 6) an important development to Meniere's disease 15% [15].

Additionally, the incidence of relapse is higher than in SSNHL (24%) [16,17].

Although there are many reports about the pathogenesis, treatment and prognostic criteria of SSNHL, few studies have examined relapsed neurosensorial hearing loss; moreover, most of the studies do not investigate the pathologies leading to relapses.

The aim of this paper is to evaluate the incidence and the risk factors of recurrent non-hydroptic SSNHL in literature.

2. Discussion

We define SSNHL relapse as an episode of SSNHL greater than or equal than the first episode of SSNHL; it consists in two types, ipsilateral and contralateral SSNHL; the ipsilateral type is defined as the first episode of idiopathic sudden deafness that had hearing improvement; after a period, a second episode of sudden deafness recurred in the same ear. In the contralateral type, two episodes of sudden deafness occurred in each ear, alternatively [18]. Kuo YL et al. compared the two types, and no significant differences existed in terms of 1) age of onset at the second episode, 2) gender, 3) laterality, 4) presence of vertigo, 5) spontaneous nystagmus, 6) abnormal ENG findings, 7) abnormal caloric results, 8) initial and final mean hearing. A significant difference was observed in abnormal VEMPs between the two types; all patients with ipsilateral type had normal VEMPs associated with improved hearing, whereas all patients with

contralateral type showed abnormal VEMPs. In conclusion, the demonstration of normal VEMPs in the lesion ear of the second episode in patients with recurrent SSNHL may indicate a good hearing outcome.

The main cause (90%) of SSNHL is idiopathic sudden sensorineural hearing loss (ISSNHL) in which the cause is not identifiable (undetermined); is commonly unilateral, and bilateral loss should lead to thinking about other causes. It is not commonly associated with other symptoms.

The incidence of ISSNHL relapse reported in the English Literature varies from 0.8% [19] to 8.3% [20].

The definite pathogenesis of ISSNHL is currently unclear.

Che-Ming Wu et al. [21] described one of the largest cohorts of patients with ISSNHL; a relapse of ISSNHL occurred in 2,281 patients (incidence 4.99%). The relapse was significantly higher (60%) in middle age patients (35-64 yo) than in younger patients (0-34 yo) (17%). In this study there is a higher prevalence of comorbid diabetes mellitus and hypercholesterolemia in the patients with a recurrence of ISSNHL than in those without, suggesting that the insufficient cochlear perfusion is an important risk factor for the recurrence of sudden deafness; otherwise, the individual comorbidities of coronary artery disease, hypertension, chronic renal disease, diabetes mellitus and hypercholesterolemia are not associated with a higher incidence of ISSNHL. There is no large difference in relapse between male and female (54.11% vs. 45.89%).

Identifiable causes are found for 7% to 45% of patients with SSNHL [22, 23, 24].

The etiology is very broad; Chau et al. [25], identified infections (13%), otologic problems (5%), traumatic (4%), vascular (3%), neoplastic (2%) as the most frequent cause of symptomatic sudden hearing loss. Capuano et al. [26] suggested that patent foramen ovale and right to left shunt are frequent in ISSNHL, particularly in young patients without comorbidities and with associated dizziness.

Infectious, traumatic, metabolic and neurologic disease are not linked to relapsed SSNHL (although increased levels of neutrophil to lymphocyte ratio (NLR) and platelet to lymphocyte ratio (PLR) have been shown in relapsed SSNHL [27]). About infections in recurrent SSNHL, some authors [28] speculate that they are caused by a reactivation of a latent HSV infection; in some patients, high values of

serum antibody titer for adenovirus 3 or human HSV-1 or VZV were detected.

Some vascular diseases can cause recurrent SSNHL, usually associated with other symptoms.

Lee and Cho [29] proposed some auditory disturbance (as recurrent sudden sensorineural hearing loss) as a warning sign of impending pontocerebellar infarction in the distribution of anterior inferior cerebellar artery (AICA) due to the ischemia of the inner ear or the vestibulocochlear nerve. In the opinion of Park JH et al. [30], in elderly patients (especially with particular risk factors) with recurrence of hearing loss and vertigo lasting several minutes, physicians may also consider the potential symptom of AICA infarction. Park H et al. [31] reported a single case of unilateral recurrent hearing loss due to medullary infarction. Bliss et al. [32] described an interesting report about recurrent contralateral hearing loss after two craniotomies for vestibular schwannoma; SSNHL on the the side opposite cerebellopontine angle surgery is a rare but well-documented occurrence. Most losses are recurrent but transient; the cause for the contralateral loss is still unclear; auto-immune mediated cochleolabyrinthitis [33], cardiovascular complication [34], allergy [35], labyrinthine rupture from elevated intratympanic pressure during general anesthetic [36] are just theories.

Some authors, as Walsted et al. [37] proposed the concept of a relative endolymphatic hydrops in the contralateral ear after vestibular schwannoma removed; they suggested that low CSF pressure can be transmitted to the cochlea through the cochlear aqueduct, creating a low perilymphatic pressure.

Some neoplasm may be related to recurrent SSNHL; for example, a vestibular schwannoma or another tumor in the internal auditory canal or at the cerebellopontine angle should be the cause [7].

MRI imaging should be performed to evaluate a vascular or retrocochlear pathology.

Electrocochleographic findings should be assessed as prognostic factors in SSNHL; in a study by Ohashi et al. [38], the patients were examined with transtympanic transpromontory ECochG within 5 days from the onset of symptoms; they concluded that an enhanced SP/AP ratio and a low initial AP threshold may be positive prognostic factors in recurrent SSNHL; furthermore the presence of initial vertigo seems to be an unfavourable factor in recurrent SSNHL.

Prognosis and recurrence rate can be influenced by many factors; metabolic syndrome [39], hyperlipidemia [40], diabetes mellitus [41,42,43,44,45] are negative prognostic factors for

recovery and recurrences; furthermore, hearing recovery is poorer after a second attack of SSNHL.

3. Conclusion

Although recurrent non-hydropic SSNHL is unfrequent, it represents a diagnostic and therapeutic challenge.

Differential diagnosis is essential to distinguish idiopathic forms from those symptomatic: the clinician should pay attention to vascular disease and retrocochlear neoplasm.

The literature established that recurrent SSNHL has a poorer prognosis than a single episode of sudden hearing loss.

Although some factors are described to affect recovery and recurrent state, further studies should be needed to explain better the related mechanisms and to evaluate the role of audiometric examinations.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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