SEAN FLANAGAN F.R.A.C.S
Conjoint Lecturer UNSW

Otolaryngology Head and Neck Surgery
Neurotology, Skull Base Surgery
Cochlear Implant Surgery

Sudden Sensorineural Hearing Loss (SSNHL)

The acute onset of hearing loss of 30 dB in three contiguous frequencies which may have occurred instantaneously or progressively over several days. [1,2]
Patients often describe a numbness or blockage as opposed to an obvious hearing loss.
The incidence is 5 to 20 per 100,000 patients per year. [3]
Untreated, spontaneous recovery has been reported to range from 30-60%. [4]
While a viral or microvascular aetiolgy is proposed, it is essentially a diagnosis of exclusion: cerebellopontine angle tumours, auto-immune disease, multiple sclerosis (MS), infectious aetiologies,intralabyrinthine haemorrhage [5], perilymph fistula and Meniere's disease must be excluded. [6]
Poor prognostic factors include a severe, flat pattern to the hearing loss [7] and associated vestibular dysfunction. [8]

Investigation

Otoscopic examination is normal. Occasionally a middle ear effusion can result in toxic inner ear damage.
A Weber tuning fork test lateralising to the worst hearing ear is highly suggestive of a sensorineural hearing loss.
Audiometry with speech discrimination
The utility of any blood tests unless there is a history of systemic autoimmune type symptoms is very marginal.[2]
MRI brain and CPA: urgent if other neurologic signs and symptoms. Up to 4% of cases will reveal a vestibular schwannoma. [9,10]

Management

Oral Steroids are the only effective therapy shown in randomised controlled trials.[11] Varying doses and length of therapy are used. [12,13] Combination therapy provides the best overall response.[14,15]
Generally the benefit to hearing occurs if therapy started within 7 days, with treatment worthwhile out to 14 days. [4]
30-50% will show no response. [16]
The general approach is to treat for a week and then repeat a hearing test.
High dose Vitamin A, C, E, Selenium, Magnesium have been reported to add minor benefit to steroids. [17,18]
No definitive benefit from antivirals or haemodilution agents has been seen.[11] [19-22]
Some supporting evidence for hyperbaric oxygen therapy as an adjunct has been published.[23-26] [The Hyperbaric Unit at Prince of Wales Hospital is the only option in Sydney]
Consideration of alternative therapies such as acupuncture can at least help from a psychologic perspective.[27]
A repeat audiogram is arranged at the 7 day mark can assist in decisions regarding ongoing/repeated steroid or other adjuvant treatments.[28]

Steroid Treatment Regimes

Oral Prednisone: Up to 1mg/kg per day, for at least a week, followed by a taper over the next couple of weeks.[14,29,30]

Intratympanic Steroid: [Dexamethasone, Methylprednisolone, Triamcinolone].[31] The theory is to allow higher intra-cochlear concentrations of medication via absorption through the round window membrane.
The regime is variable with options to perform a single transtympanic dose either under local anaesthetic or general anaesthesia, multiple transtympanic doses (3-4 doses 3-5 days apart[2], weekly dose for 3 doses[14]) or to place a ventilation tube to allow self administration. [32,33] The published concentration of steroids also varies;

St Vincent's Campus
Ground Floor, 67 Burton St
Darlinghurst NSW 2010
Australia

Shire Family ENT
Suite 16, 20-24 Gibbs St
Miranda NSW 2288
Australia

All Bookings and correspondence
Tel: +61 2 9380 5380
Fax: +61 2 9360 5419
Email: sean@sydneyentclinic.com
Web: sydneyentclinic.com/flanagan
ABN: 54035430358

dexamethasone ranges from 4mg/ml up to 24mg/ml.

	Oral Corticosteroids	Intratympanic Corticosteroids
Timing of treatment	Immediate, ideally within first 14 days. Benefit has been reported up to 6 weeks following onset of sudden sensorineural hearing loss (SSNHL)	Immediate Salvage (rescue) after systemic treatment fails
Dose	Prednisone 1 mg/kg/d (usual maximal dose is 60 mg/d) or Methylprednisolone 48 mg/d or Dexamethasone 10 mg/d	Dexamethasone 24 mg/mL or 16 mg/mL (compounded), or 10 mg/mL (stock)Methylprednisolone 40 mg/mL or 30 mg/mL
Duration/frequency	Full dose for 7 to 14 days, then taper over similar time period	Inject 0.4 to 0.8 mL into middle ear space every 3 to 7 days for a total of 3 to 4 sessions
Technique	Do not divide doses	Anterosuperior myringotomy after topical anesthetic Inject solution into the posterior inferior quadrant via narrow-gauge spinal needle to fill middle ear space Keep head in otologic position (one side down, affected ear up) for 15 to 30 minutes
Monitoring	Audiogram at completion of treatment course and at delayed intervals	Audiogram before each subsequent injection, at completion of treatment course, and at delayed intervals Inspect tympanic membrane (TM) to ensure healing at completion of treatment course and at a delayed interval

Summary of Clinical Approach to Steroid Therapy [reproduced from American Clinical Guidelines]

Success of around 40% in selected patients, defined as at least 20dB or 20%. [32,34-36] Although not all studies have reproduced these results. It is important to note that approximately 10% will show some improvement after more than 1 month [37] when analysing results of salvage therapy. [14 4,36,38,39]

Measuring Success of Treatment

Siegel's criteria for hearing recovery

	Type	Hearing recovery
I.	Complete Recovery	Patients whose final hearing level was better than 25 dB regardless of the size of the gain
II.	Partial Recovery	Patients who showed more than 15 dB of gain and whose final hearing level was between 25 and 45 dB
III.	Slight recovery	Patients who showed more than 15 dB of gain and whose final hearing level was poorer than 45 dB
IV.	No improvement	Patients who showed less than 15 dB of gain

[40]

It is important to realise that to experience true binaural hearing, that hearing thresholds must be within 15-20dB in both ears.
In rare cases, where there has been a response to steroids, but further decline once the prednisone is reduced, and long term high dose prednisone is required, steroid sparing agents such as methotrexate are considered.

Conclusion

SSNHL is an otologic emergency which requires prompt diagnosis and institution of management in order to optimise the chance of recovery. It is theorised that most cases are related to the reactivation of a virus within the inner ear, but microvascular disease, other infective agents, autoimmune processes and tumours also are occasionally implicated. The patient must be thoroughly informed as to the limitations of current treatments. Bilateral sudden sensorineural hearing loss is much rarer, but is more likely to be associated with significant systemic disease.[41]

Rehabilitation

It must be noted that the sudden loss of hearing can cause a significant psychologic impact and this in of itself must be addressed and managed.
Consideration must be given to hearing rehabilitation. Assistive listening devices and hearing aids are useful when there is measurable hearing. BiCross or a bone anchored hearing aid, BAHA is an option when there is a residual severe sensorineural hearing loss. It provides the sensation of binaural hearing in a majority of cases by re-routing

a signal to the contra-lateral ear.[42,43] The other option is cochlear implantation. This allows stimulation of the damaged cochlear allowing true binaural hearing, and is especially useful if tinnitus is a particular problem.[44-47] Vestibular rehabilitation is also an important component in the management of this condition, if the balance is also affected.

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