A CRITICAL ANALYSIS OF THE MANAGEMENT PROTOCOLS FOR IDIOPATHIC SUDDEN SENSORI-NEURAL HEARING LOSS.

Shahid A. Shah , Asif Karim, Rehman Ghani

Department of ENT, Head & Neck Surgery, Ayub Medical College, Abbottabad

Background: Sudden sensori-neural hearing loss (SSNHL) is a clinical dilemma with great diversity in presentation and poorly understood pathogenesis and hence no definitive treatment protocol as yet. Both sexes are affected, middle age to elderly being the commonest age group. A variety of causes have been implicated as responsible for this condition, but most of the times it is difficult to isolate one, and hence most of the times a battery of investigations proves to be a clinical exercise. A number of treatment protocols have been suggested and used over the years, based on presumed etiological theories, claiming varying degrees of success. **Methods:** Relevant literature available on the net regarding the management and the efficacy of various treatment regimens for ISSNHL was critically analyzed by the authors (who are professorial staff of a medical college and consultants of a teaching hospital) to develop a consensus and recommendations on the most appropriate protocol. **Results:** It was asserted that various treatment regimens have not proved beyond doubt to be superior to one another or spontaneous recovery rates. **Conclusion:** SSNHL is a medical emergency that entails thorough investigations to search for a possible cause and institution of appropriate therapy. Failing identifying a cause, i.e idiopathic group, combination therapy with steroids and antiviral drugs could prove beneficial provided treatment is instituted early. A number of placebo controlled trials consuming various modalities are needed to determine an optimal treatment of ISSNHL. Psychological and psychiatric assistance has a certain role and so has the rehabilitation in the management of these patients.

Key words: Deafness, Sensori-neural hearing loss, idiopathic, Management.

INTRODUCTION

Sudden sensori-neural hearing loss (SSNHL) has been defined and described varyingly from time to time. It was first described in the literature by De Kleyn in 1944.[1]

There had always been arguments that whether or not it has to be a loss from a previously normal level of hearing or a rapid deterioration in an already diseased ear with elevated hearing thresholds? Then how sudden it has to be? Whether happening overnight, over a week or month, need to be addressed as well. Lastly the severity of the condition is, no doubt, an important factor, as a slight loss over few hours may seem more sudden than a moderate loss over months.

However, a loss of greater than 35dB in at least three adjacent frequencies over a period of three days or less is an acceptable definition of this clinical condition to most.[2]

Reported overall incidence of SSNHL ranges from 5% to 20% per 100,000 persons per year. SSNHL may affect both sexes equally. All age groups may be affected, however 75% of patients are more than 40 years age and 1.4% are below the age of 10. The mean overall age for SSNHL is 46 years, according to a large series by Mattox and Simmons.[3]

Mostly the presentation is unilateral but it may present bilaterally. Byl in1984, in a series of 225 patients, noted bilateral SSNHL in 2% of patients [4]

50% of patients present with sudden onset, however, more than one third of people with SSNHL awaken in the morning with a hearing loss, whereas the remainder exhibit rapidly progressive hearing loss. Classically the patient experiences sudden hearing loss with pain or pressure sensation in the ear. Tinnitus is a feature in most of the cases and a significant number of patients may feel vertiginous. The amount of hearing loss may vary from mild to severe, and may involve different parts of the hearing frequency range. SSNHL may be temporary or permanent.[5]

Reports estimate that the etiology of SSNHL is diagnosed in only 10% of cases. These include perilymph fistula, vascular causes, such as embolic phenomenon, thrombosis, vasospasm, and hypercoagulable or high viscosity states, infections, autoimmune disorders with systemic manifestations, metabolic disorders, trauma, ototoxic drugs, noise induced hearing loss and tumours. However, despite extensive evaluation, majority of cases eludes definitive diagnosis and therefore, are labeled as Idiopathic sudden sensori-neural hearing loss(ISSNHL). Viral infection is presumed to be the underlying mechanism in a majority of these patients.[6]

Other possible mechanisms are autoimmune inner ear disease(AIED)[7], vascular compromise and cochlear membrane rupture.[8]

Treatment is directed towards the cause if identified. In cases where no cause is found, treatment is empirical, emphasizing mainly on improving blood supply and oxygenation of the inner ear.

Reported positive outcome is more or less same with all different combinations and not much different from spontaneous recovery rates, (65%), reported by Mattox and Simmons.[3] Byl also reported a recovery rate of about 69%.[4] Those that recover 50% of hearing in the first 2 weeks following SSNHL have a better prognosis than those who do not recover at this rate.[9] Recurrence of SSNHL is rare but possible.[10]

This study is an analysis of the published literature regarding the efficacy of the current management protocol of the ISSNHL.

Material and methods

Relevant literature available on the medscape, medline and PubMed was collected. Authors critically analyzed the literature, both in support and against the current treatment practice.

Inclusion and exclusion criteria for quality of literature were developed and the parameters for inclusion included the methodology, setting of study and the impact factor of the journal in which the study was published.

Studies citing cases treated for a defined cause were excluded. Finally, a consensus was developed regarding appropriate management protocol for ISSNHL.

RESULTS

Literature search revealed studies regarding the management of ISSNHL, based on various etiological presumptions. These studies were evaluated for support of scientific basis to their claims and results were inferred.

Haberkamp and Tanyeri reviewed the management of ISSNHL and noted that while numerous treatments have been studied aiming to improve blood flow, such as carbogen inhalation, all remain controversial or simply lack

convincing evidence of efficacy. Very few placebo controlled studies have been conducted in the treatment of ISSNHL and for this reason; there is presently a limited ability to determine what the optimal treatment is ?[11]

Schweinfurth et al, applying treatment protocols including vasodilators, Plasma expanders, anticoagulants & carbogen inhalations have shown no improvement over the rate of spontaneous recovery.[12]

Wilkins and associates, in 1987, treated 109 patients with a "shotgun" regimen that included dextran, histamine, hypaque, diuretics, steroids, vasodilators, and carbogen inhalation, and there was no significant difference between those patients receiving and not receiving treatment. The results suggest that this "shotgun" approach for treatment of sudden hearing loss offers no better outcome than is reported in the literature for spontaneous recovery.[13]

Lamm studied the efficacy of drug treatment in SSNHL, and found that cochlear blood flow was only temporarily improved during infusion of blood flow promoting drugs and cochlear oxygenation was deteriorated with most of these drugs. It was concluded that treatment of basic or accompanying diseases is currently the only effective therapy and prophylaxis of SSNHL.[14]

Kronenberg and colleagues, in a double blind clinical study, compared vasoactive treatment versus placebo in the treatment of sudden hearing loss and the results did not suggest any superiority regarding therapeutic efficacy of vasodilator to a placebo.[15]

There is no conclusive data proving carbogen inhalation to be of any benefit over spontaneous recovery, however, Fisch , found that in patients with sudden deafness, the oxygen supply to the vestibular tissues is significantly reduced but the response to carbogen is still possible and recommends carbogen inhalation for the effective, noninvasive treatment of sudden deafness.[16]

Gordin et al, in 2002, studied the efficacy of carbogen as well as MgSO4 in treating SSNHL but didn't secure enough evidence to advocate their use.[17] In 1980, Wilson and colleagues performed double-blind studies for the treatment of SSNHL with oral steroids or placebo and included additional controls who received no treatment. It was found that steroids had a significant effect on the recovery of hearing in patients with hearing loss between 40 and 90 db and that patients with isolated midfrequency losses recovered without regard to therapy.[2]

Huang and colleagues, studied the efficacy of hypaque and steroids in the treatment of SSNHL, and concluded that none of the currently available regimens produce consistently better results than the spontaneous recovery rate of 65% reported by Mattox & Simmons.[18]

Zadeh et al, designed a study to determine the efficacy of steroid and antiviral therapy in the management of SSNHL in human beings, and claim a recovery rate exceeding the spontaneous recovery rates (73%). Ninety-one percent of patients with vertigo and all patients with mid-frequency hearing loss and up-sloping hearing loss recovered with treatment.[19]

On the other hand, Tucci found no benefit of Valacyclovir plus steroids over steroids alone in a multicenter study of 84 subjects.[20]

Strokroos and colleagues in 1999 conducted an animal study to determine the efficacy of combination treatment with an antiviral and a steroid in animals whose ears were inoculated with herpes simplex virus type 1 (HSV-1) and found the combination treatment better compared to treatment with either acyclovir or prednisolone alone [21]

Hyperbaric Oxygen Therapy (HBOT) aims at increasing perilymphatic pO2. It involves rising arterial pO2 to levels of approx. 1800 mmHg.

Lamm et al., have shown in animal studies that HBOT, but not normobaric 100% oxygen breathing, could induce a rise in the perilymphatic pO2 by 500-900%.[22]

Dauman et al compared HBOT/vasodilator/ corticotherapy to vasodilator/corticotherapy alone and to haemodilution therapy and found that there was no significant difference in the outcome between different groups.[23]

Pilgramm et al compared 37 patients, being treated with haemodilution with or without HBOT, but did not find any significant advantage in the HBOT group.[24]

On the other hand, Fattori et al suggested that hyperbaric oxygen therapy was the treatment of choice.[25]

Piracetam, a rheoactive agent, has been studied in treating SSNHL, presuming increased blood viscosity as a possible cause for the insult. Piracetam and prednisolone combination was compared with steroid/vasodilator therapy. The piracetam group (n=17) showed clinical improvement in 82.3% and a mean hearing gain in 54.1%, compared with 68.7% and 49.3%, respectively, for the group without piracetam (n = 6).[26]

Discussion

Evaluation and management of SSNHL should be considered medically urgent, if not an emergency. The primary goal is to rule out any treatable cause.

It has been found that a number of different strategies have been tried and adopted by different practitioners to manage ISSNHL, based on their individual experience, with proponents and opponents of anything that has been tried so far.

Furthermore, still most of the claims are based on patient's response to treatment and there is no definite understanding of the exact nature of the pathogenesis of this clinical entity as yet, to enable practitioners develop a treatment protocol backed by scientific evidence.

Majority of clinicians would treat this entity with a combination of vasodilators, carbogen inhalation and steroids, emphasizing on increasing the blood supply and oxygenation of the inner ear, presuming ischaemia to be the main reason for the insult, to which there is no scientific evidence.

Vasodilator agents have been found on many occasions without an edge over the spontaneous recovery rates. However, Carbogen inhalation, that has not shown any superiority in short term, has been claimed as successful regarding improvement in hearing to a significant degree in long term, i.e after a year.

When a treatment of ISSNHL is used, it often consists of burst of steroids such as prednisone. Evidence to date for a good effect is mixed. Some studies suggest a better hearing prognosis for treated versus untreated patients [11,] and others a worse prognosis.[28]

Early trials of steroid therapy, whereby, low doses were used for short duration, showed disappointing results; however, later studies using higher doses for longer periods of time have been more promising.

Mostly steroids are administered as oral preparation but better prognosis has been claimed with very high doses of intravenous prednisolone.[27]

Recently, transtympanic steroids have been used with good response, in persons who were unable to tolerate oral steroids.[29]

Steroids also have shown encouraging therapeutic response in AIED, whereby patient presents with progressive bilateral sensori neural hearing loss without any identifiable cause, and absence of systemic involvement by the autoimmune process. In case of relapse, higher dose of steroids is recommended and cytoxic agent, methtraxate or cytoxan, may be added in case of continous failure of response.[30].

Whereas, McCabe prefers cytotoxic therapy, cytoxan, over steroids in the treatment of AIED, because of the higher response rate.[31]

Viral disease appears to be the basis for about 60% of all cases of ISSNHL. There is history of preceding viral infection in 30-40% cases.

Serological studies have demonstrated a statistically significant increase in viral seroconversion in patients with ISSNHL compared with controls for CMV as well as influenza B, mumps, rubeola, and varicella zoster viruses.[6.]

Antivirals seem reasonable, given the frequency that herpes family viruses have been associated with SSNHL.

Medications like acyclovir or valacyclovir may be unhelpful when the cause is a virus that is not in the herpes family, and one rarely knows at the time of the hearing loss which if any virus is responsible. It is also possible that this sort of treatment is just too late in the course of the disorder. However, combining steroids and an antiviral, if treatment can be started within three to four days of the insult, may prove a favorable choice to most.

The efficacy of HBOT has not been conclusively established. A disadvantage of these studies is that they initiated therapy as soon as possible after the onset of deafness, thereby including the large number of patients who would recover spontaneously, no matter how or even if treated.

However, success has been claimed with HBOT which is encouraging enough, but still more studies are needed to confirm the results.

There is probably no other disease for which such a variety of treatments have been proposed, and still today, many different treatment regimens, some more invasive than others, are propagated. Their therapeutic efficacy is very difficult to establish. It seems however, that the therapeutic outcome of several proposed drug treatment regimes is in the same range as the spontaneous recovery rate.

Conclusion

None of the treatment outcome has been proved convincingly superior to another or to spontaneous recovery rates.

Because of high spontaneous recovery rate, treatment is not always felt necessary, especially when impairment is minor.

A preceding viral infection is a strong indication for antiviral therapy.

Antiviral medication is beneficial, provided given early and effective against herpes group. Combination with an anti-inflammatory drug will have a synergistic effect.

Corticosteroids have shown to be effective in a particular frequency range of hearing loss, especially following prolonged treatment and at higher doses.

Recommendation

It is mandatory that any patient presenting with SSNHL should be regarded as a medical emergency.

Thorough history should be taken and physical examination conducted in search of a possible cause.

A baseline audiological assessment followed by serial monitoring of the auditory function is mandatory over the next few weeks.

All relevant laboratory investigations should be done to rule out any haematological causes.

Radiological investigation would include plain films and CT scan if trauma to the skull is the apparent cause. MRI scan is the tool to rule out intracranial pathology.

Appropriate treatment is instituted if indicated on the basis of investigations, however, if no possible cause is defined, the condition is labeled as idiopathic and steroid therapy may still be given as most of the experience and research has found it beneficial in some percentage of patients.

Furthermore, antiviral therapy may be considered and instituted as early as possible, if there is convincing clinical evidence of a preceding or associated viral insult.

Psychological/ Psychiatric assistance will be appreciated as the patient must be undergoing great mayhem. Reassurance will have significant role until patient experiences recovery in hearing.

When the hearing loss is first identified, patient should be assessed regarding rehabilitation needs and if the hearing loss persists rehabilitation begins that entails patient as well as family counseling, emphasizing communication strategies.

A number of placebo controlled trials consuming various modalities are needed to determine an optimal treatment of ISSNHL.

REFERENCES

1. De Kleyn A. Sudden complete or partial loss of function of the octavus system in apparently normal persons. Acta Otolaryngol 1944; 32:407-29

2. Wilson WR, Byl FM, Laird N. The efficacy of steroids in the treatment of idiopathic sudden hearing loss. A double-blind clinical study. Arch Otolaryngol 1980 ;106(12):772-6.

3. Mattox DE, Simmons FB. Natural history of sudden sensorineural hearing loss. Ann Otol Rhinol Laryngol 1977;86:463-80.

4. Byl FM. Sudden hearing loss: eight years experience and suggested prognostic table. Laryngoscope 1984;94:647-61.

5. Yimtae K, Srirompotong S, Kraitrakul S. Idiopathic sudden sensorineural hearing loss. J Med Assoc Thai 2001; 84(1):113-9

6. Wilson WR, Veltri RW, Laird N, Sprinkle PM. Viral and epidemiologic studies of idiopathic sudden hearing loss. Otolaryngology Head and Neck Surgery 1983;91(6):653-8

7. McCabe BF. Autoimmune sensorineural hearing loss. Ann Otol Rhinol Laryngol 1979;88:585-9.

8. Simmons FB. Theory of membrane breaks in sudden hearing loss. Arch Otolaryngol 1968;88:41-8.

9. Ito S, Fuse T, Yokota M, Watanabe T, Inamura K, Gon S, Aoyagi M. Prognosis is predicted by early hearing improvement in patients with idiopathic sudden sensorineural hearing loss. Clin Otolaryngol 2002;27(6):501-4

10. Furuhashi A, Matsuda K, Asahi K, Nakashima T. Sudden deafness: long-term follow-up and recurrence. Clin Otolaryngol 2002;27(6):458-63

11. Haberkamp TJ, Tanyeri HM. Management of sudden sensorineural hearing loss. AM J Otol 1999;20:587-95.

12. Schweinfurth JM, Parnes SM, Very M. Current concepts in the diagnosis and treatment of sudden sensorineural hearing loss. Eur Arch Otorhinolaryngol 1996;253(3):117-21.

13. Wilkins SA Jr, Mattox DE, Lyles. A Evaluation of a "shotgun" regimen for sudden hearing loss. Otolaryngol Head & Neck Surg 1987;97(5):474-80

14. Lamm K. Drug therapy of sensorineural hearing loss - critical remarks. Wien Med Wochenschr 1992;142(20-21):455-9.

15. 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(1):65-8.

16. Fisch U. Management of sudden deafness. Otolaryngol Head & Neck Surg 1983;91(1):3-8

17. Gordin A, Goldenberg D, Golz A, Netzer A, Joachims HZ. Magnesium: a new therapy for idiopathic sudden sensorineural hearing loss. Otol Neurotol 2002;23(4):447-51.

18. Huang TS, Chan ST, Ho TL, Su JL, Lee Fp. Hypaque and steroids in the treatment of sudden sensorineural hearing loss. Clin Otolayngol 1989;14(1):45-51

19. Zadeh MH, Storper IS, Spitzer JB. Diagnosis and treatment of sudden-onset sensorineural hearing loss: a study of 51 patients. Otolaryngol Head Neck Surg 2003;128(1):92-8

20. Tucci DL, Farmer JC Jr, Kitch RD, Witsell DL. Treatment of sudden sensorineural hearing loss with systemic steroids and valacyclovir. Otol Neurotol 2002;23(3):301-8.

21. Stokroos RJ, Albers FWJ, Schirm J. Therapy of idiopathic sudden sensorineural hearing loss; antiviral treatment of experimental herpes simplex virus infection of the inner ear. Ann ORL 1999;108::423-8

22. Lamm CH, Walliser U, Schumann K, Lamm H. Sauerstoffpartialdruckmessungen in der Perilymphe der Scala tympani unter normo- und hyperbare Bedingungen. HNO 1988;36: 363-6.

23. Dauman R, Cros A, Poisot D. Traitement des surdités brusques: premiers résultats d'une étude comparative. J Otolaryngol 1985;14:49-56.

24. Pilgramm MH, Lamm K. Schumann. Zur hyperbaren Sauerstofftherapie beim Hörsturz. Laryng Rhinol Otol 1985;64:351-4.

25. Fattori B, Berrettini S, Casani A, Nacci A, De Vito A, De Iaco G. Sudden hypoacusis treated with hyperbaric oxygen therapy: a controlled study. Ear Nose Throat J 2001;80(9):655-60.

26. Garcia-Callejo FJ, Velert-Vila MM, Morant-Ventura A, Orts-Alborch MH, Marco-Algarra J, Blay-Galaud L. Pathophysiological rationale for the use of piracetam in sudden deafness: Acta Otorrinolaringol Esp 2000;51(4):319-26

27. Alexiou C, Arnold W, Fauser C, Schratzenstaller B, Gloddek B, Fuhrmann S et al. Sudden sensorineural hearing loss. Does application of glucocorticoids make sense? Arch Otolaryngol Head Neck Surg. 2001;127(3):253-8.

28. Minoda R, Masuyama K, Habu K, Yumoto E. Initial steroid hormone dose in the treatment of idiopathic sudden deafness. Am J Otol 2000;21;819-25

29. Gianoli GJ, Li JC. Transtympanic steroids for treatment of sudden hearing loss. Otolaryngol Head Neck Surg 2001;125(3):142-6

30. Harris JP, Sharp P. Inner ear autoantibodies in patients with rapidly progressive sensorineural hearing loss. Laryngoscope 1990;100:516-24

31. McCabe BF. Autoimmune inner ear disease: results and therapy. Adv Otorhinolaryngol 1991; 46:78-8.

Address for Correspondence:

Dr Shahid A. Shah , Department of ENT, Head & Neck Surgery, Ayub Medical College, Abbottabad. Pakistan

Email: shahidalishah@ayubmed.edu.pk