Chapter 56
Sudden Hearing Loss and Tinnitus

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Keywords Tinnitus • Sensorineural hearing loss • Hyperacusis • Sudden deafness • Transtympanic • Steroids

Abbreviations

ABR	Auditory brainstem response
AIEDA	Autoimmune inner ear disease
FTA	Fluorescent treponemal antibody absorbed (FTA-ABS) test for syphilis
MRI	Magnetic resonance imaging
RCT	Randomized clinical trials
SNHL	Sensory neural hearing loss
SSNHL	Sudden sensory neural hearing loss
TTS	Transtympanic steroids

Introduction

Hearing loss can occur suddenly when the ear canal becomes occluded or the middle ear becomes damaged from trauma. However, the term sudden hearing loss is mainly used for suddenly occurring sensory neural hearing loss. Sudden sensory neural hearing loss (SSNHL) was first described by De Klein in 1944 [1]. SSNHL is a dramatic condition for the patient that twenty-first century medicine still has no explanation of; there is no known cure. The mechanisms, the etiology, and the treatment remain hypothetical. The SSNHL definition is also controversial among authors. The most detailed criteria have been proposed by Stokroos [2], who described SSNHL as an acute deafness with abrupt onset, generally within 3 days, of more than 30-dB hearing loss at three consecutive frequencies. Different authors have used different definitions of SSNHL [3].

Incidence of SSNHL

SSNHL occurs suddenly, over less than 3 days; it normally affects only one ear. The incidence of SSNHL has been reported to be 5–20 per 100,000 inhabitants per year in the United States [4] and 8–14.6 in Holland [5]. A recent epidemiological study conducted during 2004 in Saxony, Germany, with a population of almost half a million, showed an incidence of 160 per 100,000 inhabitants [6].

Some incidences of SSNHL have been reported in childhood. The prevalence is higher in young and healthy individuals. Many individuals with SSNHL recover spontaneously. There are many causes of hearing loss similar to etiologies that could be regarded as other forms of sensorineural hearing loss (SNHL) [4], and they have to be ruled out before the diagnosis of SSNHL is made. Endolymphatic hydrops, according to the Fetterman series, is the second most common cause of acute hearing loss (5.5%) after idiopathic sudden hearing loss. Ménière's disease was the next most frequent diagnosis (1.9%), followed by vestibular schwannoma (1.7%), perilymphatic fistula (0.7%), and autoimmune inner ear disease (0.6%). Other authors find that vestibular schwannoma accounts for 4% of acute unilateral hearing loss [7].

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Causes of SSNHL

Up until now, the etiological of most SSNHL is unknown. In only 10% of the SSNHL is it possible to find a plausible cause [4]. Many causes have been suggested, such as disturbances in the cochlear blood flow, inflammatory processes secondary to viral infections, and autoimmune reactions.

Decrease of the Inner Ear Blood Flow

Blood flow in the inner ear may be reduced because of hemorrhage or arterial occlusion, which may occur from thrombosis or vascular spasm. Pathological studies describe processes of fibrosis and cochlear ossification in individuals who have had sudden deafness [8]. The vascular causes may be suspected in individuals with a history of previous thrombi-embolisms, arterosclerosis, heart surgery, or thrombocytopenia due to aplastic anemia or leukemia.

Spontaneous recovery makes impairment of the cochlea's blood supply for longer than 1 h an unlikely cause [9].

Rupture of the Cochlear Membranes

The rupture of the cochlear membranes causes contact between the perilympha and the endolympha, altering the electrolytic balance and resulting in damage to hair cells.

A perilymphatic fistula to the middle ear is known to cause sudden hearing loss. Intense physical exercise, Valsalva maneuvers, or barotrauma can cause rupture of the oval or round window membrane. It has been estimated that perilymph fistulae may explain almost thirty percent of the SSNHL [10].

Autoimmune Inner Ear Disease

Autoimmune inner ear disease (AIED) is characterized by rapidly progressive bilateral hearing loss, usually symmetrical and fluctuant, although some individuals experience sudden hearing loss in only one ear.

Viral Theory

The viral theory is the most referred in the literature. Viral infections like mumps, rubella, herpes, or spurma retrovirus have been related to sudden hearing loss, although there is no clear evidence to confirm this theory [11–13].

Clinical Course

Sudden hearing loss starts with a rapidly progressive hearing impairment, either suddenly or within a few hours, often when the person wakes up. Sixty-three percent of people with SSNHL have ear pain initially and 41% experience aural pressure during a few days [11]. Tinnitus appears in 91% of individuals with SSNHL [4]. Tinnitus has been reported to develop days before the SSNHL occurred or it can occur simultaneously or days after to the SSNHL. Some individuals describe facial paresthesia.

Vertigo has been reported to occur in 43% of individuals with SSNHL [11]. Some patients refer a rotatory motion with vegetative manifestations during a few days. Often unsteadiness and involuntary movement of the body toward the affected side occur for weeks. The presence of spontaneous nystagmus in SSNHL has been reported to occur in half of individuals with SSNHL. The recurrence rate of hearing loss in long-term follow-up has been reported to be significantly higher (51.2%) in the group who had spontaneous nystagmus than in the individuals without nystagmus (27.9%) [14].

Diagnosis

A diagnosis of individuals with rapidly occurring hearing loss requires complete auditory examination: tone and speech audiometry, tympanometry, and stapedial reflex test. Auditory brainstem responses or MRI to rule out retrocochlear diseases and laboratory tests such as the antinuclear antibodies, erythrocyte sedimentation rate, and tests for rheumatoid factor have been proposed for diagnosis of SSNHL and to detect treatment responders [15]. However, none of the

laboratory parameters have been proven to have a high sensitivity or specificity. A test to rule out syphilis (FTA) is also recommended [16].

The assessment of the vestibular system may be useful to detect possible vestibular complications for prognosis. Vestibular-evoked myogenic potentials (VEMP) have demonstrated saccular damage in patients with SSNHL without vertigo, suggesting a saccular deterioration in those patients with profound high-frequency hearing loss [17].

Prognosis

A spontaneous recovery has been described in 45–65% of the cases [11]. Some individuals with SSNHL have a complete recovery while most have partial improvement of hearing. Specific factors that affect the prognosis of SSNHL are the severity of the hearing loss; a greater impairment on high frequencies or the presence of vestibular symptoms significantly reduces the prognosis [11, 18]. However, Fetterman [4] did not find that the audiometric profile made any differences regarding prognosis, but "U"-shaped audiograms predict higher fluctuations and recurrences of the episodes of SSNHL. Age or speech recognition threshold did not influence the course of the disease.

Treatment

The high rate of spontaneous recovery and the difference in definition of SSNHL makes it difficult to compare the results presented in published studies [19].

Blood Flow Increase

Vasodilatation: *histamine, verapamil, papaverine, novocaine, nicotinic acid, naftidrofuryl, Egb 761*.

Studies that have followed a valid design do not show significant differences between treated individuals and control groups [20–22].

Reduction in the blood viscosity: Dextran, papaverine, pentoxifiline.

A recent multicenter and randomized study evaluated the benefits of rheopheresis, a method to reduce the plasma viscosity and improve microcirculation for treatment of SSNHL [23]. The rheopheresis group (two sessions within 3 days) was compared to a group that received steroid treatment (methyl-prednisolone 250 mg per day, 3 days and tapered oral dosing) and to intravenous hemodilution (500 ml 6% hydroxyethil starch plus 600 mg pentoxifyline per day during 10 days). There was not a placebo control group in this study. None of the tested treatments were superior regarding providing overall good recovery of hearing.

Defibrinogenase Therapy: Baxtrobin

Administration of baxtrobin, a trombine-like enzyme that reduces the levels of fibrinogen and the blood viscosity [24] did not present better results than expected with placebo.

Anti-inflammatory Treatment: Corticosteroids

Systemic Steroids

Corticosteroids are the most effective treatment for SSNHL. A placebo-controlled study demonstrated the efficacy of dexamethasone or methyl-prednisolone. Seventy-eight percent of the patients with moderate and severe hearing loss who received such treatment had partial or total recovery of hearing compared with placebo [25].

In a descriptive study by Moon [26], SSNHL participants who showed any improvement after early steroid therapy were analyzed to evaluate the beginning time and the plateau time of hearing improvement. It was shown that 93.1% had an onset of improvement within 14 days of beginning the treatment. Complete recovery or completed improvement was achieved in 80.4% of the participants within 1 month and in 92.2% within 2 months after treatment [26].

After 1 month, the possibility of improvement decreases [2], but Stokroos did not find differences in starting a treatment within the first 24 h and during the

first 10 days [2]. Better recovery was found in participants who had the most hearing loss around 4 kHz [4].

Transtympanic Steroid Therapy

Many publications regarding randomized clinical trials (RCT) have demonstrated the benefit of transtympanic steroids (TTS) as a rescue treatment after systemic steroids for SSNHL. Methyl-prednisolone showed the most promising profile, when considering drug concentration in the endolymph [27]. Battaglia obtained better results in those patients who received a combined therapy, oral steroids (60 mg per day, 7 days) plus TTS (dexamethasone 12 mg/ml once per week, 3 weeks), compared to the group that received TTS plus oral placebo. This last combination was more effective than oral steroids and transtympanic placebo [28].

Antiviral Therapy

There was no difference in the benefit from treatment with antiviral drugs (acyclovir) or administration of steroids [5], nor have other studies with valacyclovir [29, 30] shown any benefit of the antiviral drug for SSNHL.

Hyperbaric Oxygen Therapy

Administration of hyperbaric oxygen treatment has been described but is controversial [2].

Surgery

If a perilymphatic fistula is the cause of SSNHL, surgical treatment of the fistula can improve hearing [31].

Other Treatments

Ozone therapy (autohaemotherapy) has been tried in a RCT for sudden hearing loss [32]. A 100 ml of the patient's own blood with a gaseous mixture of oxygen and ozone was re-injected twice a week for 10 sessions. Seventy-seven percent of the treated patients showed a significant hearing recovery compared to 40% in the placebo group. Pure-tone averages and speech reception thresholds were also significantly better.

Tinnitus and SSNHL

Tinnitus is a common symptom in SSNHL. Approximately 91% of individuals with SSNHL report that they have tinnitus in the affected ear or in both ears [4]. Tinnitus occurs at the same time as hearing loss in some individuals with SSNHL and may be the first symptom before hearing loss. Tinnitus begins some days after the hearing impairment in some individuals. The tinnitus may be caused by sound deprivation caused by the hearing loss, which is known to be able to start central nervous system reorganization processes that can lead to tinnitus (see Chaps. 10, 12 and 21) and hyperacusis (see Chaps. 3 and 4).

Tinnitus Characteristics

We have previously shown [32] that 6.6% of the first 213 patients referred to our tinnitus clinic had SSNHL. Tinnitus onset was sudden in approximately 92% of the patients. The intensity of the tinnitus fluctuated in 46% of patients. Tinnitus psychoacoustical characteristics are shown in Table 56.5.

The tinnitus in SSNHL can imply a greater handicap than the hearing loss [2]. When the hearing resolves, either through treatment or spontaneously, the tinnitus may improve or disappear. Tinnitus may be a prognostic sign for the hearing loss [33].

Tinnitus Management

Tinnitus that accompanies SSNHL can be treated in a similar way as other forms of tinnitus (see chapters in Section V). All treatments for sudden hearing loss can be effective in treating the tinnitus. The use of steroids, vasodilatation drugs, and procedures or the hyperbaric oxygen therapy is often used when tinnitus accompanies SSNHL. At early stages of SSNHL,

sound stimulation is beneficial because it can prevent reorganization processes secondary to sound deprivation in the auditory nervous system. As remapping could be the physiological substrate for tinnitus development, customized sound enrichment would help to decrease the possibility of tinnitus and hyperacusis [34, 35] (see Chaps. 74, 75 and 76).

Conclusion

Most forms of SSNHL have no known cause. In management of patients with SSNHL, it is important to rule out other causes. Retrocochlear diseases, such as vestibular schwannoma and other central nervous system tumors, have been described to occur in 4% of SSNHL. They can be ruled out through tests such as ABR or MRI. Published results regarding the prognosis of SSNHL vary among studies. Some studies show 45–65% spontaneous recovery. The severity of the hearing loss, the audiometric profile, and the presence of vestibular symptoms affect the prognosis. Delay on starting treatment is associated with a worse prognosis. Steroid treatment is proven to be the most effective treatment, although it is not largely effective. Recent studies show promising results regarding the efficacy of steroids delivered through the eardrum. More than 50% of the patients showed a significant improvement of such treatment when administered after the failure of conservative therapy. Tinnitus accompanies most incidences of SSNHL. Tinnitus usually improves when hearing is partial or totally recovered, and individuals can also benefit from other forms of tinnitus treatment when there is no improvement of hearing.

References

1. De Klein A. Sudden complete o partial loss of function of the octavus-system in apparently normal persons. Acta Otolaryngol (Stockh) 1944; 32: 409–429
2. Stokroos RJ, Albers FWJ. Therapy of idiopathic sudden sensorineural hearing loss. A review of the literature. Acta Otorhino-laryngol Belg., 1996; 50: 77–84
3. Shikowitz MJ Sudden sensorineural hearing loss. Med Clin North Am. 1991; 75(6): 1239–50
4. Fetterman BL, Sanders JE, Luxford WM. Prognosis and Treatment of Sudden Sensorineural Hearing loss. Am J Otol 1996; 17: 529–536
5. Stokroos RJ, Albers FW, Tenvergert EM. Antiviral treatment of idiopathic sudden sensorineural hearing loss: a prospective, randomized, double-blind clinical trial. Acta Otolaryngol 1998; 118: 488–95
6. Klemm E, Deutscher A, Mösges R. A present investigation of the epidemiology in idiopathic sudden sensorineural hearing loss. Laryngorhinootologie 2009; 88: 524–527
7. Hughes GB, Freedman MA, Haberkamp TJ, Guay ME. Sudden neurosensorial hearing loss. Otolaryngol Clin North Am 1996; 29: 393–405
8. Belal A. The effect of vascular occlusion on the Human Inner Ear. J Laryngol Otol, 1979; 93: 955–68
9. Perlman HB, Kimura RS, Fernandez C. Experiments on temporary occlusion of the internal auditory artery. Laryngoscope 1959; 69: 591–613
10. Duckert LG, Meyerhoff W. Sudden hearing loss. In Meyerhoff WL, ed. Diagnosis and management of hearing loss. Philadelphia WB Saunders, 1980; 12–22
11. Van Dishoeck HAE, Bierman TA. Sudden perceptive deafness and viral infection. Ann. Otol Rhinol laryngol, 1957; 66: 963–80
12. Mattox DE, Simons FB. Nautral history of sudden sensorineural hearing loss. Ann Otol Rhinol Laryngol 1977; 86: 463–480
13. Kobayashi H, Suzuki A, Nomura Y. Unilateral hearing loss following rubella infection in an adult. Acta Otolaryngol (Stockh), 1994; 514: 49–51
14. Junicho M, Fushiki H, Aso S et al. Prognostic value of initial electronystagmography findings in idiopathic sudden sensorineural hearing loss without vértigo. Otol Neurotol 2008a; 29: 905–909
15. Süslü N, Yilmaz T, Gürsel B. Utility of anti-HSP 70, TNF-alfa, ESR, antinuclear antibody and antiphospholipid antibodies in the diagnosis and treatment of sudden s12ensorineural hearing loss. Laryngoscope 2009; 119: 341–6
16. Garcia-Berrocal JR, Ramirez-Camacho R, Trinidad A. Autoimmune hearing loss: improving diagnostic performance. Acta Otorrinolaryngol Esp 2007; 58: 138–42
17. Hong SM, Byun JY, Park CH et al. Saccular damage in patients with idiopathic sudden sensorineural hearing loss without vertigo. Otolaryngol Head Neck Surg 2008; 139: 541–5
18. Byl FMJ. Sudden hearing loss: eight years of experience and suggested pronostic table. Laryngoscope, 1984; 94 (5 Pt 1): 647–51
19. Plontke SK, Bauer M, Meisner C. Comparison of pure-tone audiometry analysis in sudden hearing loss studies: lack of agreement for different outcome measures. Otol Neurotol 2007; 28: 753–63
20. Kronenberg J, Almagor M, Bendet E, Kushnir D. Vasoactive therapy versus placebo in the treatment of Sudden Hearing loss: a double blind clinical study. Laryngoscope 1992; 102: 65–8
21. Kohen W, Nickol HJ. Horsturz-zur altersabhangigkeit der therapieergebnisse unter berucksichtiguntg vom naftodrofuryl (Dusodril). HNO, 1985; 33: 36–9
22. Hoffmann F, Beck C, Schutz A, Offerman P. Ginkgoextrakt EGB 171 (Tebonin)/HAES versus Naftidrofuryl (Dusodril)/HAES. Eine randomisierte studie zur horsturztherapie. Laryngol, Rhinol Otol, 1994; 73: 149–52

23. Mösges R, Köberlein J, Heibges A et al. Rheopheresis for idiopathic sudden hearing loss: results from a large prospective, multicenter, randomized, controlled clinical trial. Eur Arch Otorhinolaryngol 2008; 266: 943–53
24. Kubo T Matsunaga T. Efficacy of defibrinogenation and steroid therapies on sudden deafness. Arch Otolaryngol Head Neck Surg 1988; 114: 649–652
25. Wilson WR, Byl FM, Laird N. The efficacy of steroides in the threatment of idiopathic sudden hearing loss. Arch Otolaryngol 1980; 106: 772–6
26. Moon IS, Kim J, Lee SY et al. How long should the sudden hearing loss patients be followed after early steroid combination therapy?. Eur Arch Otorhinolaryngol 2009; 266: 1391–5
27. Parnes LS, Sun AH, Freeman DJ. Corticosteroid pharmacokinetics in the inner ear fluids: an amimal study followed by clinical application. Laryngoscope, 1999; 109: 1–17
28. Battaglia A, Burchette R, Cuevas R. Combination therapy (intratympanic dexamethasone + high-dose prednisone taper) for the treatment of idiopathic sudden sensorineural hearing loss. Otol Neurotol. 2009; 30: 254–5
29. Tucci DL, Farner JC Jr, Kithc RD et al. Treatment of sudden sensorineural hearing loss with systemic steroids and valacyclovir. Otol Neurotol 2002; 23: 301–8
30. Westerlaken BO, Stokroos RJ, Wit HP et al. Treatment of sudden sensorineural hearing loss with antiviral therapy: a prospective, randomized, double-blind clinical trial. Ann Otol Rhinol Laryngol 2003; 112: 993–1000
31. Ragab A, Shreef E, Behiry E et al. Randomised, double-blinded, placebo-controlled, clinical trial of ozone therapy as treatment of sudden sensorineural hearing loss. J Laryngol Otol 2009; 123: 54–60
32. Ruben RJ, Yankelowitz SM. Spontaneous perilymphatic fistula in children. Am J Otol. 1989; 10(3): 198–207
33. Herraiz C. Tinnitus management with retraining therapies. Doctoral Thesis. Universidad Autonoma Madrid, June 2002.
34. Hikita-Watanabe N, Kitahara T, Horii A, Kawashima T, Doi K, Okumura SI. Tinnitus as a prognostic factor of sudden deafness. Acta Otolaryngol. 2009 May 12: 1–5. [Epub ahead of print]
35. Norena AJ, Eggermont JJ. Enriched acoustic environment after noise trauma abolishes neural signs of tinnitus. Neuroreport 2006; 17: 559–563