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://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

• 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
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

- 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

http://a-fib.com
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://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 → Embolism → Ischemia
  → Stenosis
- 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

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

• 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
    • flecaïnide (Tambocar)
    • lidocaine
    • metoprolol (Lopressor)
    • procainamide (Pronestyl)
    • propranolol (Inderal)
    • quinidine (Quinaglute, Quiniedex)
Surgical Intervention

• 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
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%)
    • 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.
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)

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!


