Hearing Disorders

Prevalence Estimates

<table>
<thead>
<tr>
<th>Percentage</th>
<th>Number (millions)</th>
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<tbody>
<tr>
<td>6.6%</td>
<td>19.8</td>
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<tr>
<td>8%</td>
<td>24</td>
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<tr>
<td>10%</td>
<td>30</td>
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"hearing impaired"  
PTA > 25 dB HL  
some hearing impairment

Prevalence by Age and Gender

Hearing Disorders in Adults

- Permanent (Sensorineural)
  - Presbycusis
  - Noise Induced Hearing Loss
  - Meniere's disease
  - Otoxic diseases & drugs
  - Other

- Fixable (Conductive)
  - Otosclerosis
  - Other

Presbycusis

- The progressive loss of hearing that occurs with age.  
  AMA Encyclopedia of Medicine
- Prevalence
  - Conservative estimate ≈ 25% of those >65 have a handicapping hearing loss

Hearing and Ageing

(Cruickshanks et al., 1998)
Classic Categories
(Schuknecht)

- Sensory
- Neural
- Metabolic
- Mechanical

Sensory Presbycusis

Sensorineural Presbycusis

Noise Induced Hearing Loss

- Permanent, sensorineural loss from chronic exposure to high-intensity sound
  – (> 80 dB A)
- Incidence
  – #1 occupational hazard
  – Estimated that at least 16 million in U.S. suffer from some form of NIHL
  – Majority of impairments in middle age

Meniere’s Disease

- Disease characterized by episodes of:
  – Vertigo
    - Nausea
    - Vomiting
  – Tinnitus
    - Roaring or buzzing
  – Hearing loss
  – Fullness
Meniere's Disease

- Incidence
  - #3 cause of sensorineural hearing loss in adults
  - Low end = 46:100,000 (Stahle et al., 1978)
  - High end = 160:100,000 (Cawthorne & Hewlett, 1954)

- Prevalence = incidence X 25

Meniere's Disease Sx

- Hearing loss - Configuration
  - Early stages
    - Classic = low frequency loss (rising)
    - Occasionally flat
    - Rarely high-frequency (sloping)
  - Later
    - Flattens out as loss progresses

Hearing Loss Configuration

Endolymphatic Hydrops

Diseases Causing Hearing Loss

- More Common
  - Bacterial meningitis
  - Sudden onset
- Less Common
  - Mumps
  - Measles

- Rare
  - Diphtheria
  - Whooping cough
  - Typhoid
  - Scarlet fever
  - Chickenpox
  - Flu
  - Cold viruses
  - Polio
  - Herpes virus
  - Other

Major Ototoxic Drugs

- Antibiotics
  - Streptomycin
  - Neomycin
  - Kanamycin
  - Gentamicin

- Chemotherapeutics
  - Cisplatin
  - Carboplatin

- “Loop” Diuretics
  - Furosemide (Lasix)
  - Bumetamide (bumex)
  - Ethacrinic acid (Edecrin)

- Aspirin
Otosclerosis

Disease of the bone of the otic capsule.

Clinical Forms:
- Fixation of the stapes (most common)
- Cochlear impairment + stapes fixation
- Pure cochlear (labyrinthine, cochlear or retrofenestral)

Stapes Fixation

Otosclerosis

Prevalence
- Youngsters ≤ 17
  ~ 1% of have SRTs ≥ 26 dB HL
- Newborns
  ~1 per 1,000 live births
"ABCDEFG's of Deafness"
(JCNH, 1981)

- A - Asphyxia
- B - Bacterial meningitis
- C - Congenital/perinatal infections
- D - Defects of the head or neck (e.g., cleft palate, pinna abnormalities)
- E - Elevated bilirubin
- F - Family history of childhood hearing impairment
- G - Gram birthweight less than 1,500 gms.

Congenital Hearing Impairment
Inherited
(Endogenous)

Incidence
- In general population = 1/2000 - 1/6000 births
- Among congenitally deaf ≈ 50% hereditary
- Pattern of inheritance
  - About 75 - 80% = recessive
  - About 20 - 25% = dominant
  - Rest = too rare to worry about

Recessive Inheritance
- Defective gene must be present at the same locus on both chromosomes in the pair for defect to be present in offspring
  - 50% = carriers
  - 25% = affected offspring
  - 25% = completely normal
Dominant inheritance

- Abnormality may be expressed when defective gene is in only one of pair of chromosomes
  - 50% = affected offspring
  - 50% = completely normal
  - No carriers

Nonsyndromic Hearing Loss Genes

- Nomenclature:
  - Loci of hearing loss genes are numbered consecutively to reflect order of discovery.
  - Mode of inheritance is denoted by prefixes:
    - DFNA (dominant)
    - DFNB (recessive)
    - DFN (X-linked)

Major Recessive Syndromes

- 50% of recessive losses are syndromic
- Pendred's Syndrome (1896)
  - Congenital hearing loss
  - Thyroid dysfunction (goiter) in adolescence
- Usher's Syndrome (1914)
  - Cochlear loss (congenital or degenerative).
  - Degeneration of inner layer of retina (retinitis pigmentosa)

Major Dominant Syndromes

- Waardenberg's Syndrome (1951)
  - 20% = unilateral or bilateral hearing loss
  - 99% = lateral displacement of medial canthi (wide set eyes)
  - 78% = flat bridge of nose
  - 25% = iris heterochromia
  - 17% = white forelock
- Branchiootorenal Syndrome
  - Renal dysfunction
  - Variable hearing loss
    - Conductive, sensorineural, mixed
    - Mild - profound

Recessive

- Estimate: at least 25 different genes involved
- DFNB1 accounts for about 50%
- DNA testing can identify deafness-causing mutations of this gene in most cases

Dominant

- Estimate: at least 22 different genes involved
For your own info:

- 90% of congenitally hearing impaired have 2 normally hearing parents.
- 30% of offspring from 2 deaf parents have hearing losses also.
- More precise predictions can only be made by expert.

Congenital Hearing Impairment

Acquired (Exogenous)

Congenital or Perinatal Infections

The TORCH Complex

- T = toxoplasmosis
- O = other (syphilis)
- R = rubella
- C = CMV
- H = Herpes

Toxoplasmosis

- Toxoplasma gondii parasite
- Transmitted across placenta
- Classic Sx Triad
  - Chorioretinitis
  - Hydrocephalus
  - Intracranial calcifications
- One estimate = 17% of infected infants will develop hearing loss

Congenital Rubella: Sx Triad

- bilateral hearing loss
- cataracts (40%)
- heart anomalies (50%)
Rubella Incidence

Herpes Group of Viruses

- Cytomegalovirus
- Herpes simplex Type I
- Herpes simplex Type II
- Epstein-Barr
- Varicella-Zoster

Cytomegalovirus (CMV)

- Incidence
  - Most common known microbiological cause of brain damage in infancy.
  - > 7000 children born each year with hearing loss from CMV
- Sensorineural Hearing Loss
  - Pattern is very variable
  - Most are bilateral, severe/profound
  - Majority show progressive loss.

High Multiple Handicap Rate:

- ~50% have one or more educationally significant disabilities in addition to h/loss.
- 22% have two additional disabilities:
  - 19% MR
  - 13% CP
  - 10% Orthopedic problems
  - 10% LD
  - 9% Emotional/behavior problems
  - 6% blindness or significant visual impairment
- Score lower on standardized tests of academic achievement than h/l peers

Herpes Simplex Type II

- Becoming one of the most common sexually transmitted diseases
  - 20-25% of the population
- Disease Process
  - 82% of neonatal infections are generalized throughout the body.
  - High mortality
  - Only 4% of infected infants survive without being affected.

Hypoxia/Anoxia

- Hypoxia - Amount of oxygen in air, arterial blood, or body tissues is below normal, but short of anoxia.
- Anoxia - Absence or almost complete absence of oxygen in air, arterial blood, or body tissues.
- Asphyxia
  - Impaired or absent exchange of O₂ and CO₂ in breathing.
  - Results in a lack of O₂ (anoxia) and increased CO₂ (hypercapnia) in the blood and tissues.
- Anemia - Deficiency of oxygen-transporting material (RBC's, hemoglobin) in the blood.
- Ischemia - Localized shortage of blood due to obstruction of the blood supply.
Asphyxia

Apgar score
A: Activity (muscle tone)
P: Pulse
G: Grimace (reflex irritability)
A: Appearance (skin color)
R: Respiration

- Two points in each area
- < 7 = "at risk"
- 3 or lower = "high risk"
- 1 min = asphyxia and need for ventilation
- 5 min = neurological impairment or death

Hearing Loss

- About 4% with severe perinatal asphyxia develop sensorineural loss
- May damage CANS
- Possible cause of auditory neuropathy?
  - Normal OAEs
  - No ABR

Elevated Bilirubin (hyperbilirubinemia; jaundice)

- Excessive amount of bilirubin in the blood.
- Any factor that causes:
  - excessive breakdown of red blood cells
  - abnormal metabolism of bilirubin by the liver
- Most Common Cause = Rh Incompatibility
  - Erythroblastosis fetalis
  - Antibodies from Rh- mother attack Rh+ protein in RBC's of child
  - Causes immature RBC's (erythroblasts) anemia
    hyperbilirubinemia

Treatment

- Phototherapy - light converts bilirubin to a water soluble form that can be excreted by the kidneys.
- Exchange blood transfusion if phototherapy fails

Kernicterus

- Neurological syndrome associated with bilirubin deposits in the CNS.
- Hearing loss:
  - Bilateral, sensorineural, high frequency
  - Possible auditory neuropathy?
- Highest multiple handicap rate of all congenital etiologies (71.1%)
- Most brain damage of any etiology

Gram Birthweight < 1,500

- Methods of estimating gestational age are unreliable.
- Consequently, prematurity is now defined in terms of birthweight:
  - Low BW < 2000 gms. (4.4 lbs)
  - Very low BW < 1500 gms. (3.3 lbs)
- Hearing loss probably associated with hypoxia/anoxia
Very Low Birthweight

- Highest MI rate of all congenital etiologies
  - 16%
  - 2.2% rate in general population
- 14.44% die as neonates
  - More are surviving as result of improved medical care

Outer Ear Malformations

- Microtia
  - Auricle
  - Grades
    - I = small, but well formed
    - II = malformed
    - III = remnant

- Atresia
  - EAC
  - Grades
    - I = lesion to EAC alone
    - II = EAC lesion, bony TM, and malformed middle ear
    - III = EAC, TM and middle ear are absent

Grade II Microtia + Atresia

Outer Ear Malformations

- Associated anomalies:
  - 12-47% have cochlear pathology
  - Tortuous VII N.

Treacher-Collins Syndrome

- Etiology
  - Dominant
  - Arrested development of structures
    - primarily from the first branchial arch
    - probably also involving the second arch
  - Occurs between the 5th and 9th weeks of gestation

Treacher-Collins Syndrome (Mandibulofacial Distosis)

1900
Treacher-Collins Syndrome

- Signs and symptoms
  - Deformities of facial bone structure
  - Malformed middle and outer ear
  - Notch in lower eyelid
  - Cleft lip & palate
  - Anomalies of bones in extremities
- Hearing loss
  - Auricle, EAC, and middle ear -- malformed or totally absent
  - Usually a maximum conductive loss
  - Occasionally, inner ear is affected as well

Audiological Treatment

- Great hearing aid users!!
- Microtia = one of few instances where BC aids are indicated

Otological Treatment

- Microtia
  - Plastic surgery
  - Plastic prosthesis
- Atresia
  - Bilateral atresia - almost always try to open one ear canal with surgery
  - When hearing is OK, risks probably outweigh the potential benefits of surgery:
    - High risk of "iatrogenic" facial paresis
    - Very difficult and dangerous if middle ear cavity is absent

External Otitis

Inflammation of the outer ear

External Otitis

- Rare, unless protective lining is damaged by some agent:
  - Moisture
    - Maceration (softening due to soaking) after swimming
    - Prolonged exposure to elevated temperature and humidity.
  - Trauma
Symptoms

• Swelling (edema)
• Redness (erythema)
• Ear pain (otalgia)
• Drainage (otorrhea)
• Skin eruptions
• Polyps (fleshy masses)
• Conductive hearing loss? Depends on patency of ear canal.

Cerumenosis

• Impacted ear wax in EAC
• Symptoms
  – Reports of dizziness and tinnitus
  – Conductive hearing loss
    • Degree depends on extent of occlusion
    • 40 dB maximum loss with total closure

Foreign Objects

"Don't stick anything smaller than your elbow in your ear"

• Pieces of food (corn, beans, rice, etc.)
• Pebbles
• Insects
• Etc.
Collapsed Canals

- When ear canals close due to pressure from the earphone headband during audiometric testing
- Most common in young children and geriatrics
- Tip Offs
  - Conductive loss (especially at high frequencies) with no history of middle ear disease
  - Variability in threshold responses

Solutions

- Inserts
- Place phones up and back
- Hand held earphone

Otitis Media

Inflammation of the middle ear

Otitis Media Incidence

- Primarily a childhood disease (< age 8)
  - As many as 10% have frequent episodes
- High incidence populations:
  - Cleft palate (≥ 50%)
  - Downs syndrome
  - Native Americans
    - 2/3 have otorrhea before age 1
    - 90% by age 2
  - Allergy (62% of allergic preschoolers)
**Eustachian Tube Physiology**

- Normally closed – passive
- Opens 1000/day
- Functions to ventilate & drain middle ear

**Eustachian Tube Maturation**

- At birth
  - 13 mm long
  - 10° angle
- Adult (reached by age 7)
  - 35 mm long
  - 45° angle
- Reason for increased vulnerability of young children to ME infection.

**Acute Otitis Media**

- Recent, usually abrupt, onset of signs and symptoms
- Effusion indicated by:
  - Bulging tympanic membrane
  - Limited or absent TM mobility
  - Air-fluid level behind the tympanic membrane
  - Otorrhea
- Signs or symptoms of inflammation
  - Distinct erythema of the tympanic membrane or
  - Distinct otalgia interferes with normal activity or sleep

**Acute OM - Early**

**Otitis Media with Effusion**

- Effusion
- No signs or symptoms of acute infection
Treatment: Acute Otitis Media  
AAP (2004)

• < 6 mo.
  – Antibiotics
• 6 mo. to 2 yrs.
  – Antibiotics if Dx is certain, or illness = severe
  – “Observation option” = defer antibiotic Rx 2-3 days
  • If Dx is uncertain and illness is not severe

Treatment: OME  
AAP, AOHNS, AAP (2004)

• “Watchful waiting” for 3 months
  – 75 – 90 % resolve spontaneously
• Still OME at 3 months – test hearing
  – If significant hearing impairment exists:
    • Speech-language eval
    • PE tubes

Serous OM

Secretory OM

SPA 608 - Lundeen©
Otitis Media

“Tympanogenic” Disease

• Four routes of infection spread:
  – Tegmental wall = fractured or eroded, or open infantal suture » meningitis.
  – Posterior wall » mastoiditis.
  – Jugular wall » systemic disease.
  – Opening of medial wall (via semicircular canal, windows, or other structure) » labyrinthitis.

Myringotomy

• Incision in the anterior-inferior quadrant of the eardrum, usually with aspiration of fluid from middle ear.
• Purpose:
  – drain fluid from middle ear
• Heals in 4 - 5 days

Tympanostomy Tubes

Pressure-Equalization (P.E.) tubes

Polyethylene tubes placed through the eardrum to keep a myringotomy incision open.

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Tympanostomy Tubes

• Purpose
  – Ventilate middle ear to compensate for eustachian tube dysfunction
  – Prevents build-up of negative pressure
  – Effective in restoring normal hearing
• Usually stay in place 6-8 months.
• If >1 year -- refer to otologist for recheck!