ACUTE HEARING LOSS

Background
1. Definition: Categorized as conductive, sensorineural, or mixed
   - Conductive hearing loss occurs when sound cannot travel through the external ear, middle ear, or both
   - Sensorineural hearing loss results from pathology of cochlea, eighth cranial nerve, or central auditory pathways
   - Mixed hearing loss is combination of both conductive and sensorineural
2. General Information: Sudden sensorineural hearing loss (SNHL) is rapid loss of hearing that occurs all at once or over a period of up to 3 days diagnosed as at least a loss of 30 decibels in three connected frequencies

Pathophysiology
1. Pathology of Disease:
   - Infections (viral, acute otitis media, external otitis, syphilis, Lyme disease) causing SNHL from toxins affecting the inner ear through direct viral invasion or latent virus reactivation
   - Immunologic hypothesis based on theory that circulating antibodies cross-react with inner ear antigens or activated T cells and damage the inner ear
   - Hydrops and perilymphatic fistula cause abnormalities of labyrinthine fluid
   - Thrombosis, embolic occlusion, and hyperviscosity state (e.g. polycythemia vera) may result in cochlear ischemia
   - Diabetes mellitus, atherosclerosis, and sickle cell anemia obstruct small vessels
2. Incidence, Prevalence
   - Estimated incidence between 5 and 20 per 100,000 persons per year
   - Typically occurs between 43 years and 53 years of age
   - Equal gender distribution according to several large case series of 7500 cases in United States, Europe and Japan
   - Approximately 1% of cases related to vestibular schwannoma, demyelinating disease or stroke
   - 10 to 15% due to other causes such as Meniere’s disease, trauma, autoimmune disease, syphilis, Lyme disease, or perilymphatic fistula
   - Majority are idiopathic
3. Risk Factors
   - Head trauma, noise, and barotrauma
   - Retrocochlear neoplasm in cerebellopontine angle
   - May be presenting symptoms of multiple sclerosis
   - Associated ophthalmologic disease should be investigated for Susac’s syndrome (rare immunological disorder characterized by encephalopathy, branched retinal arterial occlusion [BRAO], and hearing loss)
   - Ototoxic medications (aminoglycosides, diuretics, chemotherapy)
   - Conductive (otosclerosis, chronic otitis media with effusion, malleus head fixation, tympanosclerosis, myringosclerosis, tympanic membrane perforation, cholesteatoma, ossicular chain disruption)
   - Autoimmune disease (polyarteritis nodosa, relapsing polychondritis, sarcoid, ulcerative colitis, systemic lupus erythematosus, Wegener’s disease, Churg-Strauss syndrome, Behcet’s disease, Cogan’s syndrome)
4. Morbidity / Mortality
   - 15% with Sudden SNHL will have hearing loss that gets worse over time

**Diagnostics**
1. History
   - Current symptoms/PHx:
     - Unilateral or bilateral
     - Fluctuating or constant
     - Chronology
     - Current and past treatments with oral and intravenous medications, nonprescription drugs
     - Screen for systemic disease
     - Prior ear surgery, cardiac bypass, surgery, and lumbar puncture
     - Family history of hearing loss, neoplasms, renal disease, and balance disorders
     - Previous sharp or blunt head trauma, noise trauma, barotrauma
   - Sudden SNHL symptoms:
     - Immediate rapid hearing loss, or hearing loss upon wakening
     - Majority are unilateral
     - Tinnitus and ear fullness
     - Vertigo
     - Ear pain or paresthesia

2. Physical Exam
   - Visualize and palpate the auricle
   - Examine external auditory canal and tympanic membrane
   - Evaluate drum mobility and middle ear effusion with pneumatic (air inflation) otoscopy or tympanogram
   - Perform Weber and Rinne tests (512 Hz tuning fork)
   - Softly whisper simple words or numbers and ask patient to repeat
   - Instruct patient to hum and report hearing asymmetry (sound lateralizes to affected ear in conductive and unaffected ear in SNHL)
   - Evaluate cranial nerves

3. Diagnostic Testing
   - Audiology
     - Sensorineural hearing loss: sensitivity to bone-conducted and air-conducted sound stimuli are equally reduced in affected ear (thresholds elevated)
     - Conductive hearing loss: bone conduction normal bilaterally, air-conducted thresholds elevated in affected ear (sensitivity reduced)
   - Speech testing
     - Subject given list of words and asked to repeat them
     - Speech reception threshold is sound level at which 50% of spoken words are understood
     - Speech recognition score is percentage of spoken words understood at 40 dB above speech reception threshold
   - Tympanometry
     - Assesses mobility of tympanic membrane and pressure status of middle ear

4. Diagnostic Imaging
MRI
- Rule out retrocochlear abnormality (e.g. neoplasm, stroke, or demyelination)
- CT or auditory brain-stem response audiometry
  - Alternatives for patients who cannot have MRI
  - Less sensitive than MRI for detecting retrocochlear abnormality

**Differential Diagnosis**

1. **Key Differential Diagnoses:**
   - **Acute**
     - Sudden idiopathic SNHL
     - Infection (acute otitis media, external otitis, syphilis, Lyme disease, viral)
     - Perilymphatic fistula
     - Ischemia of retrocochlear structures
     - Multiple sclerosis
     - Autoimmune disease
     - Traumatic causes
     - Metabolic (chronic renal failure)
     - Hematologic (sickle cell anemia)
   - **Rapidly progressive**
     - Autoimmune inner ear disease
     - Meningeal carcinomatosis
     - Vasculitis secondary to infection (Rocky Mountain spotted fever)
     - Lyme disease
     - Otoxic exposure (aminoglycosides, diuretics, chemotherapy)

2. **Extensive differential diagnoses:**
   - **Fluctuating**
     - Perilymphatic fistula
     - Meniere’s disease
     - Multiple sclerosis
     - Migraine-associated hearing loss
     - Infection (syphilis)
     - Autoimmune (Cogan’s syndrome, systemic lupus, polyarteritis nodosa, Wegener’s syndrome, temporal arteritis, scleroderma)
     - Sarcoïdosis
   - **Gradual**
     - Presbycusis
     - Noise-induced
     - Familial
     - Retrocochlear neoplasm
     - Chronic otitis media, cholesteatoma
     - Otosclerosis
     - Endocrine (hypothyroidism, diabetes mellitus)
     - Paget’s disease
     - Metabolic (chronic renal failure, hyperlipoproteinemia)
     - Mucopolysaccharidosis
Therapeutics

1. Acute Treatment:
   - Oral corticosteroids (prednisone or methylprednisolone) tapered over 10 to 14 days
     - Immediate treatment for unilateral idiopathic sudden hearing loss and additional symptoms (dizziness or tinnitus) is 14-day course of 60 mg prednisone (with taper). EBM rating: (SOR:C)\(^7\)
     - Spontaneous recovery occurs within the first 2 weeks after onset of Sudden SNHL
     - Greatest recovery of hearing when corticosteroids is initiated within 2 weeks; minimal benefits if greater than 4 weeks from symptom onset
     - Systemic steroids cannot be considered gold standard for Sudden SNHL; benefits remains unclear\(^8,9,10\)
     - Audiogram should be done within 24 to 48 hours after treatment for documenting extent of hearing loss

2. Further Management:
   - Intratympanic corticosteroids as primary or salvage therapy\(^4,11,12,16,18\)
     - Rationale: delivers high concentration to specific tissue with less systemic effects
     - As primary treatment, appears equivalent to treatment with high-dose oral prednisone therapy
     - As salvage therapy for patients who do not improve with oral treatment, may result in hearing improvement
     - May cause discomfort, less convenient, more costly
   - Randomized trials comparing corticosteroids alone to corticosteroids plus antiviral agents failed to show added benefit from antiviral agents\(^4,6,10,13\)
   - Vasodilators and vasoactive substances showed no evidence of benefit; studies were poor quality and number of patients were small\(^4,6,9,14\)
   - No evidence of benefits from hyperbaric oxygen; its use is not recommended\(^4,6,13,15\)
   - Superiority of fibrinogen-LDL-apheresis over standard first line treatments not established\(^17\)

3. Long Term Care:\(^4\)
   - Scuba diving contraindicated due to risk of tympanic membrane rupture, hearing loss, tinnitus, and balance problems
   - Ear plugs or earmuffs should be used to protect against loud noises or music
   - Unaffected ear should have immediate otolaryngologic evaluation to assess for any signs/symptoms of pathology

Follow Up

1. Return to Office: follow up preferably face-to-face to
   - provide support,
   - assess efficacy of therapy and stage of recovery,
   - address any concerns or side effects
2. Refer to Specialist: immediate referral to otolaryngologist
3. Admit to Hospital: not specified unless other serious diagnoses (e.g. stroke) considered
Prognosis
1. Not well documented
2. Patients with Sudden SNHL may have a 1.64 times greater risk of stroke during 5 years follow up compared to patients undergoing appendectomy\textsuperscript{4,6}
3. Patients with Sudden SNHL should have audiometric monitoring repeated over the course of a year to monitor recovery, direct rehabilitation (hearing aids), and monitor signs of relapse in affected ear or development of hearing loss in contralateral ear\textsuperscript{4}

Prevention
3. Not well documented

Patient Information
1. Information for patients:

References

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