Quick Review: Sudden Sensorineural Hearing Loss

Definition: Sudden hearing loss (SHL) is a medical emergency for which definitive diagnosis and treatment is still largely unknown. It has been defined for research purposes and has been accepted by most authorities as 30 dB or more sensorineural hearing loss over at least three contiguous audiometric frequencies occurring within 3 days or less.

Etiology: The etiology of SHL can be broken down into broad categories-
1. Viral and infectious
2. Autoimmune
3. Labyrinthine membrane rupture/traumatic,
4. Vascular,
5. Neurological,
6. Neoplastic

History and Physical examination:
Evaluation and management of SHL should be considered medically urgent, if not an emergency. The primary goal is to rule out any treatable causes.
- Thorough history and physical exam, details of the circumstances surrounding the hearing loss and the time course of its onset should be elicited.
- Associated symptoms, such as tinnitus, vertigo or dizziness, and aural fullness should also be asked about.
- Patients should also be questioned about previous otologic surgery, ototoxic drug use, and previous or concurrent viral or upper respiratory tract infections.
- Any history of trauma, straining, diving, flying, and intense noise exposure should be noted.
- Past medical history of other diseases associated with sudden hearing loss should also be obtained such as diabetes, autoimmune disorders, malignancies, neurologic conditions (multiple sclerosis), and hypercoagulable states

A complete head and neck exam should be performed on all patients with sudden hearing loss. More often than not, the exam will be unremarkable, however, any processes such as middle ear effusions, infections, cholesteatoma, and cerumen impaction should be excluded.

A thorough neurological exam including Weber and Rinne, cerebellar and vestibular testing should be performed.

Diagnostic evaluation:
An audiogram (pure tone, speech, tympanometry, including stapedial reflex testing) should be performed on all patients with sudden hearing loss. Serial testing provides documentation of the progression or resolution of the hearing loss and response to treatment.

The following laboratory studies can be ordered. Initial screening tests should be directed based on history and suspected conditions, CBC, ESR, Glucose, Lipid profile, Thyroid profile, VDRL, HIV. MRI is recommended by
the majority of authors for patients with asymmetric hearing loss. MRI is useful in evaluating for acoustic tumors, multiple sclerosis and cerebrovascular accidents.

**Known Treatable Causes of Sudden Sensorineural Hearing Loss:**

**Autoimmune:** Autoimmune hearing loss may be associated with or part of systemic autoimmune diseases such as Cogan’s syndrome, Wegener’s granulomatosis, polyarteritis nodosa, temporal arteritis, Buerger’s disease (thromboangitis obliterans), and systemic lupus erythematosis, or may be primary to the inner ear.

**Traumatic:** Traumatic breaks in the membranous labyrinth are accepted causes of sudden hearing loss. Cochlear membrane breaks may be either intracochlear, as is thought to occur in Meniere’s disease, or involve the labyrinthine oval and/or round windows with a resultant perilymph fistula. The patients history will usually elicit an inciting event such as a blow to the head, sneezing, bending over, lifting a heavy object, exposure to sudden changes in barometric pressure (such as during flying or diving), or exposure to a loud noise.

**Neoplastic:** Acoustic neuromas are usually associated with gradually progressive hearing loss.

**Idiopathic Sudden Sensorineural Hearing loss (ISSNHL): Etiologic Theories:** Viral, autoimmune, vascular

**Treatment:**

The **treatments for cases with known etiologies** involve addressing the underlying condition (i.e. treatment of: acoustic neuroma with excision, ototoxicity with cessation of ototoxic drugs, multiple sclerosis with medical therapy, embolic disease with anticoagulants, sickle-cell crisis with oxygen, bacterial meningitis with antibiotics and so on).

**Therapy for ISSNHL** is a subject of controversy. The high spontaneous recovery rate of ISSNHL (47% to 63%) and its low incidence make validation of empirical treatment modalities difficult. Many treatment regimens have been proposed for ISSNHL:

- Antiinflammatory/immunologic agents (Steroids, Prostaglandin, Cyclophosphamide, Methotrexate)
- Diuretics (Hydrochlorothiazide/triamterene,Furosemide)
- Antiviral agents: Acyclovir, Valacyclovir
- Vasodilators (5% carbon dioxide with 95% oxygen, carbogen, Papaverine)
- Volume expanders/hemodilutors (Hydroxyethyl starch, Low-molecular-weight dextran)
- Defibrinogenators, Calcium antagonists