Cardiovascular Disease and Hearing Loss

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Topics

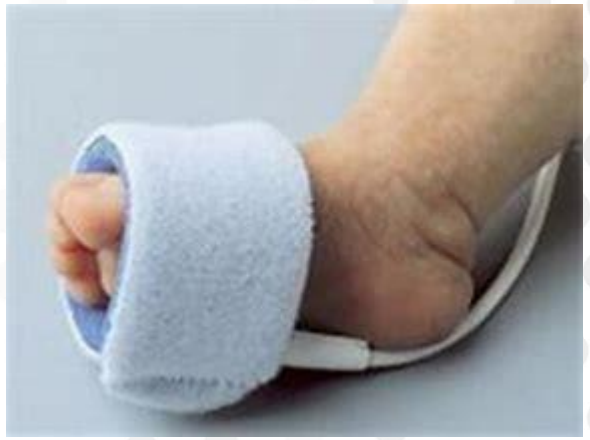
- Pediatric vs Adult Cardiac-Related Hearing Loss
- Etiologies of Comorbid Cardiac Disease/Disorders and Hearing Loss
- Hearing Loss Associated with Cardiac History
- Management Suggestions for the Patient with Cardiac History

Learning Objectives

- Describe common cardiovascular diseases in adults and the likely mechanisms for hearing loss
- Understand the mechanisms for acquired hearing loss as the result of medical or surgical intervention, particularly in the pediatric patient
- Describe the role of the audiologist in the management of comorbid cardiovascular disease/disorder and hearing loss, including possible updates to audiologic practice

Etiology – Infants and Young Children

- Congenital heart malformations
  - Most common congenital birth defect
  - ~40,000 children in US each year (8/1000)
  - ~30% will require intervention within first year of life
    - Tetralogy of Fallot (ToF)
    - Pulmonary atresia
    - Total anomalous pulmonary venous return (TAPV)
    - Transposition of the great arteries (TGA)
    - Tricuspid atresia
    - Truncus arteriosis
    - Hypoplastic left heart syndrome (HLHS)



https://ftp.cdc.gov/pub/Health_Statistics/NCHS/NHIS/SHS/2016_SHS_Table_A-1.pdf
https://www.cdc.gov/ncbddd/heartdefects/data.html

Etiology – Infants and Young Children

- Genetic Mutations (Alleles)
  - >100 individual genes associated with hearing loss that are also associated with cardiac abnormalities
  - Encode:
    - Signal transduction pathways
      - Neural network
    - Ion channels
      - Organ of Corti
      - Sodium/potassium balance in endolymph
    - Mitochondrial proteins
      - Cell energy
      - Metabolism of glucose and oxygen
    - Enzymes involved in lysosomal functions
      - Waste removal from cell
      - Impaired - toxins and cell death

Etiology – Infants and Young Children

- Syndromes including cardiac disorders and hearing loss
  - Down Syndrome (Trisomy 21)
  - Goldenhar (oculo-auriculo-vertebral) syndrome
  - Noonan syndrome
  - CHARGE syndrome
  - Velocardiofacial syndrome (VCFS, 22q11.2 Deletion syndrome, 22q-, DiGeorge syndrome)
  - Williams syndrome
  - Jervell Lange-Nielsen syndrome

Down Syndrome

https://www.cdc.gov

- Prevalence: 1/800
- Mild-moderate intellectual disability
- Facies: flattened face and bridge of nose, almond-shaped eyes that slant up
- Ears: small ears
- Skeletal: short stature, small hands and feet, small pinky fingers, short neck
- Palmar crease, low tone, often protruding tongue
- Heart defects (50% of affected individuals)
- Hearing loss: conductive most common, progressive SNHL with age

Goldenhar (Oculo-Ariculo-Vertebral) Syndrome

- Prevalence: 1/3000 – 1/5000
- Intellectual ability unaffected
- Facies: hemifacial microsomia, wider than normal mouth, cleft lip and/or palate
- Ears: partially formed or missing ears, blind auditory canal
- Skeletal: spinal and rib deformities, scoliosis
- Heart defects (5-58% of affected individuals): VSD and TOF most common
- Hearing loss: conductive most common, reports of SNHL

Photo: https://www.facebook.com/pg/Goldenhar-Syndrome-in-Children-and-Raising-Awareness

Noonan Syndrome

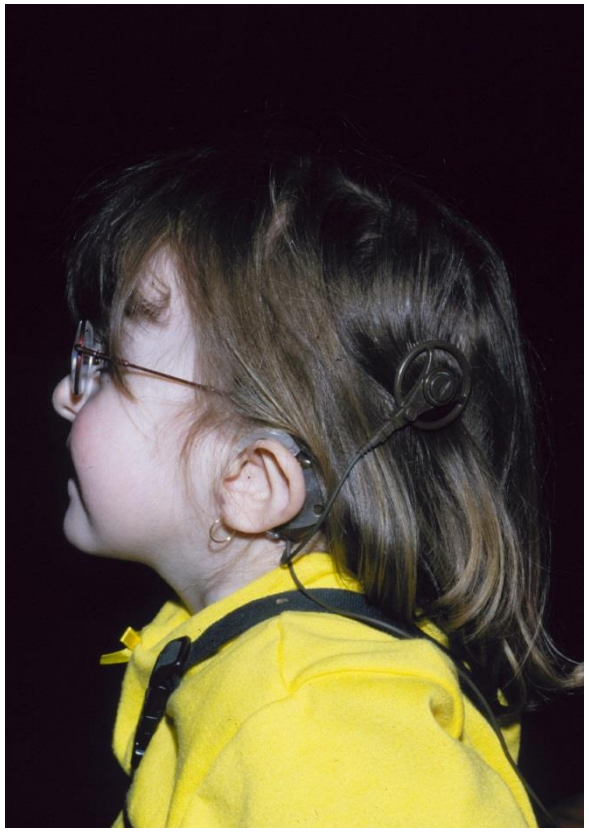
- Prevalence: 1/1000 – 1/2500
- Intellectual ability unaffected
- Facies: Deep philtrum, wide-set eyes, posteriorly rotated low-set ears, short, webbed neck and low hairline at back of neck
- Skeletal: pectus excavatum or pectus carinatum, scoliosis, short stature (within normal limits at birth)
- Heart defects: pulmonary valve stenosis (most common), hypertrophic cardiomyopathy
- Hearing loss: ~35% of affected individuals, conductive most common, SNHL in certain genetic variants which can be progressive

Photo: https://www.facebook.com/pg/NoonanSyndromeAwarenessAssociation

CHARGE Syndrome – Coloboma, Heart Defects, Choanal Atresia, Growth Retardation, Genital Anomalies, Ear Abnormalities

- Prevalence: 1/8500 – 1/10,000
- Wide range in intellectual ability
- Facies: square-shaped face, facial asymmetry, prominent forehead, flat midface
- Ears (>90% of affected individuals): Abnormal outer ears, ossicular malformations, Mondini malformation and absent or hypoplastic semicircular canals
- Cranial nerve abnormalities (swallowing problems, facial paralysis, diminished or absent sense of smell), hypogonadotropic hypogonadism, tracheoesophageal fistula
- Heart defects (75-80% of affected individuals): varied in severity
- Hearing loss: consistent with ear malformations, conductive and SNHL, ~50% severe-to-profound

CHARGE Syndrome



https://en.wikipedia.org

https://ca.wikipedia.org

https://www.chargesyndrome.org

Velocardiofacial (22q-, DiGeorge) Syndrome

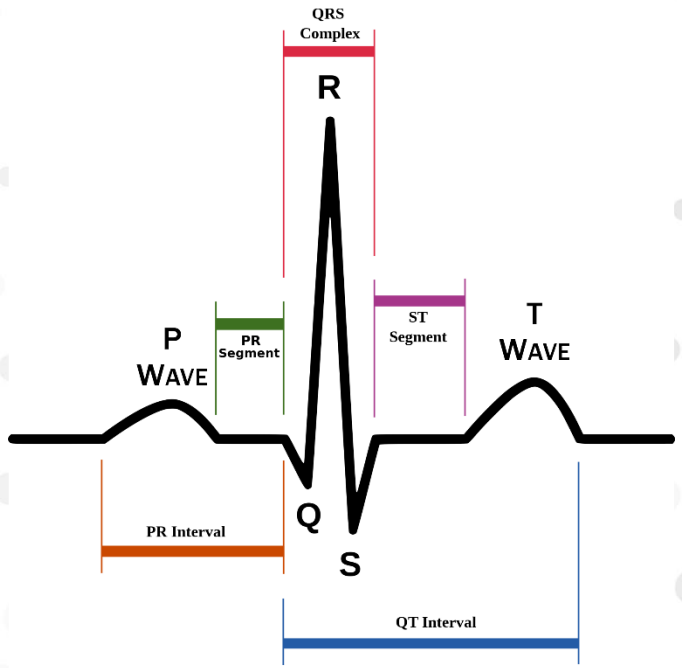
- Prevalence: 1/2000 – 1/7000 (may be underdiagnosed)
- >180 clinical features, physical and behavioral. Reliable diagnosis only through genetic testing.
- Developmental delay, psychiatric disorders
- Facies: characteristic, but not necessarily abnormal, submucous cleft palate
- Ears: structural abnormalities of middle and inner ears
- Heart defects (70% of affected individuals): strong clinical indicator, babies may be screened for 22q- in presence of congenital heart defects
- Hearing loss (~35-45% of affected individuals): usually conductive, some SNHL

Williams Syndrome

- Prevalence: 1/7500 – 1/10,000
- Mild-moderate intellectual disability or learning differences, ADD, anxiety, phobias, outgoing personality
- Facies: broad forehead, short nose with a broad tip, full cheeks, wide mouth
- Dentition: small teeth, widely spaced, crooked, or missing
- Skeletal: short stature, connective tissue abnormalities
- Heart defects (75% in affected individuals): supravalvular aortic stenosis (SVAS) most common
- Hearing loss: 63% in children; 92% in adults, about 50/50 conductive/SNHL

Photo: https://www.facebook.com/pg/williamssyndrome

Jervell Lange-Nielsen Syndrome



http://a-fib.com

- Form of Long Q-T syndrome
  - Irregular heartbeat (tachycardia) especially with exertion
  - Fainting
  - Sudden death
- Hearing loss: congenital bilateral severe-to-profound SNHL
- Prevalence: ~1.6 per million; 1/200,000 in Scandinavian countries
- Mutations in potassium channel genes. Autosomal recessive for hearing loss, but only **one copy** of defective gene necessary to affect heart rhythm.
- Referrals to Genetics and Cardiology

Etiology - Adults

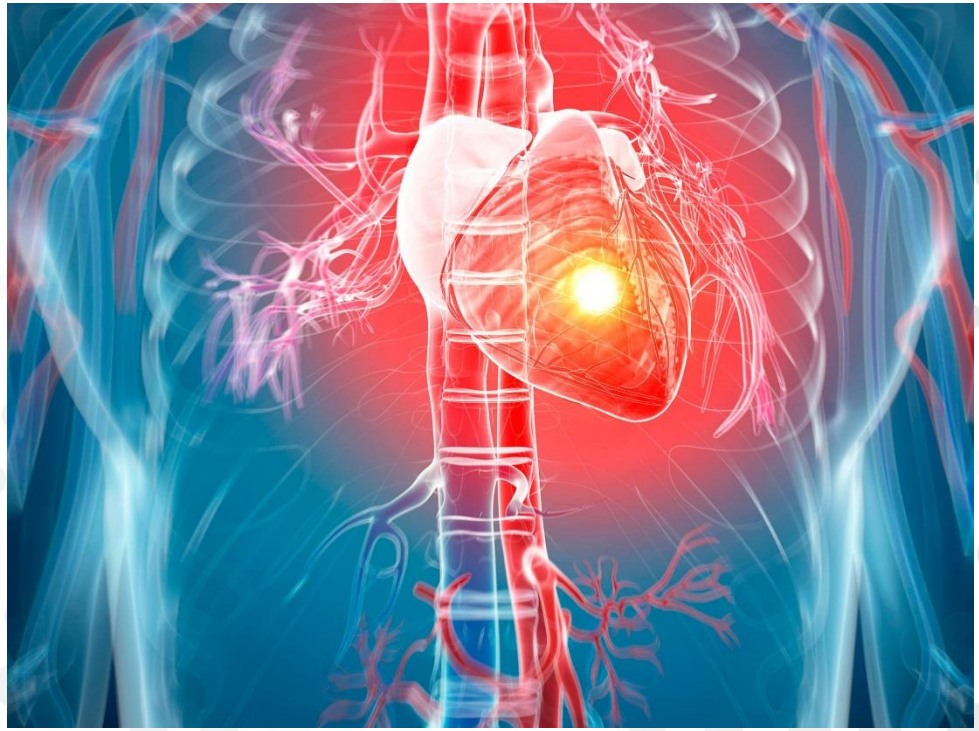
- Congenital heart malformations
- Genetic abnormalities
- Syndromes
- Cardiovascular disease (CVD)

Etiology - Adults

Adults

- Cardiovascular disease (CVD)
  - Leading cause of death in world
    - 11.5% adults in United States with CVC
    - One in four deaths in US from CVD
  - Common conditions
    - Myocardial infarction
    - Stroke

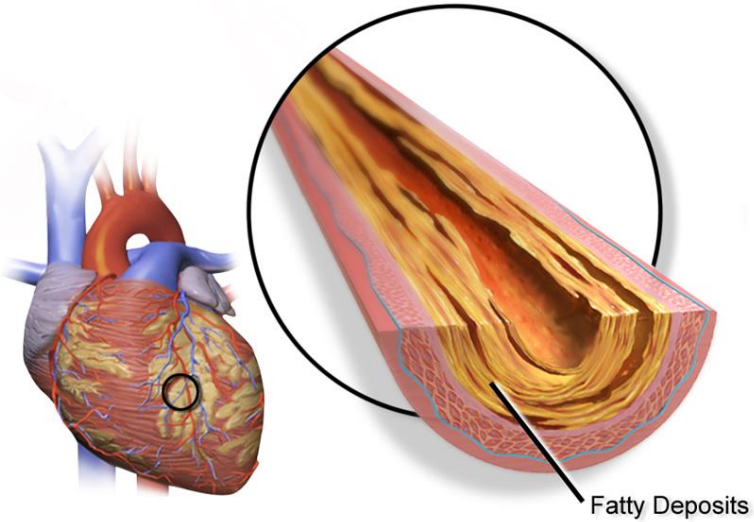
Atherosclerosis
Thrombus (Embolism)
Hypertension
Valve malformations
Atrial fibrillation



http://www.independent.co.uk

https://ftp.cdc.gov/pub/Health_Statistics/NCHS/NHIS/SHS/2016_SHS_Table_A-1.pdf
https://www.cdc.gov/ncbddd/heartdefects/data.html

Cardiovascular Disease - Adults



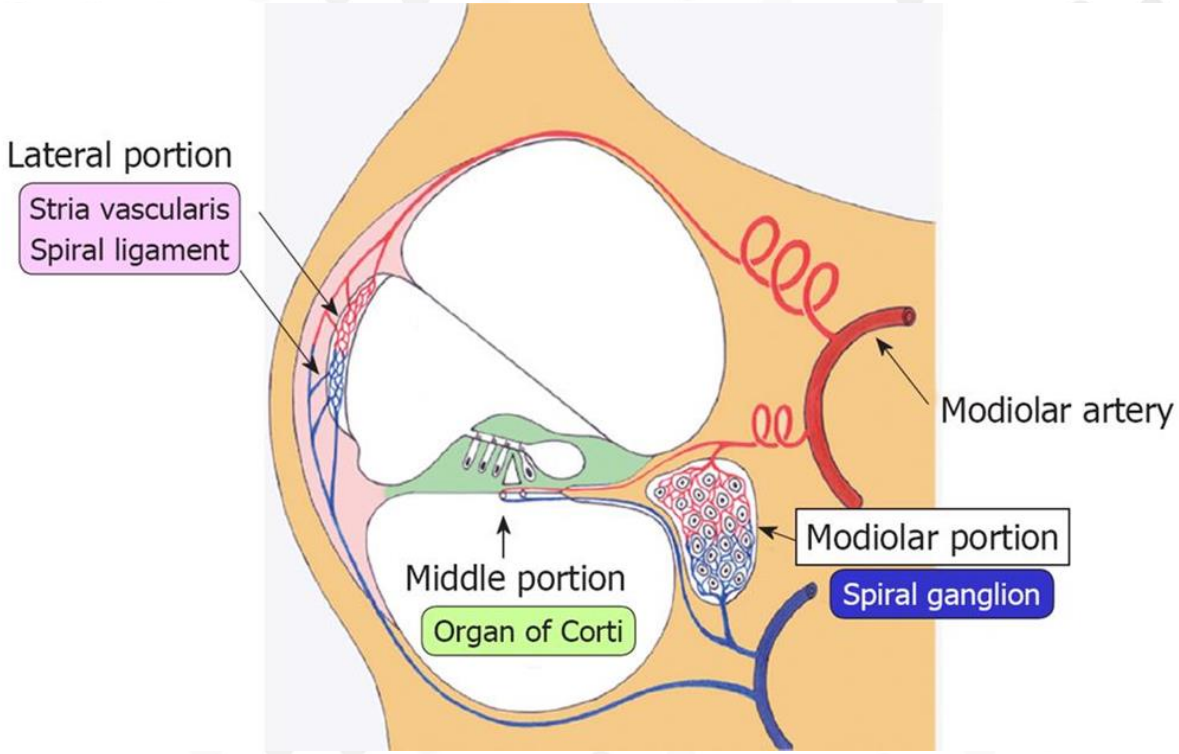
- Atherosclerosis → Embolism → Ischemia
- Hypertension → Atherosclerosis → Embolism → Ischemia
- Valve malformations
  - Mitral valve
  - Tricuspid valve
  - Aortic valves
  - Pulmonary valve

  → Regurgitation / Stenosis → Embolism → Ischemia
- Atrial fibrillation → Embolism → Ischemia
- Atherosclerosis and/or Embolism → Myocardial Infarction → Hypoxia

Illustration: https://wikimedia.org

Hearing Loss from Cardiovascular Disease

- Cochlear potential
  - Energy – glucose and oxygen
  - Sodium and potassium
- Blood supply to cochlea is highly complex
  - Terminal vessels – small capillaries
  - No collateral blood supply
  - Relatively sparse at apex, compared to base of cochlea



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Hearing Loss from Cardiovascular Disease: Predictive Value?

- Sudden onset unilateral SNHL hearing loss
  - Middle cerebral artery (Gur, et al., 2006)
  - Anterior inferior cerebellar artery (AICA)
  - Spiral modiolar artery is branch of AICA

  } Embolism Micro-Embolism (Gyo, 2013)
- Flat SNHL
  - Strial (metabolic) presbycusis
  - Involvement of stria vascularis along entire cochlea
- Low frequency SNHL (Friedland, et al., 2009)

Hearing Loss from Cardiovascular Disease

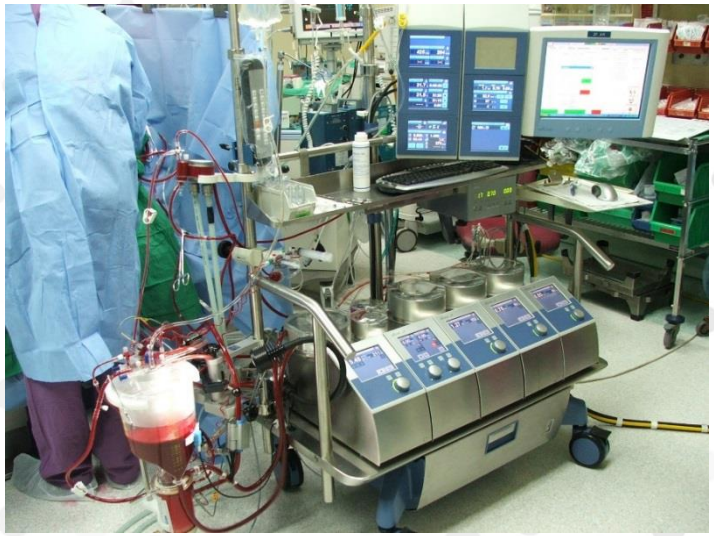
- Risks
  - Sedentary lifestyle
  - Smoking
  - Diet, obesity
  - Family history
  - Comorbid disease

Medical Intervention

- Lifestyle changes
- Medication
  - Ototoxic Diuretics (Generally High-Dose IV)
    - bendroflumethazide (Corzide)
    - bumetadine (Bumex)
    - chlor-thalidone (Tenoretic)
    - ethacrynic acid (Edecrin)
    - furosemide (Lasix)
  - Cardiac Medications Associated with Tinnitus
    - celiprolol
    - flecainide (Tambocar)
    - lidocaine
    - metoprolol (Lopressor)
    - procainamide (Pronestyl)
    - propranolol (Inderal)
    - quinidine (Quinaglute, Quiniedex)

https://www.ntdaily.com

Surgical Intervention



https://commons.wikimedia.org

- Cardiopulmonary bypass (CPB)
  - Heart stopped for bloodless surgical field
  - Blood thinned; reversed when heart restarted
- Deep hypothermic circulatory arrest
  - Body temperature reduced to lower metabolic rate and demand for oxygen
  - Offers cerebral protection
- Extracorporeal membrane oxygenation (ECMO)
  - Similar function to CPB, but used during healing process
  - Support for lungs, or lungs and heart

http://www.pediatriabasadaenpruebas.com

Consequences of Surgical Intervention – Infants and Young Children

- Neurodevelopmental disabilities
  - Most common sequelae, variable
  - Factors
    - Cardiac diagnosis (Gaynor, et al., 2010)
    - Perioperative factors (Gunn, et al., 2016)
    - Patient characteristics (Goff, et al., 2012; Laraja, et al., 2017)
      - Gestational age
      - Low birthweight
      - Head circumference
      - Failure-to-thrive
    - Postoperative factors (Gunn, et al., 2016)
      - Repeated surgeries
      - ECMO

http://apuntesdeunapsicopedagoga.wordpress.com

Consequences of Surgical Intervention - Children

- Hearing Loss
  - Permanent childhood hearing loss (PCHL)
    - 1-3 per 1000 general population (0.1 – 0.3%)
    - 2-4 per 1000 NICU survivors (0.2 – 0.4%)
  - Grasty, et al (2018)
    - 6.9% with SNHL (30% with high-frequency loss)
    - Tested at age 4 years (80% previously undiagnosed)
      - Younger gestational age
      - Longer postoperative duration of stay
      - Confirmed genetic anomaly (22 q-)
  - Bork, et al. (2018)
    - 5.9% with SNHL
      - Cardiac defect (HLHS)
      - Ototoxic medications
      - Hypoxia

Consequences of Surgical Intervention - Adults

- Central nervous system impacts
- Hearing loss – very few reports in the literature
  - High frequency SNHL?
  - Middle and high frequency SNHL with use of ECMO (Aytacoglu, et al., 2006)
  - Sudden unilateral SNHL (Arenberg, et al., 1972; Plasse, et al., 1980; Plasse, et al., 1981)
    - Basilar artery – common location for atherosclerotic disease
    - High venous pressure during anesthesia
    - Arterial plaque rupture
  - Asymptomatic/subclinical signs (Munjal, et al., 2013)
    - Extended high frequency audiometry
    - OAEs

Observations

- Role of audiologist on management team
- Conveying results to medical/surgical team
- Role of PCP and Medical Home model
- Audiologist as “Hearing Health Home”
  - Referrals or recommendations for additional assessments, both medical and developmental/educational
  - Management of hearing loss impact on language, education, socialization, occupation, etc.

http://praxis.blog

Recommendations for Management of Patients

- Increase detail of cardiac history to medical history
  - Cardiovascular disease
  - Cardiac surgery
  - Thrombotic event (deep venous thrombosis, pulmonary embolism, stroke)
  - Syndromes associated with heart malformations and hearing loss
- Include for indications for medical referrals
  - Strial presbycusis
  - Sudden unilateral SNHL
  - Family history of profound bilateral SNHL
- (Re)familiarize with adult and pediatric risk indicators for hearing loss, including updated ototoxic medication list, comorbid conditions, and syndromes, especially those associated with late-onset or progressive hearing loss

Risk Indicators for Hearing Loss in Infants and Young Children (Late Onset)

- Caregiver concern
- Family history of permanent childhood hearing loss
- Neonatal intensive care of >5 days, or regardless of length of stay:
  - ECMO
  - Assisted ventilation
  - Ototoxic medications or loop diuretics
  - Hyperbilirubinemia requiring transfusion
- In-utero infections (CMV, herpes, rubella, syphilis, toxoplasmosis)

Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. Joint Committee on Infant Hearing. *Pediatrics* 2007;120;898

Risk Indicators (cont'd.)

- Craniofacial anomalies
- Physical findings (such as white forelock) associated with a syndrome known to include hearing loss (Waardenburg S.)
- Syndromes associated with hearing loss, progressive hearing loss or late-onset hearing loss
- Neurodegenerative disorders
- Postnatal infections associated with sensorineural hearing loss
- Head trauma
- Chemotherapy

Thank You!

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