SUDDEN HEARING LOSS

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HEARING LOSS FACTS

- **Men** are more likely to experience hearing loss than women.
- Approximately 17 percent (**36 million**) of American adults report some form hearing loss.
- About 2 to 3 out of every 1,000 children in the United States are born deaf or hard-of-hearing.
  - Nine out of every 10 children who are born deaf are born to parents who can hear.

NIDCD
National Institute on Deafness and Other Communication Disorders
Research to improve the lives of people with communication disorders

HEARING LOSS FACTS

- The NIDCD estimates that ~15 percent (**26 million**) of Americans between the ages of 20 and 69 have **high frequency** hearing loss due to exposure to loud sounds or noise at work or in leisure activities.
- Only **1 out of 5** people who could benefit from a hearing aid actually wears one.
- Three out of 4 children experience ear infection (otitis media) by the time they are 3 years old.

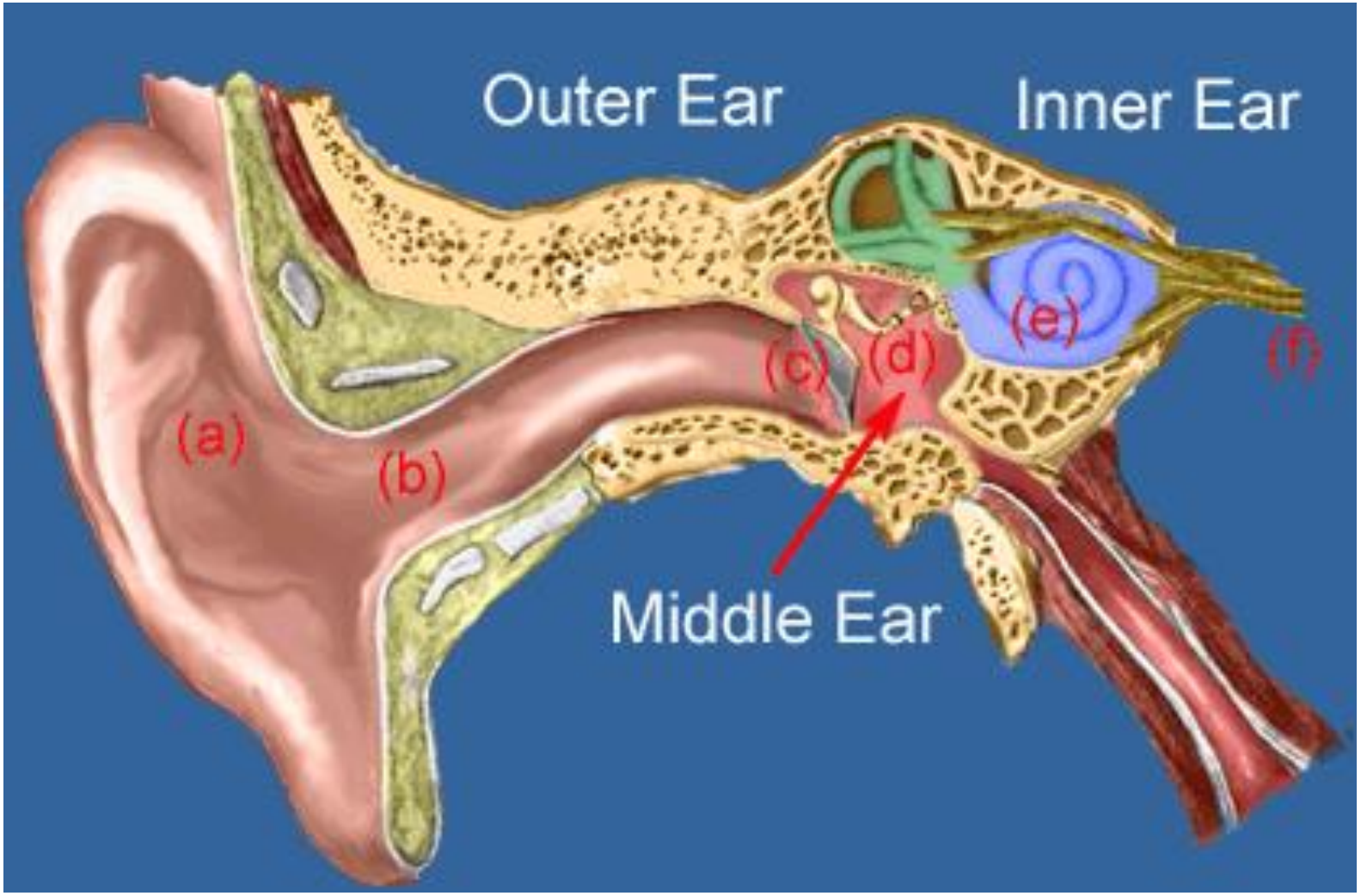
HEARING LOSS FACTS

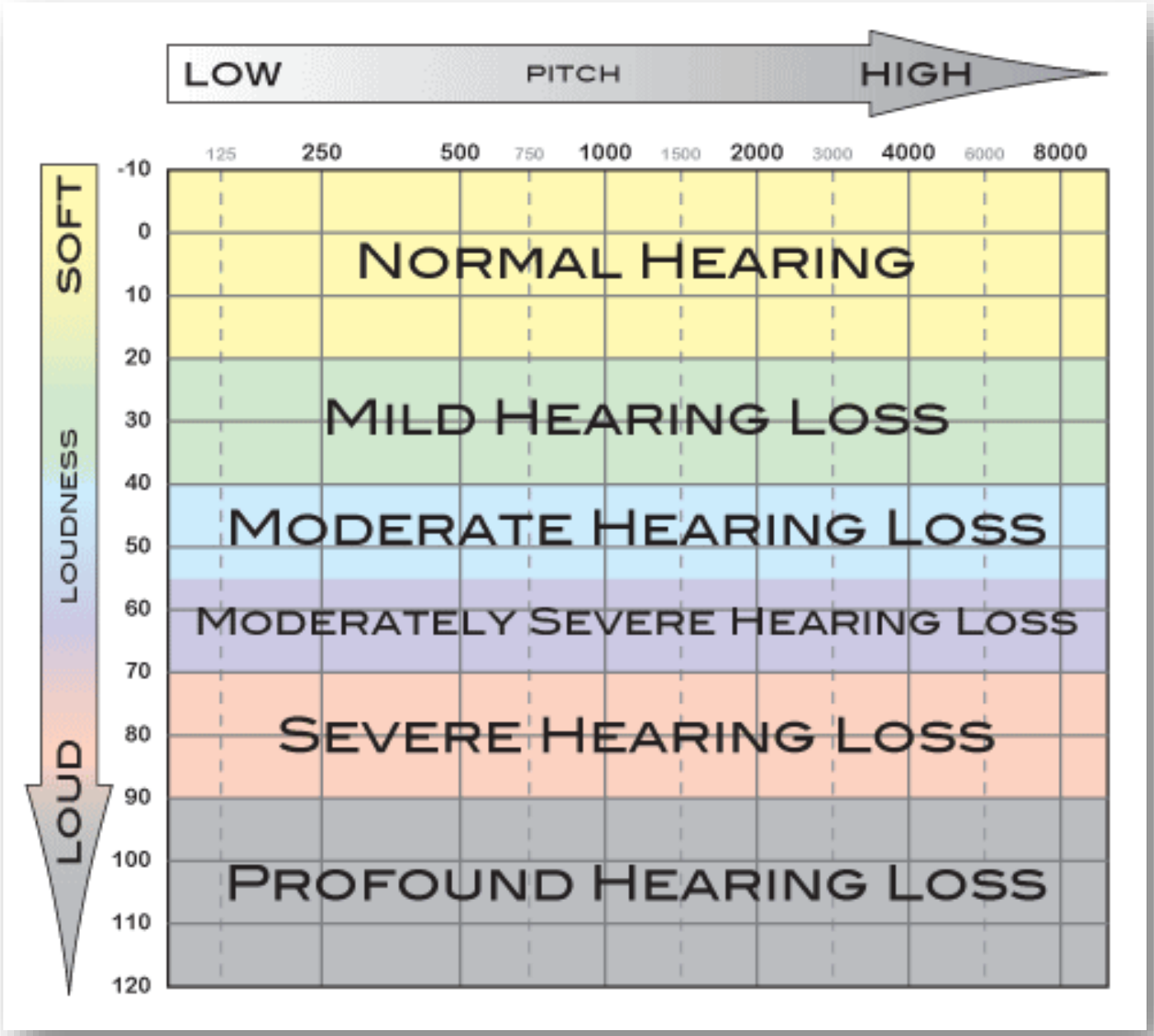
- Strong relationship between **age** and reported hearing loss:
  - 18 percent of American adults 45-64 years old, 30 percent of adults 65-74 years old, and 47 percent of adults 75 years old or older have a hearing impairment.
- Roughly **25 million** Americans have experienced tinnitus.
- Approximately 4,000 new cases of sudden deafness occur each year in the United States.

HEARING LOSS FACTS

- Approximately 615,000 individuals have been diagnosed with **Ménière's** disease in the United States. Another 45,500 are newly diagnosed each year.
- One out of every 100,000 individuals per year develops an acoustic neuroma (vestibular schwannoma).

HEARING ANATOMY & FUNCTION




LOW
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SOFT
LOUDNESS
LOUD
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30
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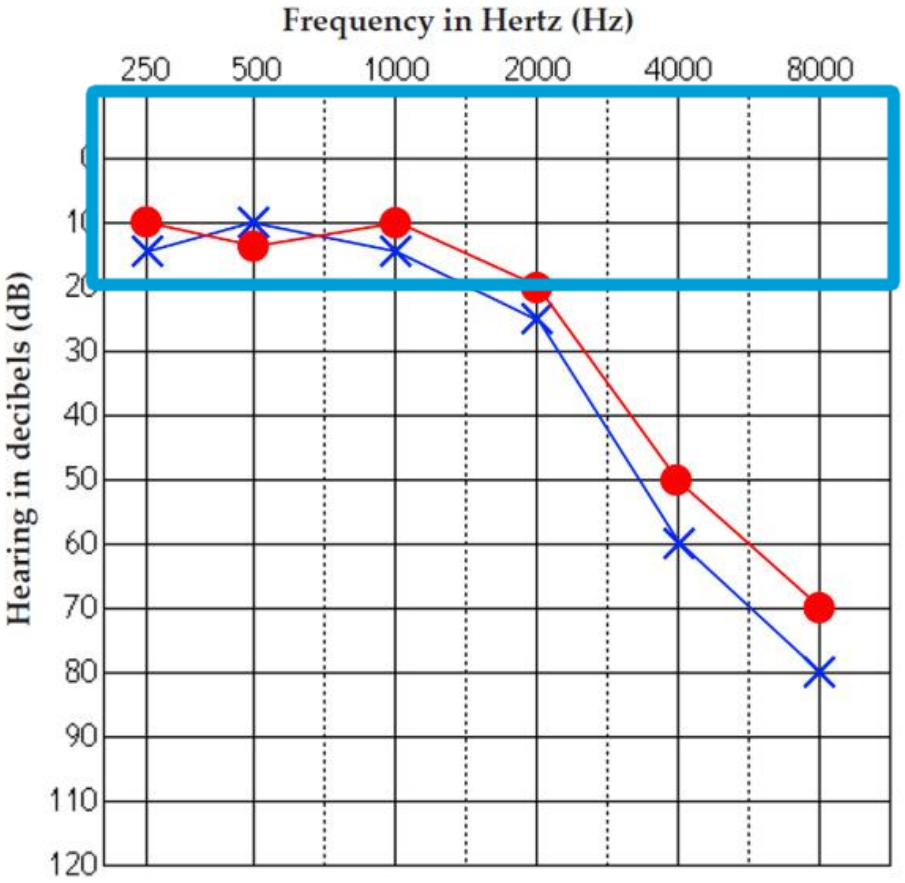
70
80
90
100
110
120
NORMAL HEARING
MILD HEARING LOSS
MODERATE HEARING LOSS
MODERATELY SEVERE HEARING LOSS
SEVERE HEARING LOSS
PROFOUND HEARING LOSS

AUDIOGRAM



Red Line = Right Ear

Blue Line = Left Ear

TYPES OF HEARING LOSS

1. **Sensorineural** Hearing Loss (nerve loss)
2. **Conductive** Hearing Loss
3. **Mixed** Hearing Loss (both nerve and conductive loss)

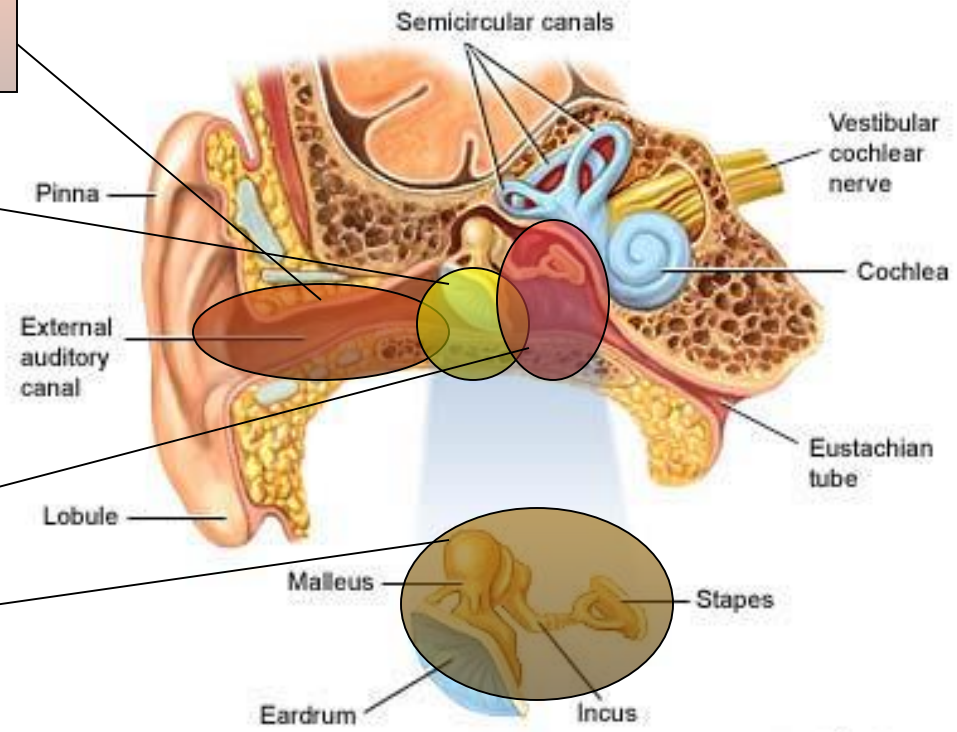
CONDUCTIVE HEARING LOSS

External Ear Canal
Cerumen impaction, foreign body

Tympanic Membrane
Perforation, tympanosclerosis, Hematoma

Middle Ear
Otitis Media with Effusion, Cholesteatoma

Ossicles
Otosclerosis



A.D.A.M.

SENSORINEURAL HEARING LOSS ETIOLOGIES

1. Infectious:
   - Meningitis, Herpes virus, HIV, Mumps, Rubella, Rubeola, Mycoplasma, Toxoplasmosis, Syphilis, Lyme disease
2. Autoimmune
   - Lupus erythematosus, Cogan's syndrome, Wegener's granulomatosis
3. Traumatic
   - Perilymph fistula, T-bone fracture, Acute blast injury

SNHL ETIOLOGIES

4. Vascular
   - Vertebrobasilar insufficiency (VBI), Sickle cell disease, Hyperviscosity syndromes, Waldenstrom's macroglobulinemia, Polycythemia vera, thrombocythemia
5. Neurologic
   - Multiple sclerosis, Migraine
6. Neoplastic
   - Acoustic neuroma, Meningioma, Metastasis, Leukemia, Myeloma

SNHL ETIOLOGIES

7. Iatrogenic
   - Ototoxic Meds, Otologic surgery
8. Congenital
   - Hereditary, Toxic, Infectious, Spontaneous
9. Toxic
   - Chronic noise
10. Idiopathic Sudden SNHL

SNHL: HISTORY

- Onset (sudden vs. progressive)
- Duration
- Fluctuations in hearing
- Associated symptoms:
  - tinnitus, **vertigo**, imbalance, aural fullness
- h/o otologic surgery or recurrent AOM, head trauma, vascular disease, autoimmune disease
- Family history of hearing loss

SNHL: PHYSICAL EXAM

- Full head and neck exam
- Otoscopic exam
  - Pneumatic otoscopy / Tympanometry
- Tuning forks (Weber & Rinne)
- Cranial nerve exam
- Cerebellar exam/Balance testing as appropriate

SNHL: PHYSICAL EXAM

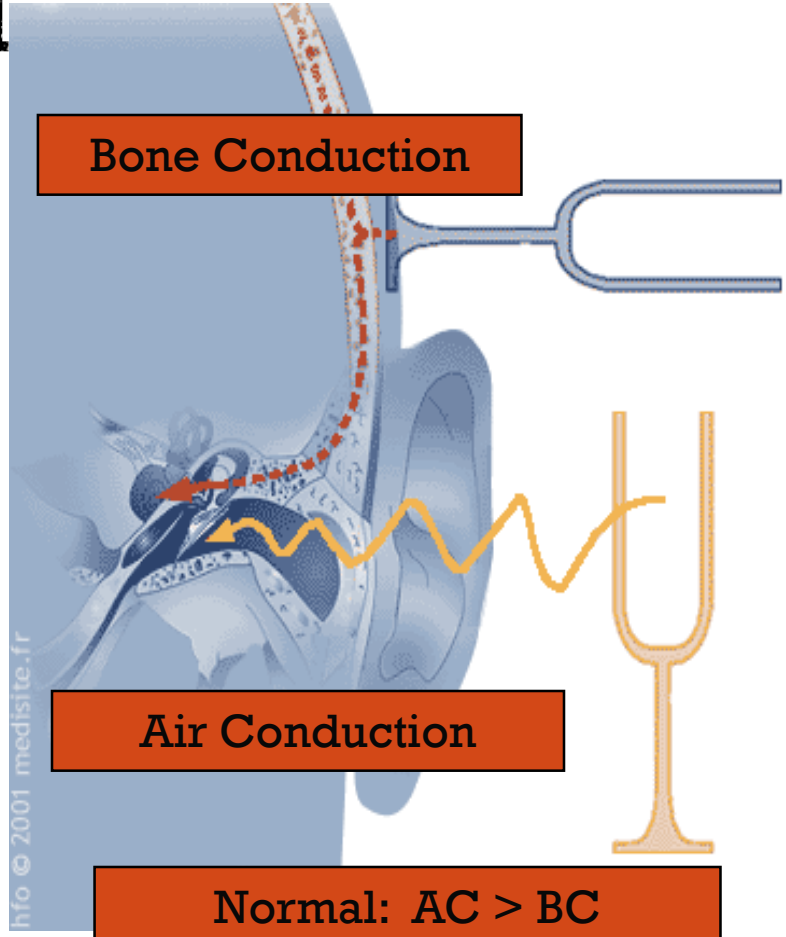
WEBER TEST: typically with a 512 Hz tuning fork

- Normal = sound heard centrally or in both ears
- unilateral SNHL should lateralize to *better* hearing ear,
- unilateral CHL should lateralize to *diseased* ear

SNHL: PHYSICAL EXAM

Rinne Test: compare air conduction (AC) and bone conduction (BC); place tuning fork ≤1 cm of the EAC (AC)and then place on the mastoid (BC);

- Positive = SNHL (GOOD)
  - tuning fork heard better by AC = normal or most SNHL
- Negative – CHL
  - BC is perceived louder than AC = CHL >15–30 dB HL or severe to profound SNHL with cross-over



WEBER & RINNE TEST REVIEW

Table 4. Recommended Technique for Weber and Rinne Testing

Weber Test	Rinne Test
1. Place vibrating tuning fork (256 or 512 Hz) at midline of forehead or on maxillary teeth (not false teeth) 2. Ask where the sound is heard; it is normal to hear at the midline or "everywhere" 3. If the sound lateralizes to one ear then: a. There is a CHL in that ear, OR b. There is SNHL in the opposite ear	1. Place vibrating tuning fork (256 or 512 Hz) over the mastoid bone of one ear, then move the tuning fork to the entrance of the ear canal (not touching the ear) 2. The sound should be heard better via air conduction (at the entrance to the ear canal). 3. If the sound is heard better by bone conduction, then there is a CHL in that ear. Repeat for the other ear.

Abbreviations: CHL, conductive hearing loss; SNHL, sensorineural hearing loss.

SNHL: WORKUP

- Full audiogram with pure tones, speech recognition, and word recognition
- For sudden sensorineural hearing loss or asymmetric sensorineural hearing loss: MRI + gadolinium

PRESBYACUSIS

- Age-related hearing loss
- 40% U.S. population >75 y/o affected
- Often familial ($\geq$50%)
- Bilateral and symmetric

PRESBYACUSIS: TREATMENT

- Treatment
  - Hearing aids
  - Assistive listening devices
  - Cochlear Implantation

NOISE INDUCED HEARING LOSS (NIHL)

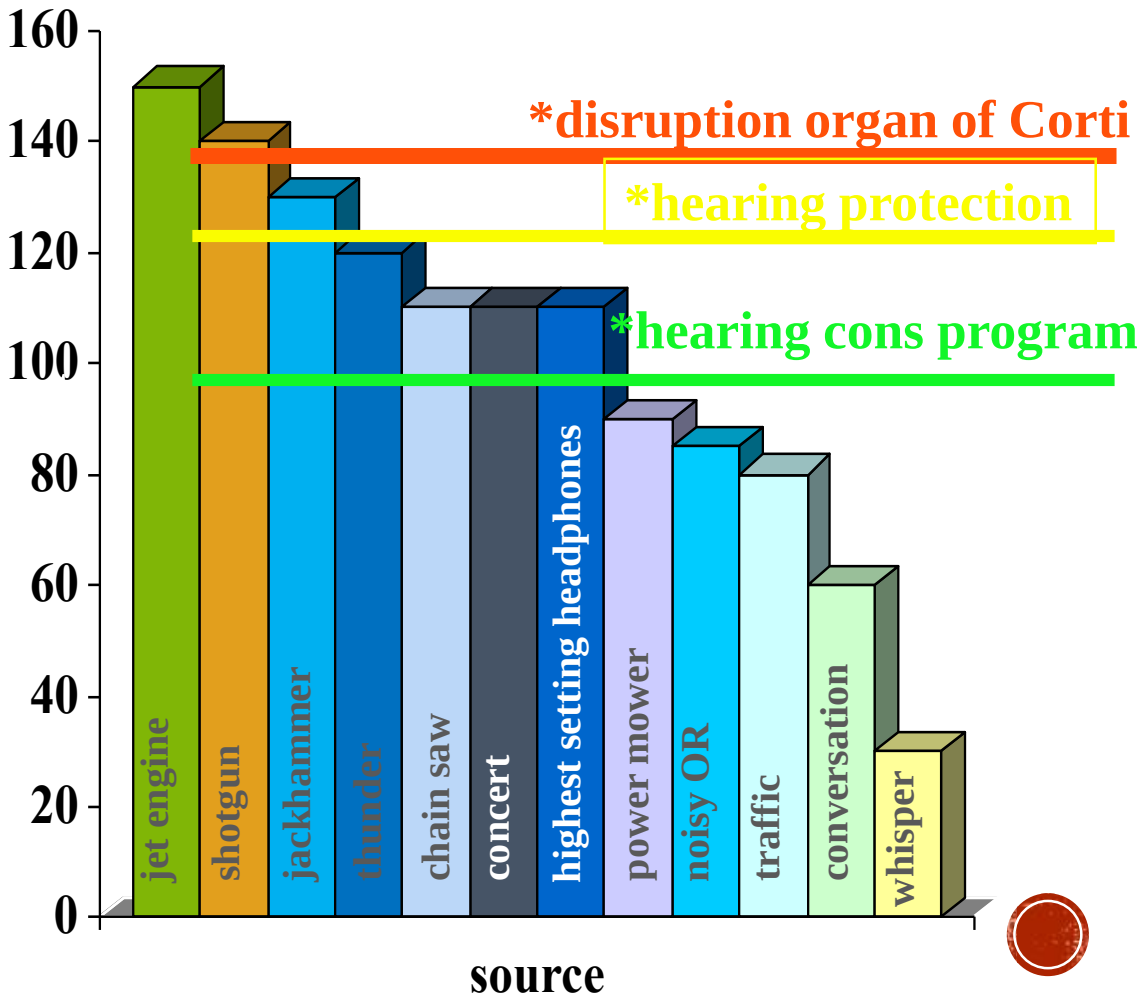
- Most *common* cause of *preventable* SNHL
- Most frequently occurs from exposure through years (> 90dB)
- Can result from single exposure to very loud noise (>120-130 dB)
- Typically bilateral and symmetric

NIHL: BACKGROUND

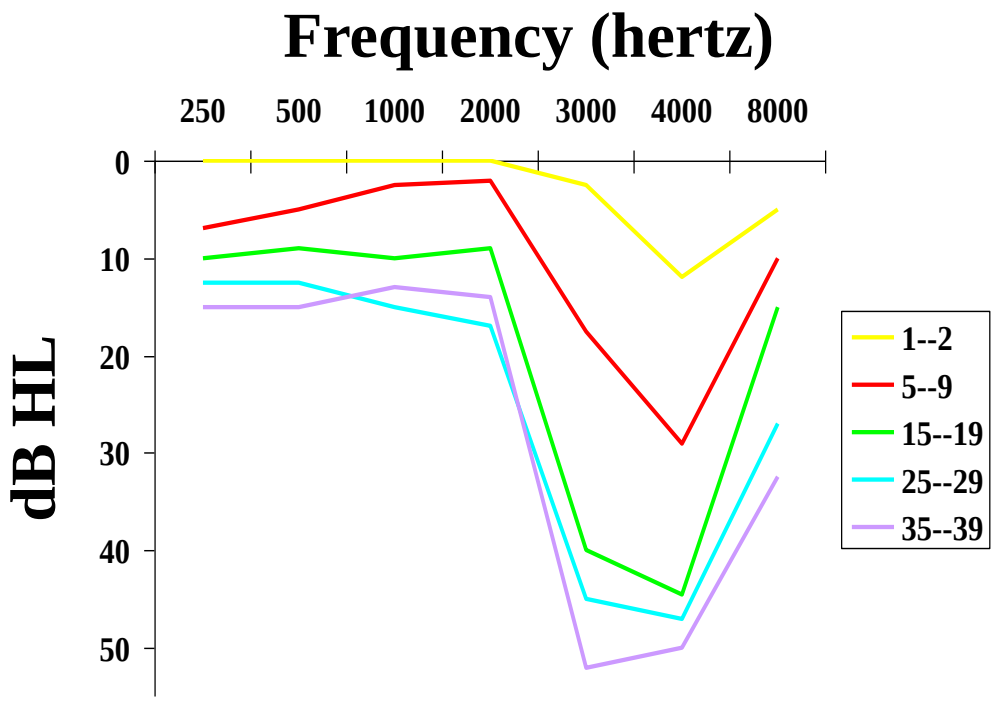
3 types noise:
- Continuous
- Impulse (eg., gun)
- Impact

2 types exposure:
- Chronic
- Acute

2 types damage:
- TTS (temporary)
- PTS (permanent)



CHRONIC NIHL: AUDIOGRAM



Years Exposure

OTOTOXIC MEDICATIONS

- **Macrolides**
  - High frequency SNHL, tinnitus, vertigo
  - Usually reversible within 2 weeks
  - Unknown mechanism
- **Vancomycin**
  - High freq SNHL progresses to bilateral profound SNHL
- **ASA**
  - Doses > 2700 mg/day
  - Affects stria vascularis, reversible

OTOTOXIC MEDICATIONS

- Antineoplastics/cisplatinum
  - Begins with high freq HL, progresses as total dose accumulates
  - Irreversible when profound deafness occurs
  - Can be vestibulotoxic
  - Affects OHC
- Loop diuretics/ethacrynic acid
  - Affects stria vascularis, rarely permanent
- Phosphodiesterase type 5 inhibitors (Viagra > Levitra, Cialis)
  - Unknown mechanism; question of nitric oxide effects on ear

PERILYMPHATIC FISTULA (PLF)

- Definition: Communication between perilymph space and middle ear/mastoid
- Etiology
  - Increased pressure/trauma→communication→Decreased perilymph volume→2^{ndary} endolymphatic hydrops→symptoms
- Potential causes (rare):
  - Otologic surgery (stapedectomy)
  - **Head trauma**
  - **SCUBA** diving
  - Congenital ear malformation
  - **Forced valsalva** / suppressed sneezing

NEOPLASIA

- **Acoustic tumors**:
  - Most common: **Acoustic Neuroma** (misnomer) = Vestibular Schwanomma
  - Usually present with gradually progressive SNHL
  - 1% of patients with asymmetric SNHL have acoustic tumors

AAO-HNS GUIDELINES – 2012

AMERICAN ACADEMY OF OTOLARYNGOLOGY–HEAD AND NECK SURGERY FOUNDATION

Guideline

Otolaryngology–Head and Neck Surgery
146(1S) S1–S35

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http://otojournal.org
SAGE

Clinical Practice Guideline: Sudden Hearing Loss

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Abstract

Objective. Sudden hearing loss (SHL) is a frightening symptom that often prompts an urgent or emergent visit to a physician. This guideline provides evidence-based recommendations for the diagnosis, management, and follow-up of patients who present

of medical interventions, and the limitations of existing evidence regarding efficacy; and (3) counsel patients with incomplete recovery of hearing about the possible benefits of amplification and hearing-assistive technology and other supportive measures. The panel made *recommendations* that clinicians should (1) assess patients with presumptive SSNHL for bilateral SHL, recurrent episodes of SHL, or focal neurologic findings; (2) diagnose presumptive ISSNHL if audiometry confirms a 30-dB hearing loss at 3 consecutive frequencies *and* an underlying condition cannot

CLINICAL PRACTICE GUIDELINE SUMMARY

Table 3. Summary of Evidence-Based Statements

Management of Patients with Sudden Hearing Loss (Evidence-Based Statement)	Statement Strength
Diagnosis	
Exclusion of conductive hearing loss (Statement 1)	Strong recommendation
Modifying factors (Statement 2)	Recommendation
Computed tomography (Statement 3)	Strong recommendation against
Audiometric confirmation of idiopathic sudden sensorineural hearing loss (Statement 4)	Recommendation
Laboratory testing (Statement 5)	Strong recommendation against
Retrocochlear pathology (Statement 6)	Recommendation
Shared decision making	
Patient education (Statement 7)	Strong recommendation
Treatment	
Initial corticosteroids (Statement 8)	Option
Hyperbaric oxygen therapy (Statement 9)	Option
Other pharmacologic therapy (Statement 10)	Recommendation against
Salvage therapy (Statement 11)	Recommendation
Follow-up	
Outcomes assessment (Statement 12)	Recommendation
Rehabilitation (Statement 13)	Strong recommendation

Robert J. Stachler et al. Otolaryngology -- Head and Neck Surgery 2012;146:S1-S35

Otolaryngology–Head and Neck Surgery

IDIOPATHIC SUDDEN SENSORINEURAL HEARING LOSS (ISSNHL)

- Theories:
  - Viral
  - Autoimmune (autoimmune inner ear disease – AIED)
  - Vascular
  - Intracochlear membrane breaks

ISSNHL: VIRAL

- Current belief – viral cochleitis causes the majority of cases of ISSNHL
- 1983 – Wilson and colleagues
  - Viral seroconversion rates greater in patients with ISSNHL (63%) compared to control (40%)
    - Influenza B
    - Mumps
    - Rubeola
    - VZV

ISSNHL: VIRAL

- 1981- Veltri *et al*.
  - 65% seroconversion
- 1986 – Schuknecht and Donovan
  - Temporal bone studies (n. 12)
    - ISSNHL vs. cases of known viral labyrinthitis
    - Similar pathologic findings
      - Atrophy of the organ of Corti, tectorial membrane, stria vascularis, cochlear nerve, and vestibular organ

ISSNHL: VIRAL

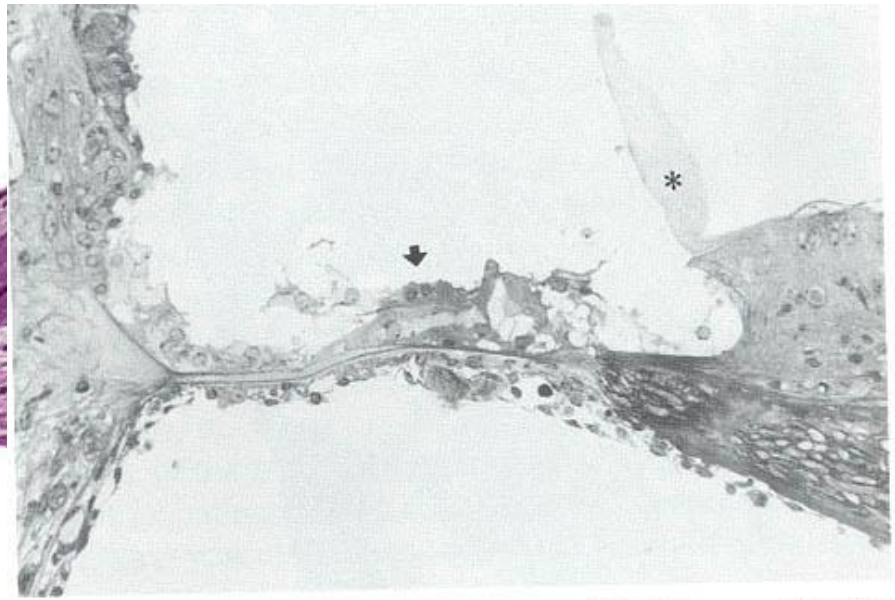

FIG. 2. Light microscopy of the organ of Corti in experimental herpes simplex virus type 1 labyrinthitis. Extensive destruction of the organ of Corti and supporting cells. The tectorial membrane is disrupted from the sensory cells but has a normal appearance (arrow) (bar = 100 μm).

ISSNHL

- New - **Sleep apnea** linked to sudden hearing loss
  - No causility proven
  - Sleep apnea causes major inflammation in the bloodstream/brain promoting vascular complications
  - Need for prospective studies, and causality studies with treatment trials (i.e., improved sleep apnea, improved hearing).

Arch Otol Head Neck Surg. 2012; 138 [1]:55.

ISSNHL: TREATMENT

- 90% of cases will be Idiopathic
- Treat known causes by addressing the underlying condition

ISSHNL: TREATMENT

- Therapy for ISSNHL is **controversial**
- Difficult to study
  - High spontaneous recovery rate
  - Low incidence
  - Makes validation of empiric treatment modalities difficult

ISSNHL: TREATMENT

- Proposed treatment modalities
  - Anti-inflammatory – steroids, cytotoxic agents
    - e.g., **Prednisone** 1mg/kg/day (80mg) PO QD taper over 2 weeks.
  - Diuretics
  - Antiviral agents

General Guidelines for Corticosteroid Therapy for Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL)

Table 9. General Guidelines for Corticosteroid Therapy for Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL)[a]

	Oral Corticosteroids	Intratympanic Corticosteroids
Timing of treatment	Immediate, ideally within first 14 days. Benefit has been reported up to 6 weeks following onset of sudden sensorineural hearing loss (SSNHL)	Immediate Salvage (rescue) after systemic treatment fails
Dose	Prednisone 1 mg/kg/d (usual maximal dose is 60 mg/d) or Methylprednisolone 48 mg/d or Dexamethasone 10 mg/d	Dexamethasone 24 mg/mL or 16 mg/mL (compounded), or 10 mg/mL (stock)Methylprednisolone 40 mg/mL or 30 mg/mL
Duration/frequency	Full dose for 7 to 14 days, then taper over similar time period	Inject 0.4 to 0.8 mL into middle ear space every 3 to 7 days for a total of 3 to 4 sessions
Technique	Do not divide doses	Anterosuperior myringotomy after topical anesthetic Inject solution into the posterior inferior quadrant via narrow-gauge spinal needle to fill middle ear space Keep head in otologic position (one side down, affected ear up) for 15 to 30 minutes
Monitoring	Audiogram at completion of treatment course and at delayed intervals	Audiogram before each subsequent injection, at completion of treatment course, and at delayed intervals Inspect tympanic membrane (TM) to ensure healing at completion of treatment course and at a delayed interval
Modifications	Medically treat significant adverse drug reactions, such as insomnia Monitor for hyperglycemia, hypertension in susceptible patients	May insert pressure-equalizing tube if planning multiple injections, but this increases risk of TM perforation May consider adding round window transport facilitator

[a]This table is designed to provide guidance for systemic and intratympanic steroid treatment for SSNHL. Treatment is routinely individualized by provider and per patient. The most important principles pertain to early institution of high enough dosages of treatment. Prednisone 1 mg/kg/d or its equivalent and/or adequate concentration of intratympanic dexamethasone or solumedrol should be administered.

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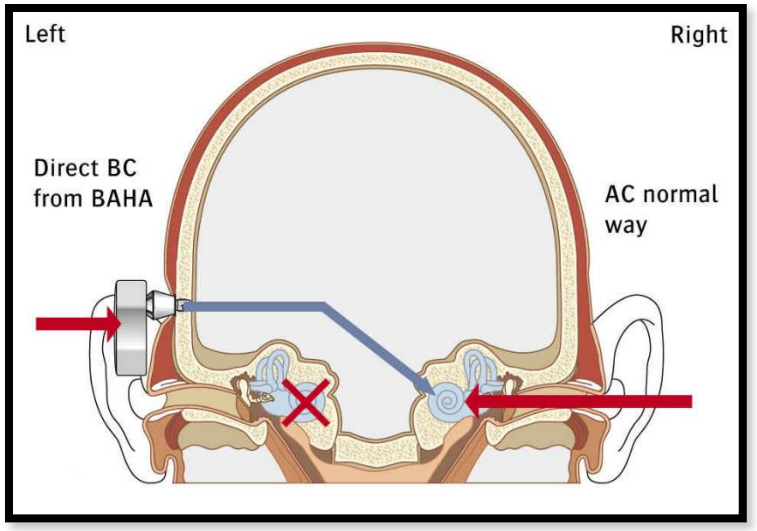
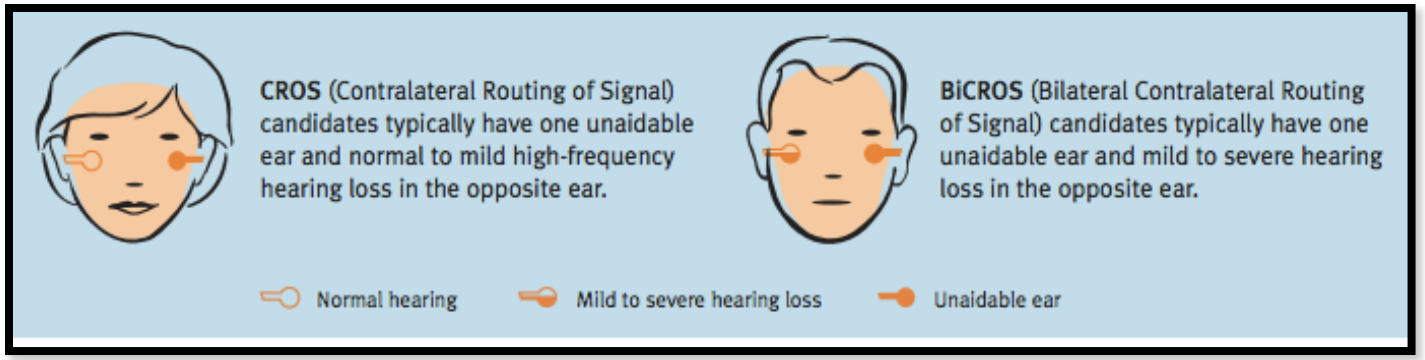
TREATMENT

- BAHA (Cochlear / Oticon)
  - Bone Anchored Hearing "Aid"
  - Surgically implanted pin in the skull, using a vibrating digital hearing amplfication device.
- CROS/BICROS
  - Contralateral Routing Of Signal





Selected Conditions That May Be Associated with Bilateral Sudden Hearing Loss.

Table 6. Selected Conditions That May Be Associated with Bilateral Sudden Hearing Loss

Cause	Other Features
Meningitis (infectious, inflammatory, neoplastic)	Headache, fever, abnormal cerebrospinal fluid (CSF) studies, possibly other cranial nerve palsies[211]
Autoimmune inner ear disease	Fluctuation of hearing may sometimes occur; vertigo may occur in some cases.[41]
Lyme disease	Erythema chronicum migrans, abnormal CSF, fluctuating bilateral audiovestibular symptoms[212]
Syphilis	Abnormal fluorescent treponemal antibody absorption (FTA-abs) test, bilateral fluctuating hearing loss, tabes dorsalis, multiorgan involvement [213]
Ototoxic medications	Vestibular loss, oscillopsia[214,215]
Trauma	Significant head trauma, barotrauma, temporal bone fractures[214]
Herpes zoster oticus (Ramsay-Hunt syndrome)	Otalgia, pinna and/or ear canal vesicles, facial nerve paresis, positive viral titers, positive viral cultures[216]
Human immunodeficiency virus (HIV) otitis	Positive HIV titers, altered T cell counts, and often other cranial neuropathies may be associated with mastoiditis out of proportion to clinical complaints.[217,218]
Lead poisoning	Learning disabilities, other stigmata of lead poisoning[219]
Genetic disorders	May be syndromic or nonsyndromic[220,221]
MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes)	Periods of confusion, elevated serum lactic acid levels around times of attacks, stroke-like spells, magnetic resonance imaging (MRI) white matter signal changes, migraine-like headaches, seizures, diabetes, mitochondrial gene mutation (Mt-RNR1, Mt-TS1, POLG genes)[222,223]
Other mitochondrial disorders	Variable phenotypes[224]
Bilateral synchronous internal auditory artery occlusion associated with vertebrobasilar vascular disease	Vertigo, dysarthria, facial weakness, ataxia, nystagmus, unilateral numbness, abnormal computed tomography or magnetic resonance angiogram of the vertebrobasilar vasculature[48,50,225-227]
Cogan syndrome	Nonsyphilitic interstitial keratitis of the cornea, hearing loss, vertigo[40]
Neoplastic (neurofibromatosis II, bilateral vestibular schwannomas, intravascular lymphomatosis, others)	Abnormal brain MRI or cerebrovascular imaging study[228-230]
Sarcoidosis	Pulmonary symptoms, bilateral vestibular loss, elevated serum angiotensin-converting enzyme level or abnormal Gallium scan[231,232]
Hyperviscosity syndrome	Mucous membrane bleeding, neurologic and pulmonary symptoms, associated retinopathy[233]

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AUTOIMMUNE INNER EAR DISEASE (AIED)

- 1979 – McCabe
  - Described patients with bilateral rapidly-progressive SNHL (BRPSNHL)
  - Proposed the term – **autoimmune inner ear disease** (AIED)
  - Evidence of **autoimmunity**
    - Lymphocyte inhibition test
    - Substantial hearing improvement with steroids

AIED

- Clinical characteristics
  - Middle-aged **females**
  - **BRPSNHL**
  - Absence of systemic immune disease
  - 50% with **dizziness**
  - Light-headedness and ataxia more common than vertigo
  - Episodes – multiple, daily
  - Hearing loss sudden, rapidly progressive, or protracted

AIED: EXAMPLES

"RUSH LIMBAUGH'S severe-to-profound, bilateral, rapidly progressive hearing loss generated considerable public interest in sudden deafness. In his case, its cause was reportedly an autoimmune disease of the cochlea."

- CNN.com

"FOXY BROWN, real name Inga Marchand, has revealed that she is slowly losing her hearing . She first noticed a problem when her label boss, Jay-Z told her the sound levels on her new record were way too high when she had thought they were perfect."

- Hip Hop News

AIED

- Diagnosis
  - Based on Hearing loss and response to treatment
  - Hughes –
    - Lymphocyte transformation test
      - Sensitivity – 50-80%
      - Specificity – 93%
      - Positive predictive value 56-73%
    - Western blot
      - Sensitivity – 88%
      - Specificity – 80%
      - Positive predictive value – 92%

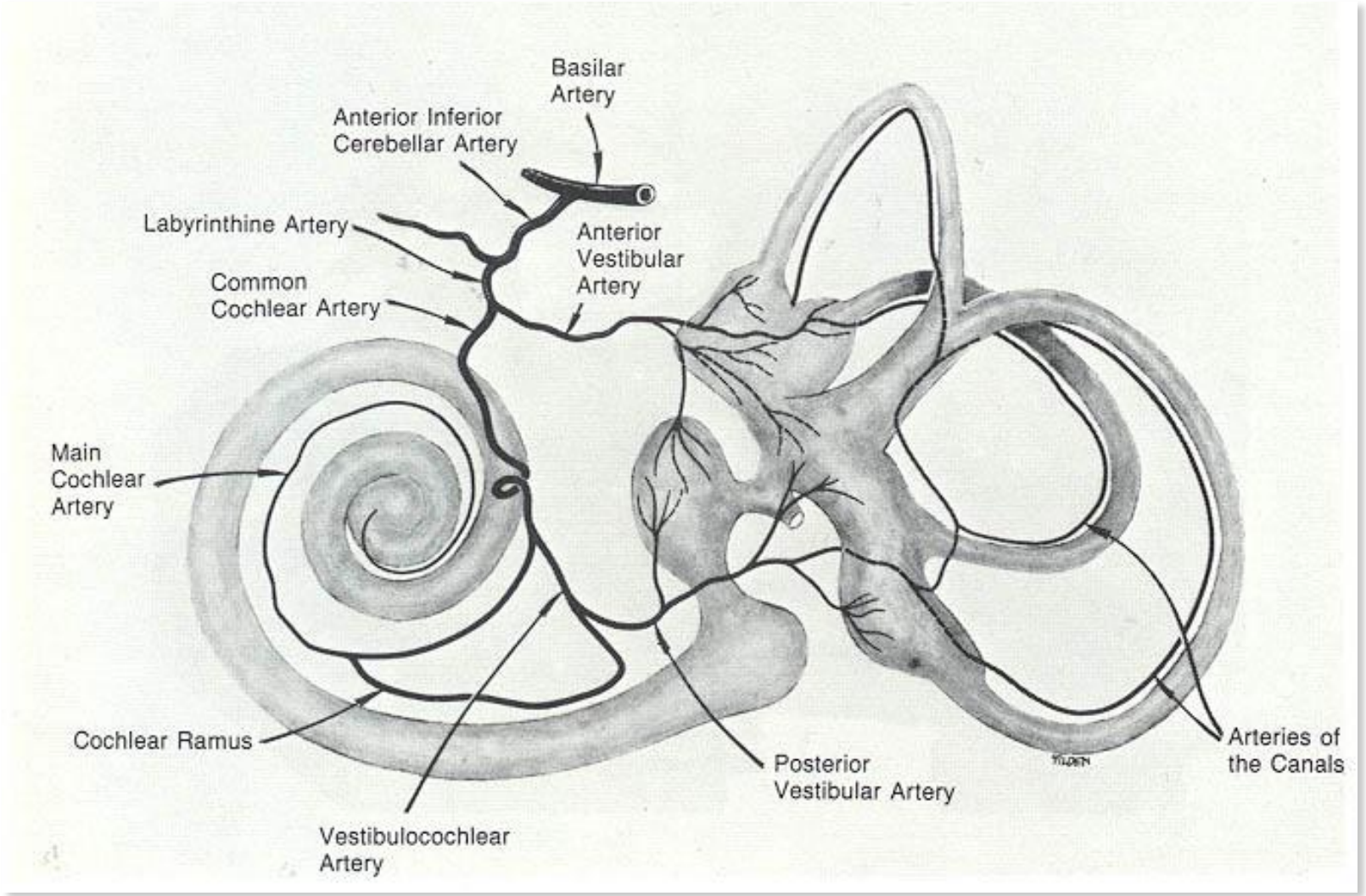
AIED TREATMENT

1. **Prednisone** 1mg/Kg/day for 4 weeks
2. Slow taper
3. Relapse during taper – restart
4. Slow taper
5. If relapse during taper – **Cytotoxic** agent
   - Methotrexate
   - Cyclophosphamide
   - Monitor electrolytes, LFTs, blood counts

VASCULAR

- Etiologies - Embolism, vasospasm, hypercoagulable states/sludging
- Pathophysiology – anoxia to vestibulocochlear apparatus
- Cochlea is intolerant to disruption of blood supply
  - 1957 Kimura and Perlman
    - Clamped the labyrinthine artery in guinea pigs
    - Demonstrated irreversible loss of cochlear function after 30 minutes of disruption

VASCULAR ANATOMY



VASCULAR

- Abnormal **circulatory** states
  - Sickle-cell disease
  - Waldenstrom's macroglobulinemia
  - Hearing loss is usually reversible with tx
  - AICA strokes
  - Cardiopulmonary bypass

PROGNOSIS

- 47%-63% spontaneously resolve
  - Combined patients with all audiogram types
- **Prognostic** variables:
  1. Time since onset
  2. Audiogram type (severity of hearing loss)
  3. Vertigo
  4. Age

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