

Sudden Sensorineural Hearing Loss

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The definition of sudden sensorineural hearing loss (SSNHL) is dependent on the purpose of the literatures and aim of each research to find out the improvement of their treatments. The National Institute on Deafness and Other Communication Disorders (NIDCD) in the United States defines SSNHL as the rapid loss of hearing threshold at least 30 dB in three consecutive frequencies within 3 days and it is considered as the medical emergency.¹ This definition is not universally accepted in all articles.² However, the Division of Otology, Neuro-otology and Balancing Problems at Siriraj Hospital determines SSNHL as the sudden loss of hearing threshold >30 dB in at least one frequency within 3 days.

The incidence of SSNHL in the United States has been reported by Byl to be 5-20 per 100,000 people per year.³ The incidence of SNHL at Siriraj Hospital between 2006-2008 shows 4,109 patients, in which SSNHL is 315 among 4,209 of the SNHL patients (7.67%). The female to male preponderance is about 1.56:1. The highest occurrence might be the cool season in Thailand, which may be caused from the outbreak of influenza and the bird flu according to the report of the Department of Disease Control (<http://thaigcd.ddc.moph.go.th/index.html>) in Thailand. (Fig 1)

The aetiology of SSNHL

The aetiology of SSNHL can be classified into 4 theories, which are the viral infection, the vascular disturbance, the autoimmune process and the labyrinthine

membrane rupture theories. The two most common theories that are usually postulated are the viral infection and the vascular disturbance theories.

The congenital infection that relates to the hearing loss (TORCH infection: cytomegalovirus, rubella, herpes virus or toxoplasmosis)² and the evidence that has been demonstrated in terms of the increasing antibody titer to several viruses⁴ introducing the concept of viral infection theory. Stokroos RJ, et al.,⁵ demonstrated the atrophy of the organ of the corti, stria vascularis as well as tectorial membranes after injection of herpes simplex virus type I into the perilymph of guinea pigs, which were similar to the pathological finding of the temporal bones of the patients who were diagnosed as viral labyrinthitis. Furthermore, one third of patients show the viral prodromal symptoms prior to the SSNHL⁶ and the history of upper respiratory tract infection prior to the onset of SSNHL which was reported by Mattox and Simmons⁷ in about 25% of patients also supported the viral infection theory.

The vascular supply of the inner ear is the end-on-artery, which is the labyrinthine artery. On account of vascular problems, the disturbance of the blood circulation may lead to the cochlear damage. Kimura RS, et al.,⁸ showed the animal models of labyrinthine artery occlusion resulting in a rapid and severe loss of the cochlear functions. The limitation of insufficiency of blood circulation can impair cochlear function after 60 seconds and the structural changes occurred after 30 minutes, which are the ganglionic cells, spiral ligaments as well as tectorial membranes affected. The decrease of cochlear blood flow more than one hour caused the permanent loss of both structures and functions.^{7,9,10} Moreover, patients with small cerebellar infarctions may present similar vestibular lesions including sudden hearing loss.¹⁰ The vascular insufficiency may cause the total and permanent or total and temporary problems of the auditory function depending of the severity of the vessel occlusion.⁹

The evidences of autoimmune mediated inner ear diseases were demonstrated in the animal models by Harris et al.¹¹ The model using bovine temporal bone antigen stimulated guinea pigs by systemic immunization. The model demonstrated the changes in the physiological measure of hearing and cochlear morphology. Furthermore, in humans, McCabe BF¹² presented 18

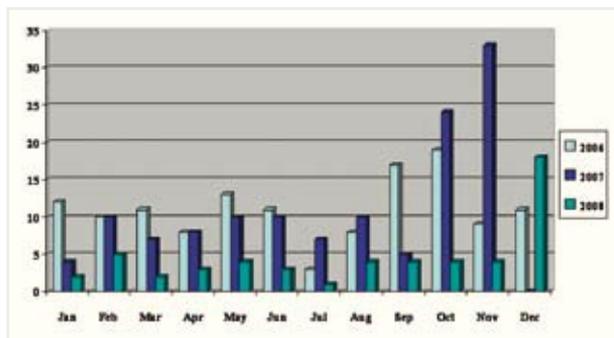


Fig 1. Incidence of SSNHL at Siriraj Hospital between 2006-2008.

patients with autoimmune SNHL who responded to corticosteroid and cyclophosphamide therapy. The SNHL related with known associated autoimmune diseases such as Cogan's syndrome, Wegener's granulomatosis, and temporal arteritis has been shown.²

The labyrinthine membrane rupture theory, it seems controversial. The presentation of either temporal bone fracture, or post stapedectomy may damage the internal membranes of the cochlear leading to cause SNHL.² The perilymphatic fistula from rupture of the round or oval window membrane including the barotrauma may cause the fluctuating of hearing loss. Nevertheless, there is no histological demonstration of postmortem temporal bone of patients who had SSNHL with possible rupture of the cochlear membrane.⁹

Management

The spontaneous recovery rate of SSNHL is about 32-70%¹³ that will impact the efficacy of treatment, thus the management of this disease is still controversial. Most of SSNHL studies can be divided into¹⁴

- The corticosteroid vs. placebo
- The corticosteroid vs. no treatment
- The corticosteroid plus other treatment vs. placebo plus the same other treatment
- The corticosteroid plus other treatment vs. the same other treatment

The other treatments that have been found in the studies such as the specific antiviral treatment are the vasoactive and hemodilution therapies (e.g. Carbogen inhalation, Ginkgo biloba) or the hyperbaric oxygen based upon the theories of aetiology.

The specific action of steroids on the cochlear is still unclear and needs further studies, but its action in decreasing inflammation and edema are chosen to treat this condition.¹⁴ The dosage and duration of treatment depends on the protocol of each study or academic institute. At Siriraj Hospital, the dosage of prednisolone used in treatment of ISSNHL is about 1 mg/kg daily for 7-14 days. The golden period for treatment is about 2 weeks after onset of SSNHL. Wilson WR, et al.,¹⁵ reported the improvement rate of 61% by the steroid treatment compared to the placebo 32%. On the other hand, Cinamon U¹⁶ showed no statistical significance between patients treated with steroid and placebo. These two studies were the randomized controlled studies, which all had confounding factors. The Cochrane review 2006¹⁴ concluded that the value of steroid treatment is still unclear because of no valid randomized controlled studies. Nevertheless, systemic corticosteroid seems to be the most accepted in most of the academic centers. In North America, steroid treatment has been proposed by some authors to be the gold standard of therapy.¹³

In the systematic reviews by Colin AE,¹³ the specific oral antiviral agents, such as valacyclovir or acyclovir, have been reported in two randomized controlled trials. Two of those have not shown any benefit on the SSNHL.

The Cochrane review 2007¹⁷ reported 6 randomized controlled trials demonstrating another method to improve perilymph oxygenation which is hyperbaric oxygen therapy (HBOT; 100% oxygen in a pressure chamber at 250 kPal for 60-90 minutes per session).¹⁸ The authors concluded that there were methodological

limitations of each study and HBOT might improve hearing and tinnitus, consequently these lead to the implications for research.

Neither carbogen inhalation (95% oxygen plus 5% carbon dioxide), vasoactive nor hemodilutional therapies (e.g. pentoxifylline, dextrans, Ginkgo biloba or nifedipine) showed positive results in the treatment of SSNHL.

The new promising therapy

It has been found that the formation of the reactive oxygen species (ROS) resulting from the noise trauma and ischemia to the cochlear including ototoxic drugs destroy the structure of cochlear, but the antioxidants including Glutathione (GSH) can protect the structural damages from ROS. Recently, Nagashima R & Ogita K²⁰ found that corticosteroids had the therapeutic effect at the spiral ganglion in murine cochlear resulting by scavenging of ROS by significant increase of the GSH level after steroid treatment. Additionally, Shimazaki T, et al.¹⁹ demonstrated that the glucocorticoid receptor was widely detected in lateral wall tissues, organs of the corti, spiral limbus, spiral ligament, and spiral ganglion, but the most abundant site was at the spiral ligament. It seems that corticosteroids have the capability for treatment of SSNHL, so the new route has been proposed.

Intratympanic steroid injection (IT) for treatment of SSNHL had been proposed by Silverstein H, et al.²¹ In guinea pig models, comparison of the concentrations of hydrocortisone, dexamethasone, and methylprednisolone in the inner ear fluid following oral, intravenous and intratympanic administration have been established by Parnes LS, et al.²² They found that the highest concentration of the corticosteroids in the cochlear fluid was intratympanic application, which also reached the inner ear fluid faster than the other administration as well. Moreover, methylprednisolone reached the highest concentration and remained for the longest period of time in the endolymph compared to the other corticosteroids.²³

It seems likely that methylprednisolone should be the best drug for intratympanic application. Many studies have been conducted on methylprednisolone, in which the results are successful or fail. Nevertheless, there were many studies performed on dexamethasone. The trend in using dexamethasone is increasing in the developing countries because it is more economic and abundant.

In the IT literatures, many authors showed the effectiveness of the transtympanic route for SSNHL. They proposed their own protocols in application of IT both frequency and drugs (dexamethasone vs. methylprednisolone). Although the groups of patients in each research are small, it seems likely to be more effective treatment comparing to the conventional treatment. Thus, the IT may be use for SSNHL in term of primary treatment for SSNHL, adjunctive therapy given collaterally with systemic steroid or salvage therapy after failure of systemic steroids.²

CONCLUSION

Sudden sensorineural hearing loss (SSNHL) is commonly defined as the sudden onset of a decrease of at least 30 dB of hearing threshold on one's own within 24 to 72 hours in three consecutive frequencies.

However, the definition of this problem is different between institutes and protocols in researches. SSNHL may be called sudden idiopathic sensorineural hearing loss because the majority of patients are unable to find the aetiologies, though the theories of the disease have been proposed, which are the viral infection, the vascular insufficiency, the autoimmune process and the labyrinthine membrane rupture theory.

The treatment of this disease is still controversial. Corticosteroids seem widely use for therapy. The other options of treatment to serve each theory that have been postulated such as hyperbaric oxygen, volume expander and the antiviral agents did not show the statistical significance in treatment. The new approach of treatment that many researchers demonstrated is the intratympanic steroid injection. Nevertheless, the result of this method of treatment is still controversial and needs further intensive trials.

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