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Sudden Sensorineural Hearing Loss: An Otologic Emergency

Introduction

Sudden sensorineural hearing loss (SSNHL) is a potentially disabling condition that requires prompt recognition and management. As frontline clinicians, GPs are often the first to evaluate patients presenting with rapid hearing decline. Early intervention is crucial for optimal recovery, underscoring the importance of understanding its presentation, diagnostic pathway, and current management paradigms.

Definition

- **SSNHL** is classically defined as a *rapid loss of ~30 dB across three consecutive frequencies within 72 hours*, confirmed by audiometry. However,
- It is commonly unilateral, but bilateral cases can occur.

Presentation

- **Key Symptoms:**
  - Sudden unilateral hearing loss (most common).
  - +/- Tinnitus
  - +/- Aural fullness
  - Sometimes vertigo or dizziness.
  - No pain or external ear pathology.

Clinical Note: Patients may report difficulty understanding speech, even with normal volume. The onset is often abrupt, and patients may notice the change immediately upon awakening or during daily activities.

Causes

The etiology of SSNHL is idiopathic in approximately 60-70% of cases, but potential causes include:

- **Viral infections** (e.g., herpes simplex virus).
- **Vascular compromise** affecting the cochlear blood supply.
- **Autoimmune inner ear disease.**
- **Trauma.**
- **Neoplastic causes**, such as vestibular schwannoma.
- **Other causes:** Ménière's disease, ototoxic medications, Syphilis.

Note: Often, no definitive cause is identified (70%).

Epidemiology

- **Incidence**: SSNHL affects approximately **5 to 20 per 100,000 people per year** in Australia and elsewhere. It accounts for about **1 in 5,000 to 1 in 30,000 individuals** annually.
- **Age & Demographics:** It occurs across all age groups but is most common between 40-60 years. Slightly more prevalent in males.
- **Recovery Rates:** Spontaneous recovery occurs in approximately **30-60%** of cases, particularly with early treatment.

Diagnostic Approach

1. **High suspicion of Sensorineural Hearing Loss; eliminating causes of Conductive Hearing loss**
   - Often, examination is normal
   - Rule out conductive hearing loss findings ie infections, perforations, effusions, trauma
   - Tuning Fork (512Hz) Test
     - Weber's Test - Lateralises to contralateral ear in SNHL
     - Rinne's Test
       - Air Conduction > Bone Conduction (Normal) in SNHL
       - **Clinical Note:** False negative seen in total SNHL ('dead' ear)
         - Bone conduction will appear louder as sound is conducted through bone to contralateral ear

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2. Immediate Referral for Audiometric Evaluation

- Conducted urgently, ideally within 24-48 hours.
- Confirms cochlear origin (Sensorineural) of hearing loss.
- Helps quantify severity and monitor response to therapy.

3. Neuroimaging

- **MRI with gadolinium contrast** is the gold standard for excluding retrocochlear pathology such as vestibular schwannoma.
- **Should be performed as soon as possible, especially if:**
  - Initial audiometry is inconclusive.
  - There is associated vertigo or neurological signs.
  - Hearing loss persists despite treatment.

4. Laboratory Tests

- Not routine but may include blood glucose, lipid profile, autoimmune panels, VDRL depending on clinical suspicion.

Management Principles

1. Prompt Initiation of Corticosteroids - do not wait for investigation results if suspicious

High-dose corticosteroids are the cornerstone treatment:

- Oral prednisolone: 1 mg/kg/day (up to 60-75 mg/day) for 7-14 days, with gradual tapering.
- The best outcomes are observed when treatment starts within 2 weeks of symptom onset.

2. Salvage Therapy: Intra-tympanic Steroid Injection

Consider referring for intra-tympanic steroids if:

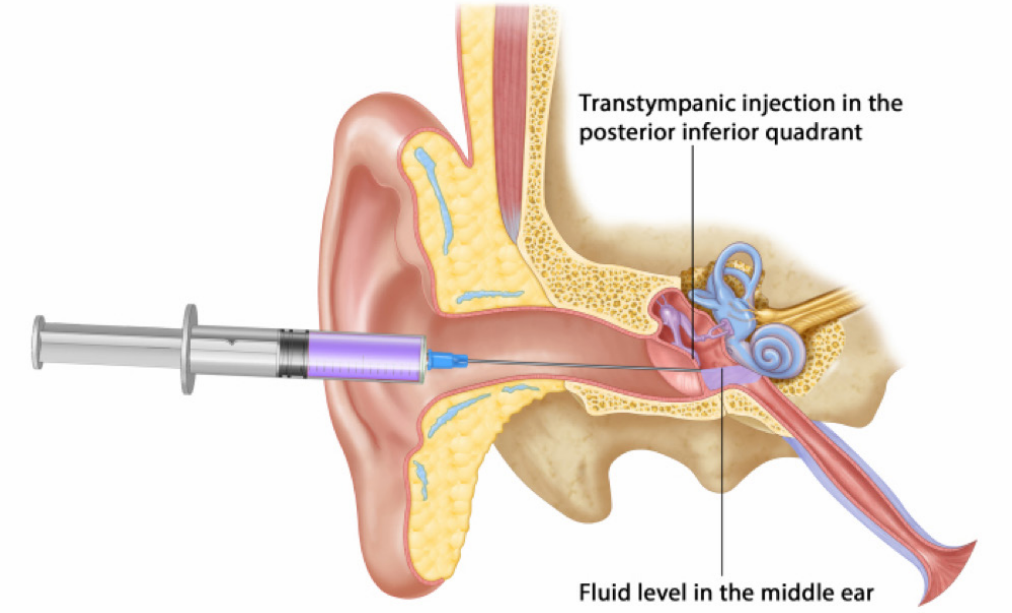
- systemic steroids are contraindicated
- initial therapy is unsuccessful or
- if delayed presentation (>30 days)

What is intra-tympanic steroid injection?

- It involves delivering steroids directly into the middle ear via a tympanic membrane (eardrum) perforation or through a minimally invasive technique.
- The steroid solution (commonly dexamethasone or methylprednisolone) diffuses across the round window membrane into the cochlea, achieving higher local concentrations with fewer systemic effects.

Procedure Details:

- **Preparation**: The patient is usually seated; topical anesthesia is applied.



- **Injection:** Under microscopic vision, a fine needle or cannula is used to inject the steroid solution through the tympanic membrane into the middle ear.
- **Post-procedure:** The patient can usually return home immediately. The injection may be repeated over several sessions depending on response.

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Evidence for intra-tympanic steroids:

- Multiple studies, including randomized controlled trials, demonstrate improved hearing recovery when intra-tympanic steroids are added to systemic therapy
- **Evidence:** A 2020 Cochrane review indicates intra-tympanic steroids improve hearing recovery, especially in combination with oral steroids.
- **Guidelines:**
  - American Academy of Otolaryngology–Head and Neck Surgery (AAO-HNS) 2019 recommends prompt corticosteroid therapy.

What happens after?

- Unlikely to occur again, but counselling regarding management of potential future episodes to optimise outcomes
- Repeat Audiogram in 6-12 months to check hearing has stabilised
- Consider hearing rehabilitation
  - Hearing Aids: Note if age <26 years or >75 years, eligible for government subsidy
  - Cochlear Implant - worth considering sooner for patients with viral aetiology, as cochlear sclerosis occurs
- Consider **neuroimaging** to exclude retro-cochlear pathology.
- **Refer to ENT promptly** for specialist assessment, including possible intra-tympanic therapy.

Summary

- Recognize SSNHL as an **urgent condition.**
- Initiate **prompt corticosteroid therapy.**
- Ensure **early referral** for **audiometry** within 24-48 hours.
- Consider **neuroimaging** to exclude retro-cochlear pathology.
- **Refer to ENT promptly** for specialist assessment, including possible intra-tympanic therapy.

References

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