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Research Article

Sudden Sensorineural Hearing Loss: An Updated Review

José Luis Treviño-González PhD MD*, Felix Maldonado MD, Germán A. Soto-Galindo MD, Jesús Eduardo Hernández de León MDa, Laura Lisset Reyes Suárez MDa

Deparment of Otolaryngology and Head and Neck Surgery, Universidad Autónoma de Nuevo Leon, México

***Corresponding Author:** Jose Luis Trevino-Gonzalez, Department of Otolaryngology and Head and Neck Surgery, Universidad Autónoma de Nuevo León, Medicine School and University Hospital, Vista Hermosa, Monterrey, Nuevo León, México.

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Abstract

Sudden sensorineural deafness is defined as a hearing loss greater than 30 dB in 3 contiguous frequencies of pure tones that occur within 72 hours. It is a rare but disabling entity, affecting approximately 5 - 27 people per 100,000 globally per year.

The etiology is identified only in a few cases, being a viral infection the most associated cause. The diagnosis is based primarily on the medical history, physical examination, and pure tone audiometry. The mainstay of treatment is corticosteroids, either systemic or intratympanic. This treatment can be accompanied by hyperbaric oxygen therapy to improve the response. For the follow-up, serial audiometric studies are suggested at the end of the treatment and six months, in search of residual hearing loss.

We examined the most recent studies on epidemiology, clinical manifestations, diagnosis, and treatment of sudden sensorineural hearing loss to provide a comprehensive review on this topic to improve the understanding of this entity among clinicians.

Keywords: Sudden Sensorineural Hearing Loss; Idiopathic Sudden Sensorineural Hearing Loss; Treatment; Epidemiology; Therapy

Abbreviation

ISSNHL: Idiopathic Sudden Sensorineural Hearing Loss

Introduction

Idiopathic sudden sensorineural hearing loss (ISSNHL) is generally defined as a sensorineural hearing loss of greater than 30 dB across 3 contiguous pure-tone frequencies occurring within 72 hours. ISSNHL is considered an otolaryngologic emergency. Its real incidence is unknown since 30 - 65% of cases recover spontaneously, therefore, some patients do not seek medical attention. SSNHL has a prevalence of 10 per 100000 persons in Korea and 5 to 20 per 100000 persons in the United States [1,2]. The incidence of ISSNHL is approximately 5 to 20 cases per 100000 persons per year. The highest incidence occurs within patients aged between 50 - 60 years. At present, pathogenesis, clinical signs, treatment, and prognostic components of ISSNHL are not clear, with spontaneous recovery rates ranging from 32% to 70% [3,4].

The main cause of ISSNHL is not well understood [5]. Due to the relatively low incidence of ISSNHL, it is not possible to determine with certainty either its etiology or adequate therapy. Viral infection, vascular occlusion, irregular cellular stress responses within the cochlea, and immune-mediated mechanisms are among the most frequently suspected etiologies of ISSNHL [6]. Concerning the

viral cause, the herpes virus family (Herpesviridae) is probably the most associated family of viruses in ISSNHL pathogenesis [7].

Materials and Methods

Pubmed and Google Scholar databases were reviewed using the following keywords: Sudden sensorineural hearing loss/treatment/epidemiology/therapy/complications from 2015 up to 2020.

Initially, 772 articles were identified, 679 articles were found in the Google Scholar database, 93 in the PubMed database; of which we selected 75. Articles selected for review were published in English or Spanish and at last, 33 articles were chosen for the writing of this review.

Figure 1: Review flow chart.

Results and Discussion

Etiology and physiopathology

About 90% of cases of SSNHL are idiopathic. Its etiology is multifactorial and viral infections, autoimmune disorders, and vascular abnormalities such as leukemia, sickle-cell anemia and iron-deficiency anemia might be involved. Anemia can lead to insufficient oxygen supply to the organs, therefore organs that demand more oxygen are more susceptible to an anemic insult predisposing to infection and inflammation. Due to the high metabolic demands of the cochlear hair cells involved in mechanoelectrical transduction to acoustic stimuli, the cochlea is one of the most vulnerable organs. In medical literature, it is well described that oxidative stress and inflammation are the main pathophysiology mechanisms for noise-induced, ototoxic drug-induced, and age-related hearing loss [8].

Regarding the autoimmunity hypothesis, it seems that mechanisms not well identified may play a role, given that the blood-labyrinth barrier has its autoimmune privileges, and there are only a few macrophages in this area. Despite this, the immune-mediated mechanism of ISSHNL has been verified using immunosuppressants as an effective therapy [9].

A possible association between viral infection and ISSNHL has been proposed by many studies. Herpes simplex virus, human immunodeficiency virus, hepatitis virus, measles virus, rubella virus, mumps virus, Lassa virus, and enterovirus are the most related and described in the literature. There are three proposed mechanisms by which viruses might cause sudden hearing loss. The first one is through the invasion of the cochlear nerve, the fluid spaces, or the soft tissue of the cochlea. The second is through the reactivation of a virus lying in the latent phase within tissues of the inner ear. The last one is by triggering an antibody response that cross-react with an ear antigen, causing activation of cellular stress pathways in the cochlea [6].

The underlying etiology is found in only 10 - 15% of the patients at the initial presentation. After a follow-up, the underlying etiology is found in up to 30% of the cases. Although there are plenty of scientific studies about this subject, there are only a few of them that approach etiology and physiopathology, leaving room for further research about this [10].

Clinical presentation

ISSNHL is more common between 50 to 60 years old population with no sex preference. In most cases it is unilateral, bilateral involvement is reported in less than 5% of the cases. Patients usually also a sudden hearing loss upon awakening. Tinnitus accompanies hearing loss in 80% of the cases and vertigo in 30%, being associated with a peripheral vestibular dysfunction. Ear fullness can be found in up to 80% of the patients affected by ISSNHL. A normal otoscopic examination in both ears is expected in ISSNHL, accom-

panied by a sensorineural hearing loss pattern of the affected ear given by Weber and Rinne tuning fork tests [16].

Diagnosis

The medical record should focus on past personal medical history including infections, otologic diseases, trauma and surgeries, medications, history of neoplasia, and vascular diseases.

In view of the clinical suspicion of ISSNHL, an otoscopy should be performed which must show absence of pathology in the external and middle ear in both ears. Tuning fork tests are compatible with a sensorineural hearing loss of the affected ear with, Rinne´s test (256 or 512 Hz tuning fork) positive in the affected ear and a Weber´s test (256 or 512 Hz tuning fork) lateralized to the healthy ear. Nevertheless, a false negative Rinne can be found in extreme cases (the patient does not hear the tuning fork at all) [34].

Audiometry should confirm a sensorineural hearing loss pattern, showing a decrease in auditory thresholds of 30Db in three consecutive pure tone frequencies from baseline hearing (Figure 2).

Differential diagnosis

Numerous different factors may be responsible for acute hearing loss. Differential diagnosis is based on the patient's history, ear microscopy, tuning test (Weber and Rinne), pure tone audiometry, and neuro-otological imaging [11]. MRI with gadolinium should be requested to rule out any retrocochlear deficiency, ponto-cerebellar angle tumors or demyelinating diseases. A fine-cut computed tomography with contrast can be considered as an alternative [12,13].

In the differential diagnosis, major depression should be considered, as it may lead to apathy and social disengagement. Patients with dementia should also be evaluated for hearing loss as hearing impairment is commonly present. Cognitive screening should be performed to detect these entities.

In table 1, we described some of the differential diagnosis and common causes of sudden hearing loss [14].

Treatment

Considering the evaluation of the ratio of risk versus benefit, oral steroids are generally prescribed as a first-line treatment [15].

Figure 2: Audiometries showing a sensorineural hearing loss pattern.

Systemic steroids

The first-line treatment is oral systemic steroids. However, it has not been well established the proper length of oral steroid therapy. In medical literature, it ranges between 5 to 24 days. Oral prednisolone is often prescribed at a dose of 60 mg per day (i.e. approximately 1 mg/kg) [15-17].

Type of hearing loss	Cause
Idiopathic	Unknown
Infectious	Epstein-Barr virus, group A streptococcus, herpes zoster virus, HIV; Lyme disease, syphilis, meningitis, herpes simplex virus
Otologic	Meniere disease, an autoimmune condition
Trauma	Barotrauma, ear trauma, head trauma
Vascular	Cerebrovascular disease
Neoplastic	Neurofibromatosis 2, schwannoma, angioma, meningioma
Other	Genetic causes, mitochondrial disorder, ototoxins, pregnancy

Table 1: Common causes of sudden sensorineural hearing loss.

Prednisolone for long-term therapy may not be beneficial for patients because of the excessive dose intake. According to some studies, after 2 months of treatment, hearing outcomes are not expected to continue improving [17].

Trans-tympanic steroids

High concentrations of steroids in the inner ear can be obtained using intratympanic steroids without the risk of adverse effects of systemic steroids [18]. In a supine position, the patient's head is to be turned 30 degrees away from the physician, steroids are administered in the inner ear, under local anesthesia. Under direct visualization through an operating microscope and using a syringe connected to a 22- (3.5 IN; 0.7 90 mm) or 25-gauge spinal needle, a 0.4 - 0.6 ml of methylprednisolone can be injected into the tympanic cavity through the posterior inferior quadrant of the tympanic membrane. To avoid vertigo, heating the solution to body temperature is recommended before the injection. After the application, the position of the patient is kept for 30 minutes. The patient is evaluated after one week of the first dose. Another dose might be needed to be administered if no improvement or partial improvement is observed [19].

Antivirals

There was no effect on the remission rates found in patients on treatment with combined therapy [7]. Therefore, it is not recommended to prescribe antivirals to patients with ISSNHL [20].

Heparin

Higher hearing recovery rates in selected patients with a PTA at 90 to 100 dB have been observed as a result of the use of treatment with adjuvant heparin therapy in combination with systemic steroid therapy when compared to combined systemic and local corticosteroid therapy alone [21].

Treatment in diabetic patients

In patients with a history of Diabetes Mellitus, steroids are known to exacerbate hyperglycemia or even develop it in those without any history of metabolic disorders. Therefore, intratympanic corticosteroids are prescribed as the main treatment for sudden hearing loss in patients with diabetes [22,23].

Metformin

In patients with Diabetes Mellitus, metformin has been reported to play a protective role against cerebral or cardiovascular disease and to decrease the risk of stroke. In the study conducted by Dr. Hsin-Chien Chen., *et al*. the relationship between metformin use and the lower incidence of SNHL among DM patients was found [24].

Hyperbaric oxygen

The treatment with hyperbaric oxygen is based on its vasodilator effect on Corti's organ and other internal ear structures thus acting against the process of vascular compromise and oxidative stress, which are hypothesized to be major factors in the pathophysiology ISSNHL. This can be appropriately added to the standard treatment with corticosteroids, particularly for those patients with moderate to profound hearing loss [25,26].

Follow-up

Follow-up audiometric tests for patients with ISSNHL should be received at the end of treatment and within 6 months of completion of treatment. This is currently recommended by the American Academy of Otolaryngology and Head and Neck Surgery and the aim of this recommendation is to recognize patients who may benefit from other options for hearing loss and tinnitus, as well as to determine potential etiologies that may occur much later after the initial episode of hearing loss [27].

Rehabilitation

The guideline of the American Academy of Otolaryngology and Head and Neck Surgery proposes physicians to counsel patients

with ISSNHL who have residual hearing loss or tinnitus about the possible benefit of audiology therapy and other supportive measures. Recently, cochlear implantation has emerged as an option for patients with SSD, allowing for restoration of auditory input to the deaf ear [27,28].

Prognosis

Among the most important prognostic factors, tinnitus has been observed to be a good prognosis factor, whereas vertigo has one of poor prognosis. The only preventable prognostic poor factor is the late start of treatment [29]. During follow-up, an ascending pattern in the audiogram has been associated with a higher recovery rate [30]. Siegel's criteria are currently used for the assessment of recovery. These criteria are described in table 2 [31].

Type	Hearing recovery
1. Complete recovery	Final hearing better than 25 dB
2. Partial recovery	More than 15dB recovery, final hearing 25 - 45 dB
3. Slight improvement	More than 15 dB recovery, final hearing poorer than 45dB
4. No improvement	Less than 15 dB recovery, final hearing poorer than 75 dB

Table 2: Siegel's criteria.

Complications

Affective disorders

Recently, multiple studies have shown that ISSNHL is substantially correlated with the risk of developing affective disorders. The possibility of opposite ear affection and the risk of recurrence of hearing loss has been reported as a cause of anxiety. Depressive symptoms may arise given the long duration to hearing recovery and the social isolation that patients report after the hearing loss episode. Patients with history of ISSNHL typically have trouble hearing speech over background noise and even identifying sound sources [32,33].

The scores for anxiety and depression scales were higher in women than in men in the DR Fatih Arslan., *et al*. study, but there was no statistically significant difference. This result demonstrates that doctors should consider the high risk of developing affective disorders in patients with ISSNHL and take specific steps to reduce their risk of affective disorders [32,33].

Conclusion

SSHL is a rare entity, considered an emergency in the otolaryngology field, of which etiology and physiopathology are poorly understood. Diagnosis is based on three fundamental pillars: medical history, physical examination, and pure-tone audiometry. As for treatment, it is mainly based on the use of corticosteroids, whether systemic or intra-tympanic. The use of any other adjuvant drug as part of the first line scheme is not recommended. Follow-up with pure-tone audiometry at the end of treatment and 6 months for residual hearing loss is recommended.

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Conflict of Interest

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Informed Consent

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