



Sudden Sensorineural Hearing Loss: An Updated Review

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Abstract

Sudden Sensorineural Hearing Loss is an unexplained and suddenly appearing hearing loss that occurs over less than a 72 h period and most cases are unilateral and idiopathic. It often presents in the prime of mid-life between 43 years to 53 years of age, with equal sex distribution. This diminish of hearing can be severe in some patients while mild in others and it may be accompanied by tinnitus and vertigo, among other symptoms. It has a multifactorial etiology and, in most cases, uncertain, this has led to many different therapeutic approaches, including corticosteroids, antiviral agents, hyperbaric oxygen, and others. Glucocorticoids are considered first line of therapy, more commonly administrated orally or other times locally by intratympanic injections. Intratympanic injections are commonly used when hearing does not recover after systemic treatment. Combination of systemic and local glucocorticoid therapy may be considered in patients with profound loss of hearing. However, neither of these approaches has been established as official or superior due to a lack of strong evidence for the efficacy of any of these treatments. More clinical trials are needed to establish a definitive management. We reviewed the latest and most recent studies that compare different treatment methods and provide an update on Sudden Sensorineural Hearing Loss' epidemiology, clinical manifestations, diagnosis and treatment.

Keywords: Sudden sensorineural hearing loss; Intratympanic steroids; dBHL

Introduction

Sudden Sensorineural Hearing Loss (SSNHL) is a suddenly appearing, generally unilateral hearing loss of at least 30 dB in three sequential frequencies in the standard pure tone audiogram, over 72 h or less [1]. "Probable" SSNHL is one in which two or three frequencies are reported, with losses of 10 dBHL to 20 dBHL, occurring in less than 12 h [2].

Sudden sensorineural hearing loss is considered one of the few otology emergencies, and although some patients have a complete or partial spontaneous recovery, 32% to 65% of patients do not recover [3].

The impact of an Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL) on a patient can be devastating; this event often presents in the prime of mid-life between 43 years to 53 years of age, with equal sex distribution [4]. The loss of function in an organ that's essential for communication can be a terrifying experience and significant stressor. The study by Kim reveals that an often-overlooked sequel is the onset of affective disorders such as depression and anxiety [5].

Sudden sensorineural hearing loss is not a simple disease entity but rather is likely to be a clinical symptom of multiple pathological conditions [6]. The etiology of SSNHL is thought to be multifactorial, mostly idiopathic (ISSNHL). Membrane damage of the labyrinth, viral infection; such as CMV, rubella virus and mumps virus; ototoxicity, genetic mutations, chronic inflammation, cellular stress theories, immunological disease, impairment of the vascular microcirculation, neoplastic, and nonorganic origin have been suggested as possible causes [7,8].

Epidemiology

According to Ahmadzai et al. [3] the incidence in the US is approximately 27 per 100,000 persons annually. It is frequently present in patients between 25 years and 60 years old, but there is a peak on prevalence in patients between 46 years and 49 years old. Prevalence in patients under 18 years of age it's of 6.6%, under 14 years it's of 3.5%, and under 9 years its of 1.2% [9]. SSNHL is more frequent between 43 years to 53 years of age but it can occur at any age [10].

There are approximately 4000 new cases per year in the US [11]. National surveys in the UK have estimated the incidence of sudden sensorineural hearing loss between 5 and 30 cases per 100 000

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per year (Shin & Rauch, 2008). Wu et al. [7] describes an incidence of 10.21 per 100,000 in Taiwan. As a major incidence in autumn and as age increases [2]. Nakashima et al. [12] described an annual incidence of 60.9 per 100,000 in Japan. Klemm et al. [13] describes a prevalence of 160 per 100,000 cases per year in Dresden, Germany.

Clinical Manifestations

Most patients with SSNHL describe rapid hearing loss when awakening. Some patients don't notice the loss of hearing and refer a sensation of full ear. Most SSNHL cases are unilateral but up to 3 percent may be bilateral. About 90 percent of patients with unilateral SSNHL refer tinnitus. A 20% to 60% of patients refer vertigo. During physical examination signs of obstruction shouldn't be present. Patients that refer posterior neck pain should be evaluated for vertebral artery dissection [10].

Diagnosis

Diagnosis is based on history, physical examination, tuning fork tests, and audiometry. Patients should be evaluated to distinguish sensorineural hearing loss from conductive hearing loss [11]. Patients that refer acute hearing loss with no specific condition identifiable should be considered to have sudden sensorineural hearing loss. When SNHL is suspected, it becomes a priority to initiate treatment as soon as possible.

In order to obtain a correct diagnosis, a thorough case history should be obtained. History of prior hearing loss, ear drainage, fever, trauma, headache and ophthalmological symptoms should be asked.

Audiometric evaluations are essential at first contact and during follow up in patients with a SSNHL. Weber and Rinne tuning fork tests can help distinguish between Conductive Hearing Loss (CHL) and Sensorineural Hearing Loss (SNHL). During the Weber test, SSNHL can be identified when it lateralizes to the affected ear. However, if lateralization it's not present, SSNHL it's not excluded and further evaluation it's needed.

Idiopathic sudden SNHL can be confirmed when hearing loss of at least 30 dB at three consecutive frequencies on audiometry is present, without an underlying condition identified by history and physical examination.

An appropriate physical examination includes an otoscopic examination for inspection of the ear canals and tympanic membranes to exclude foreign bodies, otitis media, perforated tympanic membrane, otitis externa or cholesteatoma. A neurologic examination should be performed to exclude a stroke in the AICA as a cause of Sudden Hearing Loss (SHL) [10]. Most of the otoscopic evaluation of patients with SNHL will be normal while patients with CHL will present alterations. If present, impacted cerumen should be removed before evaluation. Audiometry, tympanometry, pneumatic otoscopy, and Weber and Rinne tests are also helpful for arriving to a proper diagnosis [11].

According to Plontke, some features associated with differential diagnosis are: sudden bilateral hearing loss, fluctuating hearing loss, ophthalmological symptoms, isolated hearing loss in the lower frequencies, focal neurological symptoms, abnormal findings in the SNC imaging, and recent head trauma [6]. According to the AAO-HNS, a gadolinium-enhanced brain MRI, auditory brainstem response, or follow-up audiometry should be done in patients with ISSNHL to assess for retrocochlear pathology, being MRI the

most sensitive tool for diagnosis. Plontke described a prevalence of vestibular schwannoma of 4 percent in a group of patients with SHL in whom an MRI was performed. Screening for vestibular schwannoma allows earlier tumor identification and higher probabilities to preserve hearing [6].

Some causes that may be associated with sudden bilateral sensorineural hearing loss are metabolic, vascular, autoimmune inner ear disease, infectious (such as meningitis, syphilis, HIV, Lyme, Ramsay-Hunt syndrome), neoplastic, toxic (such as lead poisoning), trauma. A history of a fluctuating hearing loss presenting with SSNHL should suggest other diagnosis such as Meniere disease, autoimmune disease, Cogan syndrome, and hyperviscosity syndromes. SHL accompanying focal weakness, dysarthria, hemiataxia, encephalopathy, severe headaches, diplopia or another focal neurological symptom is an indicator of a SNC etiology [11].

The American Academy of Otolaryngology Head and Neck Surgery (AAO-HNS) doesn't recommend a CT scan of head and brain during initial evaluation of a patient in whom SSNHL is suspected. It doesn't provide sufficient resolution for detecting brainstem infarctions or tumors of the cerebellopontine angle and should only be performed in patients who have a contraindication for MRI [10].

The AAO-HNS doesn't recommend routine laboratory tests in patients with SSNHL. Laboratory studies should be ordered on findings from physical examination and history. For example, presence of Sexually Transmitted Diseases (STDs), history of exposure in an endemic area of Lyme's disease [10].

Treatment

There is controversy and a wide variety of treatment options in patients with ISSNHL due to a considerable rate of 45 to 65 percent of spontaneous recovery and a lack of strong evidence of a clearly superior therapy. The use of systemic corticosteroids, intratympanic corticosteroids, antiviral drugs, hyperbaric oxygen among others are well described in scientific literature, we reviewed articles where these methods were used and described.

Intratympanic (IT) glucocorticoids have been used for patients resistant to initial treatment in which hearing does not improve after a trial of oral glucocorticoids. IT glucocorticoids can be used as initial therapy in patients with a contraindication or intolerance to high dose systemic glucocorticoids. Combination therapy with systemic and local steroids is reserved for patients that have a profound hearing loss [10]. Hasan Demirhan et al. [14], compared the use of intratympanic methylprednisolone vs. dexamethasone using two groups, both groups received standard treatment for five days which consisted of 600 mg of pentoxifylin, enoxaparin Na 60 mg/0.6 mL, 500 mL of dextran 40, vitamin B complex supplementation and 30 mg of lansoprazole, and both groups received oral methylprednisolone at a dose of 1 mg/kg/day reducing the dose by 10mg every 3 days until reaching 10 mg or lower, group B which consisted of 60 patients received also the intratympanic steroid dividing them into two clinics: clinic A was injected 0.5 mL to 0.7 mL of methylprednisolone (66.6 mg/mL), and clinic B, 0.5 mL to 0.7 mL of dexamethasone (4 mg/mL). Both clinics had 5 injections, one every 72 h.

Patients that showed a moderate hearing loss had a higher rate of success with the dexamethasone approach, but this result showed no statistically significant difference. On the other hand, patients with a severe hearing loss also showed a greater success rate with this

therapy, with a statistically significant difference compared to the methyl prednisolone and the systemic only approach [14].

In Tokyo Japan, Hideaki Suzuki under the supervision of University of Occupational and Environmental Health studied the difference in effectiveness when applying 2 intratympanic injections vs. 4 intratympanic injections of dexamethasone 4 mg/mL in the treatment of SSNHL. They review the results in 191 patients and found no difference between both therapeutic approaches [15].

A study made on the University of Hawaii Institutional gathered 53 patients with SNHL dividing their therapeutic approach in 3 groups: oral corticosteroids, a combination of intratympanic and oral corticosteroids, and the final group only received intratympanic corticosteroids. They didn't find a significant difference between any of the groups, but significance was found between patients who had an earlier therapy compared to those who had a later treatment. Showing the importance of early onset of the corticosteroid treatment [16].

L. Sutton in Germany and Austria surveyed 908 specialists, who had 21 questions for otorhinolaryngologists on the frequency of intratympanic steroid use 49.1%. Of this 49.1% the 73.7% did not use them in the initial approach, 20.6% used them in conjunction with oral steroids and only the 5.8% used the intratympanic steroid as monotherapy; the most used intratympanic steroid was dexamethasone 4 mg/mL, with 61%, the rest is divided into different types of steroids without demonstrating the best one [17].

Jan Peter Thomas et al. [18] studied the use of tympanotomy and sealing of the labyrinthine window in SHL as a salvage therapy after ineffective systemic corticosteroid application in a study that included 136 patients which failed to show improvement after the use of systemic corticosteroids. After surgery 25 subjects showed a complete recovery of their audition, these was more remarkable in patients without comorbidities (systemic hypertension, coronary heart disease, vertigo and nystagmus) and under the age of 60; 105 patients experienced a >10 db improvement on the pure tone audiometries, and only 6 patients showed no improvement at all [18].

Awad Z describes in his review four randomized trials. Two trials compared intravenous acyclovir to a prednisolone, the third trial compared valacyclovir to prednisolone and the last one compared the addition of intravenous acyclovir to hydrocortisone with hydrocortisone alone. The total of participants were 257 participants, they reported no statistically significant advantage in the use of antivirals [19].

Patients with incomplete hearing recovery should be counseled about the possible benefits of amplification, hearing assistive technology, and other supportive measures [11].

Discussion

Sudden Sensorineural Hearing loss is a disease known to have a multifactorial etiology causing a variety of treatments to be used in its approach. Glucocorticoids are considered the first line of therapy, more commonly using only oral glucocorticoids, other times accompanied by intratympanic injections. Many studies have been made comparing these two approaches and comparing them to other therapeutic methods, not finding a definitive therapy yet, but many different pathways and forms of treatment that prove to be effective.

We can conclude after reviewing different results of a variety of

articles that the main factor in success with any form of glucocorticoid treatment is the earliness of its application, the sooner the therapy is set the better results and higher probability of improvement or even total recovery of the patient's hearing. In addition to the rapidness of the treatment application, another important factor to consider is that in most research papers found about this matter, patients with comorbidities such as diabetes mellitus, systemic hypertension, history of cardiovascular disease, among others, showed a considerably lower rate of improvement with most of the therapeutic approaches.

Sudden Sensorineural Hearing loss still leaves an open field for new research to be made on the most effective form of treatment; however, what appears to be certain is that the use of an early application of corticosteroid therapy is to this day the best form of treatment.

Objectives

Review the most recent studies on the subject to provide an update on its definition, epidemiology, clinical manifestations, diagnosis and treatment.

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