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Congenital Heart Defects: An Overview and Impact on Feeding and Development

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Moderated by:
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Disclosure

- The presenter receives a salary for her work as an Associate Professor and Director of Clinical Education at the University of Louisville. She also received a stipend from SpeechPathology.com to present this course.

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Learner Outcomes

1. Participants will be able to describe 3 characteristics of congenital heart disease.
2. Participants will be able to describe 3 congenital heart disease diagnoses.
3. Participants will be able to identify 3 ways congenital heart disease impacts feeding and/or development

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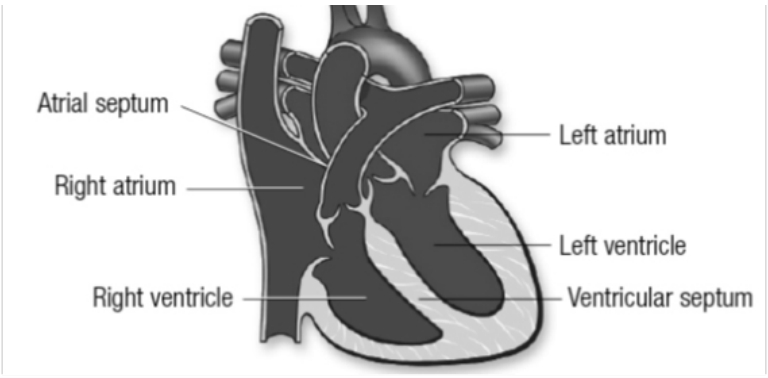
The Normal Heart



The Valves

- Valves
  - Mitral
  - Tricuspid
  - Aortic
  - Pulmonary

Blood Flow – Super Simplified

- Right side of the heart receives blood from the body
- Right side pumps blood from the body to the lungs to get receive oxygen
- Left side of the heart receives the oxygenated blood and sends it out to the body
- Before each heart beat the heart fills with blood
- The muscle contracts and move the blood along

How the Heart Feeds the Body

- Blood delivers O_2 to the cells of the body
- The cells in the body use the O_2 and that makes carbon dioxide (CO_2)
- CO_2 & other gets carried away in the blood vessels
- In the lungs the CO_2 is removed on exhalation
- On inhalation blood is oxygenated and can be used again

When the Heart Doesn't Work Well

- Cells do not get the O_2 they need
- CO_2 does not get expelled efficiently
- Everything in the body has to work harder (including the heart!)
- Necessary bodily functions are impaired

Congenital Heart Disease (CHD) Facts

- Most common birth defect
- Occurs in 8:1,000 births
- Diagnosis most often in infancy/early childhood
- Approximately 25% of infants with CHD require invasive treatment/1st year of life

Common Early Symptoms of CHD

- Slow feeding
- Breathlessness
- Irritability
- Pallor and sweating
- Failure to gain weight

Common Early Signs of CHD

- Cyanosis
- Tachycardia
- Tachypnea
- Cardiac Murmur
- Cardiomegaly
- Shock

Cyanosis Presentation

- Bluish discoloration of skin
- $SaO_2 < 85\%$
- Most significant sign of serious cardiac anomaly

Classification of CHD

- Cyanotic CHD
  - Decrease pulmonary blood flow
  - Mixed blood flow
- Acyanotic CHD
  - Increase pulmonary blood flow
  - Obstruction of blood flow

Cyanotic CHD

Decreased Pulmonary Blood Flow

- Tricuspid Atresia
- Tetralogy of Fallot (ToF)

Mixed Blood Flow

- Transposition of the Great Arteries
- Total Pulmonary Venous Return
- Truncus Arteriosus
- Hypoplastic Left Heart Syndrome

Tetralogy of Fallot (ToF)

- Defect with 4 problems
- Hole between 2 lower chambers (Ventricular septal defect)
- Obstruction from heart to lungs (Pulmonary stenosis)
- Aorta lies over the hold in the lower chambers (aorta enlarged and appears to arise out of both R & L ventricles)
- Muscle surrounding lower right chamber becomes overly thick (working so hard becomes thickened)

ToF Management

1. Determine if child's O_2 is in safe range
2. If critically low then provide a prostaglandin infusion to keep PDA open (this helps increase pulmonary blood flow)
3. If O_2 levels are adequate/mild cyanosis may go home in first week of life
4. Complete repair at approximately 6 months of age
5. If decline in O_2 stats surgery performed earlier

Surgical Repair of ToF

- Closure of VSD with synthetic Dacron patch so blood flows normal from left ventricle to aorta
- Resection of pulmonary stenosis and right ventricle to enlarge outflow pathway
- Additional intervention may be required if additional problems

Survival Rate of Infants/Children

with ToF

- In the absence of additional problems more than 95% of infants successfully undergo surgery in first year of life
- Long term cardiac function is excellent
- Lingering issue with a leaky pulmonary valve (some backflow of blood into the R ventricle causing it to work harder)
- Follow up intervention may be required (surgery or balloon dilation)

Hypoplastic Left Heart Syndrome (HLHS)

- Left side of heart does not form correctly
- May have co-occurring atrial septal defect
- Effects ability to pump O_2 rich blood to body
- May be diagnosed in utero or first few days of life

Management of HLHS

- Medication
- Nutrition
- Surgery
  - Norwood (within 2 weeks of birth)
  - Bidirectional Glenn Shunt (~4-6 months)
  - Fontan Procedure (between 18 months – 3 years)

Survival Rate of Infants/Children

with HLHS

- Surgery is not curative and lifelong complications may exist
- Survival to 1 year of age (55.2%) CDC
- Survival to 8 years of age (50.4%)
- Lowest chance of survival across multiple ages compared to children with any other birth defect studied

Management of HLHS

- Medication
- Nutrition
- Surgery
  - Norwood (within 2 weeks of birth)
  - Bidirectional Glenn Shunt (~4-6 months)
  - Fontan Procedure (between 18 months – 3 years)

Acyanotic CHD

Increased Pulmonary Blood Flow

- Atrial Septal Defect (ASD)
- Ventricular Septal Defect (VSD)
- Atrioventricular Canal Defect
- Patent Ductus Arteriosus (PDA)

Obstruction of Blood Flow From Ventricle

- Pulmonary Stenosis
- Aortic Stenosis
- Coarctation of the Aorta

Ventricular Septal Defect (VSD)

- Wall that forms between two ventricles does not fully develop
- Can have VS defects in more than one place
- Blood flows from L ventricle through defect to R ventricle and into lungs
- Excess blood pumped in lungs creates extra work
- Occurs 42:10,000
- Usually diagnosed after birth

Management of VSD

- Medicine
- Nutrition
- Surgery (depends on size, problems resulting from defect, and/or may close spontaneously)

Patent Ductus Arteriosus (PDA)

- Fetal ductus arteriosus fails to close
- Result is shunting of oxygenated blood from aorta to pulmonary arteries
- In the presence of other congenital heart defects the PDA may be purposefully kept open

Management of PDA

- Observation – if signs of increased cardiac workload/pulmonary vascular changes then closure recommended
- PDA closure – dependent on size of PDA, age of patient, degree of shunting, symptomology
- Premature infant – Indomethacin or Ibuprofen
- Term infant <5kg symptomatic – Digoxin and Furosemide, if not suitable size for device closure then surgical ligation
- Infants/children >5k-Percoutaneous occlusion (if not possible for particular child then surgical ligation)

Pulmonary Stenosis

- Thickened/fused heart valve that does not fully open
- Pulmonary valve allows blood flow out of the heart into pulmonary artery and then into the lungs
- Pressure much higher than normal in R ventricle-increased effort to pump blood into lungs
- If unable to access pulmonary valve-blood will travel other routes
- May be diagnosed in utero or shortly after birth

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Management of Pulmonary Stenosis

- Medication (to keep PDA open)
- Nutrition
- Treatment dependent on severity
  - May perform cardiac catheterization – balloon to expand or stent to keep PDA open
  - Surgery to widen or replace the valve

Genetic Syndromes CHD

Genetic Syndromes Associated w/CHD

- Down syndrome – 40-50% (CHD)
- Turner syndrome – 25-45% (CHD)
- Williams syndrome – 75-80% (CHD)
- Noonan syndrome – 70-80% (CHD)

Down syndrome

- 1:700 babies born in U.S. diagnosed with Down syndrome
- Most common chromosomal condition
- Developmental delays
- Higher incidence of infection, respiratory, vision, hearing problems
- Higher incidence of thyroid problems

Down syndrome and CHD

- Most commonly occurring heart defects in children with Down syndrome
- Atrioventricular septal defect
- Ventricular septal defect
- Persistent PDA
- ToF

Turner syndrome

- Affects 1:2000 females
- Small for gestational age
- 3rd-10th percentile infancy
- 3rd percentile childhood
- Below 3rd percentile, no growth spurt
- Most symptoms occur due to loss of genetic material from one of the X chromosomes

Turner syndrome and CHD

- Coarctation of Aorta
- Bicuspid Aortic Valve
- Aortic Stenosis
- Hypoplastic Left Heart Syndrome

Williams syndrome

- 1:7500-10,000 people
- Affects many parts of the body
- Mild to moderate intellectual disability
- Unique personality characteristics
- Distinctive facial features
- Visual-spatial difficulty
- Tend to do well with spoken language

Williams syndrome and CHD

- Peripheral Pulmonary Stenosis
- Aortic Stenosis

Noonan syndrome

- 1:1,000-2,500 people
- Short stature (5—75 %)
- Skeletal malformations
- Distinctive facial features (wide-spaced eyes, deeper philtrum, low-set ears with posterior rotation, poor dental alignment, micrognathia, webbing

Noonan syndrome and CHD

- Pulmonary Stenosis
- Hypertrophic Cardiomyopathy
- Atrial Septal Defect

Malnutrition in Children with CHD

- Inadequate intake
- Increased energy needs
- Inefficient nutrient absorption/utilization

Inadequate Intake

- Side effects of medication
- Fatigue during feeding
- Swallow problems
- Oral aversion
- Neurological dysfunction secondary to prematurity/operative complications

Inadequate Intake

- GERD
- Early satiety
- Tachypnea
- Fluid restriction
- Frequent periods of NPO
- Recurrent respiratory infections
- Psychosocial issues

Increased Energy Needs

- Chronic metabolic stress
- Post-op metabolic stress
- Tachypnea
- Tachycardia
- Cardiac hypertrophy
- Increased sympathetic activity
- Infections, fever, sepsis

Inefficient Nutrient Absorption

- Vomiting
- Edema of the small bowel (as result of right sided heart failure) leading to malabsorption
- Excessive nutrient loss
- Gut mucosal atrophy leading to malabsorption in children with pre-existing malnutrition

Medication Related Side Effects

- Lidocaine – Nausea, vomiting
- Warfarin (Coumadin) – diarrhea, nausea, GI pain/cramps
- Bumetanide (Bumex) – GI cramps, nausea, vomiting, electrolyte abnormalities
- Digitalis (Digoxin) – Nausea, vomiting, anorexia, feeding intolerance, electrolyte imbalance
- Fentanyl – nausea, vomiting

Factors Impacting Life Expectancy

- Advances in:
  - Surgical techniques
  - Cardiac catheterization
  - Interventional cardiology
  - Noninvasive imaging
  - Early diagnosis/Fetal assessment
  - Complex critical nursing care

Neurodevelopmental Complications

- Learning disabilities
- Visual motor integration
- Motor delays

(Marino et al, 2012; Wernovsky, 2006)

Attention Deficit Hyperactivity Disorders

- Common diagnosis in children with CHD (Shillingford, et al., 2008)
- Early claims - stimulant meds for ADHD result in cardiovascular damage
- Current status – safe to use stimulant meds with CHD (Cooper et al., 2011; Marino et al., 2012)
- Recommendation to consult cardiologist prior to initiation (Batra et al., 2012)

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Impact of Complexity of CHD

- Lower incidence of neurodevelopmental disabilities/milder forms of CHD
- Higher incidence of neurodevelopmental disabilities/complex forms of CHD

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CHD and the School-Age Child

- Promotion of health in this population includes:
- Chronic disease management
- Health maintenance education
- Preventative focus for future problems

Impact of CHD on School Performance

- Increased absence – medical appointments
- Increased absence – medical complications
- Neurodevelopmental complications

Parents and the Child with CHD

- Report worry/fear
- Report anxiety/apprehension
- Impacts parenting skills and discipline

(Duncan & Caughy, 2009; Lee & Rempel, 2011)

Potential Long-Term Complications

- Follow up surgery/cardiac catheterization
- Heart failure/ventricular dysfunction
- Hyperviscosity of blood
- Stroke/thrombosis
- Endocarditis
- Arrhythmia
- Sudden death
- Myocardial infarction
- Systemic &/or pulmonary hypertension
- Renal problems
- Limited physical activity
- Dependence on meds
- Need for SBE prophylaxis

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