ACUTE HEARING LOSS

Background
1. Definition: Categorized as conductive, sensorineural, or mixed1,5
   o Conductive hearing loss occurs when sound cannot travel through the external ear, middle ear, or both
   o Sensorineural hearing loss results from pathology of cochlea, eighth cranial nerve, or central auditory pathways
   o 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 frequencies2

Pathophysiology
1. Pathology of Disease:3
   o 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 reactivation13
   o Immunologic hypothesis based on theory that circulating antibodies cross-react with inner ear antigens or activated T cells and damage the inner ear13
   o Hydrops and perilymphatic fistula cause abnormalities of labyrinthine fluid
   o Thrombosis, embolic occlusion, and hyperviscosity state (e.g. polycythemia vera) may result in cochlear ischemia
   o Diabetes mellitus, atherosclerosis, and sickle cell anemia obstruct small vessels
2. Incidence, Prevalence4
   o Estimated incidence between 5 and 20 per 100,000 persons per year
   o Typically occurs between 43 years and 53 years of age
   o Equal gender distribution according to several large case series of 7500 cases in United States, Europe and Japan
   o Approximately 1% of cases related to vestibular schwannoma, demyelinating disease or stroke
   o 10 to 15% due to other causes such as Meniere’s disease, trauma, autoimmune disease, syphilis, Lyme disease, or perilymphatic fistula
   o Majority are idiopathic
3. Risk Factors3
   o Head trauma, noise, and barotrauma
   o Retrocochlear neoplasm in cerebellopontine angle
   o May be presenting symptoms of multiple sclerosis
   o Associated ophthalmologic disease should be investigated for Susac’s syndrome (rare immunological disorder characterized by encephalopathy, branched retinal arterial occlusion [BRAO], and hearing loss)
   o Ototoxic medications (aminoglycosides, diuretics, chemotherapy)
   o Conductive (otosclerosis, chronic otitis media with effusion, malleus head fixation, tympanosclerosis, myringosclerosis, tympanic membrane perforation, cholesteatoma, ossicular chain disruption)
   o 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
   o 15% with Sudden SNHL will have hearing loss that gets worse over time

Diagnostics
1. History
   o Current symptoms/PHx:\(^3\)
     ▪ 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
   o Sudden SNHL symptoms:\(^4\)
     ▪ Immediate rapid hearing loss, or hearing loss upon wakening
     ▪ Majority are unilateral
     ▪ Tinnitus and ear fullness
     ▪ Vertigo
     ▪ Ear pain or paresthesia
2. Physical Exam\(^1,4\)
   o Visualize and palpate the auricle
   o Examine external auditory canal and tympanic membrane
   o Evaluate drum mobility and middle ear effusion with pneumatic (air inflation) otoscopy or tympanogram
   o Perform Weber and Rinne tests (512 Hz tuning fork)
   o Softly whisper simple words or numbers and ask patient to repeat
   o Instruct patient to hum and report hearing asymmetry (sound lateralizes to affected ear in conductive and unaffected ear in SNHL)
   o Evaluate cranial nerves
3. Diagnostic Testing
   o Audiometry\(^1,4\)
     ▪ 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)
   o Speech testing\(^1,5\)
     ▪ 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
   o Tympanometry\(^5\)
     ▪ Assesses mobility of tympanic membrane and pressure status of middle ear
4. Diagnostic Imaging\(^4\)
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MRI
- Rule out retro cochlear 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 retro cochlear 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 retro cochlear 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)
     - Sarcoidosis
   - Gradual
     - Presbycusis
     - Noise-induced
     - Familial
     - Retro cochlear 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 unclear⁸,⁹,¹⁰
     - 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 therapy⁴,¹¹,¹²,¹⁶,¹⁸
     - 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 agents⁴,⁶,¹⁰,¹³
   - Vasodilators and vasoactive substances showed no evidence of benefit; studies were poor quality and number of patients were small⁴,⁶,⁹,¹⁴
   - No evidence of benefits from hyperbaric oxygen; its use is not recommended⁴,⁶,¹³,¹⁵
   - Superiority of fibrinogen-LDL-apheresis over standard first line treatments not established¹⁷

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.
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.

Prevention
3. Not well documented

Patient Information
1. Information for patients:

References


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