INTRODUCTION

The diagnosis and management of ISSNHL has been always controversial and is likely to remain so. This pessimistic view is put forward on the basis that when the definition and diagnosis of a condition is difficult, defining optimal management is always problematic. Despite being a rare presentation, but is very problematic. Despite being a disease of unknown/putative causes and unpredictable outcome, standard documentation/ definitions/ plan of management are needed. A large array of empiric treatments has been used and serious side effects can occur from such empiric treatment(s). The lack of one or more uniformly accepted treatment(s)/investigation(s) potentially increases the cost of management/ risks on patients. Even the presence of clinical guidelines doesn’t mean that all physicians are aware of it, and its implementation may be difficult, impractical or not feasible. Therefore, it is important for each hospital/ local department to provide clinicians with evidence-based local protocol/ agreement in evaluating patients with ISSNHL, with particular emphasis on management. As being a disease of unknown/putative causes and unpredictable outcome, standard documentation/ definitions/ plan of management are needed. A large array of empiric treatments has been used and serious side effects can occur from such empiric treatment(s). The lack of one or more uniformly accepted treatment(s)/investigation(s) potentially increases the cost of management. In 90% of cases, or even more, the underlying cause is unknown or uncertain at the time of presentation, it is appropriate, therefore, to approach these cases in a common way [2].

Patients with ISSNHL may present in emergency departments or referred to otolaryngology clinics. Though there may be available guidelines, it is important to standardize the management in a local hospital/ department, as not every physician might be aware of the available guidance, and there is sometimes a fear from doctors to do nothing and/or no treatment policy. Also, the moral demand of beneficence must outweigh any anxiety the physician might have about others’ criticisms of their actions, therefore, a standard protocol might be the safe gate for every doctor to justify his/her management.

Many individual departments and individual otolaryngologists treat patients with ISSNHL according to a protocol which has been agreed locally or which they have developed over the years. In this protocol we use mainly the statements of the clinical guidelines for sudden hearing loss published in 2012 American Academy of Otolaryngology Head and Neck Surgery Foundation (AAOHNSF) [2] Not only is the protocol in line with recent EBM and guidelines, it is used by local hospitals/ departments in actual practice to provide safe and high standard care. Though it is intended to represent the local care for patients’ management through easy clear steps that every physician can follow, as it is important to standardize the management of ISSHL in a local hospital/ department, but it can mainly be a nucleus upon which further audit/research can proceed to build up more EBM and measure its practicality.

Definition of ISSNHL

Though it is a rare disease with incidence 5 to 20 per 100,000 populations [3,4], a definition of ISSNHL was necessary as it is easier to recognize than it is to define. Lots of definitions have been introduced, but perhaps Wilson et al definition in 1980 remains the most universally acceptable definition that is still used in literature excessively [5]. In this protocol a modification is suggested in the definition to make it suspected ISSNHL, which is a standardized documentation of the diseases and its idiopathic nature, pending appropriate investigations or time to exclude other causes

It entails definition of suspected ISSNHL:

1. New hearing loss of at least 30 dB in three consecutive frequencies
2. Over 72 hours or less
3. An underlying cause cannot be identified by initial history and physical examination.

As ISSNHL has to be a new hearing loss, it is crucial to standardize documentation locally, consequently, clinicians must decide the degree of certainty they are comfortable with when making a decision that the hearing loss in the poorer ear is de-novo [6]. The levels of certainty of the pre-morbid hearing thresholds are outlined in Table (1) [6].

As suggested by the first statement of AAOHNSF guidelines that clinicians should distinguish Sensorineural hearing loss (SNHL) from conductive hearing loss (CHL) in a patient presenting with sudden hearing loss [2]. However, a practical time-efficient scheme of doing so through proper initial history taking and physical examination (including neurological examination) can rule out many possible causes. The scheme includes basic clinical steps to follow, even if audiological evaluation is not readily available in emergency sets. Therefore, the clinical physician can document easily with more confidence the suspected idiopathic nature. The scheme of doing this is outlined in Table (2).

Etiology

Listing the putative causes of ISSNHL is out of the scope of this protocol, but it is important to emphasize that bilateral ISSNHL is relatively rare [7,8] and should raise concern for certain specific causes, some of which are outlined in Table (3). Identification of such patients is crucial, as they have a high likelihood of alternative and potentially serious life-threatening underlying cause.

Investigations

As per available EBM and recent guidelines it is important to discourage routine laboratory tests, as they do not improve management/outcome of patients with ISSNHL but nonetheless have associated cost and potential harms related to false-positive results [2].

It is also important that unless history/examination strongly suggestive of alternative cause, clinicians should not order CT scan of the head/brain in the initial evaluation (This does not apply to patients with focal neurological findings). However, retro-cochlear lesions have to be excluded in all cases by obtaining a magnetic resonance imaging (MRI) of brain, internal auditory canals (IACs) and cerebello-pontine angle (CPA). The specific MRI protocol used will often depend on the local radiological resources/experience available [9-14]. In this protocol a request an urgent MRI is to be done, once initial history/examination is not suggestive of alternative cause, even if MRI is not readily available in emergency/ clinic at time of presentation

Ten to twenty percent of patients with a vestibular schwannoma may present with ISSNHL, [15] but the rate of vestibular schwannoma in patients who present with ISSNHL is somewhat lower, ranging from 2.7% to 10.2% of patients who are evaluated with MRI.

Though of its rarity, it is an important cause to exclude it through an MRI scan. Firstly, it is crucial to rule out an important statistically significant cause that can be managed or monitored later. Secondly excluding it avoids leaving the patient terrified of potential intracranial causes of his/her sudden hearing loss and leaves in the limbo of living with an unknown cause of such sudden deafness [16-18].

TREATMENT

Oral steroids

There has been counter-indicatory evidence of the value of oral steroids in ISSNHL. An early systematic review, Conlin and Parnes, in 2007, found no effectiveness of corticosteroids [19], while recent meta-analysis and Cochrane database reviews of various medical treatments, including corticosteroids, showed a slight improvement with oral steroid therapy compared with placebo [20,21].

Steroids therapy is one of the few treatment options that have evidence of its efficacy. The greatest improvement in hearing occurs during the first two weeks, although benefit has been reported up to 6 weeks following onset of deafness [2,22-24].

Counseling patients about options of treatment, including no treatment policy and the uncertainty of the outcome are very important. Also clear discussion about possible risks of steroids should be highlighted and well understood by the patient. The idea in discussion and decision making should be that when so little is known about the efficacy of treatment, the safety of individual/tailored treatments is particularly important. In this protocol, it is suggested that unless contraindication/unsafe, advise is to use oral Prednisolone 1mg/kg/day (max 60mg/day) for 7 days tapered over the next week, not to divide doses, and perform Audiogram at completion of treatment course, in a consultant lead ENT clinic [2].

Intra-tympanic (IT) steroid injection

Studies showed that initial IT steroid injections are not more effective than using oral steroids alone, and even combination of both together initially didn't show much difference in hearing improvement [25,26].

Patient who have no improvement, including those given corticosteroid treatment and/or observation treatment can be offered IT injection of steroids, as it has been proposed by...
Table 1: Degree of certainty about pre-morbid hearing [4].

<table>
<thead>
<tr>
<th>Degree of certainty about pre-morbid hearing</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Very certain: Patient had previous Pure tone Audiometry (PTA)</td>
</tr>
<tr>
<td>2. Certain: Patient history not suggestive of previous ear problems or hearing loss (HL)</td>
</tr>
<tr>
<td>3. Fairly certain: Patient had a longstanding HL and reports that the current episode of HL is subjectively poorer.</td>
</tr>
<tr>
<td>4. Uncertain: the clinician unsure 1, 2, 3 OR feels there was some pre-morbid HL but never documented/ unsure.</td>
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</tbody>
</table>

Modified criteria from Burton MJ, et al. [4]

Table 2: Initial evaluation scheme for those presenting with Sudden hearing loss.

Initial evaluation scheme for those presenting with Sudden hearing loss

Exclude other pathology through:
- History
- Examination including: Otoscopy, tuning fork tests (TFTs) and neurological examination
- Pure tone Audiometry (PTA) (If out of hours, TFTs and free field audiometry and arrange PTA)

Exclude conductive hearing loss: PTA/ TFTs
If history suggestive of acute Stroke= discuss case with on call stroke/medical team urgently
Patient should be discussed with ENT doctor and assessed by ENT team within 24 hours

Table 3: Possible causes of sudden bilateral sudden ISSNHL may be: [28]

<table>
<thead>
<tr>
<th>Possible causes of sudden bilateral sudden ISSNHL may be:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Metabolic</td>
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<tr>
<td>• Vascular</td>
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<tr>
<td>• Autoimmune</td>
</tr>
<tr>
<td>• Infectious</td>
</tr>
<tr>
<td>• Toxic</td>
</tr>
<tr>
<td>• Traumatic</td>
</tr>
<tr>
<td>• Inflammatory</td>
</tr>
</tbody>
</table>

Table 4: Studies of Intra tympanic Steroid as Salvage Therapy.

<table>
<thead>
<tr>
<th>Study/ number of patients</th>
<th>Time of initiating therapy</th>
<th>Dose/Method of Injection</th>
<th>Improvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ahn et al (2008) 2</td>
<td>&lt;2 weeks after oral failure</td>
<td>0.3-0.4 mL of 5 mg/mL Dexamethasone 2 times per week for 2 weeks</td>
<td>(43.8%) early ITD (30.0%) mid-ITD (15.4%) late ITD</td>
</tr>
<tr>
<td>Ho et al (2004) 32</td>
<td>Within 2 weeks after oral methylprednisolone</td>
<td>1 mg/mL dexamethasone once per week for 3 weeks</td>
<td>53.3%</td>
</tr>
<tr>
<td>Slattery et al (2005), 20</td>
<td>Up to 3 months after oral steroids</td>
<td>62.5 mg/mL methylprednisolone, 4 injections over a 2-week</td>
<td>55%</td>
</tr>
<tr>
<td>Choung et al (2006), 33</td>
<td>&lt;28 days after oral steroids</td>
<td>5 mg/mL dexamethasone, 2 injections per week for 2 weeks</td>
<td>38.2%</td>
</tr>
<tr>
<td>Dallan et al (2006), 8</td>
<td>unknown</td>
<td>40 mg/mL methylprednisolone, single injection</td>
<td>75%</td>
</tr>
<tr>
<td>Xenellis et al (2006), 40</td>
<td>&lt;2 weeks after IV prednisolone</td>
<td>0.5 mL of 40 mg/mL, 4 injections over 2 weeks</td>
<td>47%</td>
</tr>
</tbody>
</table>

Abbreviations: ITD, Intratympanic Dexamethasone; IV, Intravenous

a number of authors as an option to obtain additional hearing recovery. There is no general consensus on the definition of the failure of initial management or the subsequent improvement after IT steroid injections, with limited and variable guidance from the literature as to what level of residual hearing loss qualifies a patient for salvage (Tables 4, 5).

Despite the limitations and variability of the existing research, the majority demonstrated a consistent benefit in restoration of hearing beyond that afforded by initial therapy.

In this protocol the decision to use of salvage IT steroid injections should be considered, if no improvement to initial therapy (and/or oral steroids declined/contraindicated) [27-32].

Clinicians should take into consideration the risks and benefits of the treatment. Suggested usage is IT steroids injections, once weekly dexamethasone, for a maximum of 3 weeks, after discussion of possible risks/benefits with the patient)

Follow up

Another follow up pure tone audiogram should be done
Table 5: (Continue) Studies of Intra tympanic Steroid as Salvage Therapy.

<table>
<thead>
<tr>
<th>Study/ number of patients</th>
<th>Time of initiating therapy</th>
<th>Dose/Method of Injection</th>
<th>Improvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haynes et al (2007), 40 patients</td>
<td>&lt;40 days</td>
<td>24 mg/mL dexamethasone, single injection</td>
<td>26.7%</td>
</tr>
<tr>
<td>Roebuck and Chang (2006), 29</td>
<td>After 5 to 7 days of oral steroids</td>
<td>24 mg/mL dexamethasone, single injection</td>
<td>33%</td>
</tr>
<tr>
<td>Plaza et al (2007), 9 patients</td>
<td>&lt;5 days after IV methylprednisolone</td>
<td>20 mg/mL methylprednisolone, 3 injections over 1 week</td>
<td>55%</td>
</tr>
<tr>
<td>Kilic et al (2007), 19 patients</td>
<td>After a 3-week course of high-dose systemic corticosteroid</td>
<td>0.5 mL of 62.5 mg/mL, 5 injections over 12 days</td>
<td>73.6%</td>
</tr>
<tr>
<td>Gouveris et al (2005), 21 patients</td>
<td>&lt;2 weeks after oral steroids</td>
<td>0.3-0.4 mL of 8 mg/mL dexamethasone every 2 days</td>
<td>33%</td>
</tr>
<tr>
<td>Silverstein (1996), 8 patients</td>
<td>&lt;30 days</td>
<td>Dexamethasone via microwick, 3 times per week for 3 to 4 weeks</td>
<td>25%</td>
</tr>
</tbody>
</table>

**Abbreviations:** ITD: Intratympanic Dexamethasone; IV: Intravenous

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**Definition of suspected ISSNHL:**

New hearing loss of at least 30 dB, in three consecutive frequencies, over 72 hours or less AND An underlying cause cannot be identified by initial history and physical examination

**Initial assessment:**

- History
- Examination including: Otoscopy, tuning fork tests (TFTs) and neurological examination
- Pure tone Audiometry (PTA) (if out of Hours, TFTs and arrange PTA)
- Exclude conductive hearing loss: PTA/TFTs
- Patient should be discussed with ENT and assessed by ENT within 24 hours

**Management:**

- Request MRI Internal Auditory Canal and CPA
- Unless history strongly suggestive of alternative cause/contraindication/ declined:
  - **Commence oral steroids** after discussion of possible risks/benefits with the patient
  - Use oral Prednisolone 1mg/kg/day (max 60mg/day) for 7 days, tapered over the next week
  - Arrange follow-up for patient with repeat PTA on arrival in ENT clinic (within 10-14 days)
  - Consider blood investigations for specific patients where history is suggestive of autoimmune hearing loss

If no improvement (and/or oral steroids declined/contraindicated), consider **Intra-tympanic steroids injections** (after discussion of possible risks/benefits with the patient) (once a week, for three weeks followed by repeat PTA)

Counsel patients about possibility of incomplete recovery of HL/ possible benefits of HAs and other supportive measures (if required)

**Figure 1** Flowchart protocol for management of ISSNHL.
within 6 months of diagnosis with ISSNHL.

Follow up for patients with ISSNHL is very important to assess their residual hearing thresholds, the need for hearing aids, and for counseling. Increase awareness that counseling and education for patients is needed [33-36]. Clinicians should counsel patients with incomplete recovery of hearing about the possible benefits of amplification and hearing assistive technology (HAT), including benefits of different Hearing Aids (HAs), and other supportive measures (if required) [2,37-40].

CONCLUSIONS

Figure 1 shows the protocol for management of ISSNHL. This protocol represents a good practice with high levels of clinical effectiveness in use. It is feasible, applicable and in line with recent EBM and guidelines. It can be used by local hospitals/departments in actual practice to provide safe, high standard, cost-effective health care and with reasonable risk-benefit. The protocol is aimed to be a one page sheet, to be clear and easy to follow on display panels in clinics/emergency departments or on intranet to be viewed or printed easily.

Though it is intended to represent the local care for patients’ management through easy clear steps that every physician can follow, as it is important to standardize the management of ISSNHL in a local hospital/department, but it can mainly be a nucleus upon which further audit/research can proceed to build up more EBM and measure adherence of clinicians to it.

REFERENCES


