Overview and Nutrition Management of Congenital Heart Disease in Pediatric Patients

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Outline

- Introduction
- Definitions, prevalence, etiology, & diagnosis
- Normal Heart Anatomy & Physiology
- Types of Congenital Heart Defects: Common & Complex
- Anatomy & Physiology
- Medical & Surgical Intervention
- Nutrition Management
- Challenges & Complications in CHD Patients
- Case Study
- Summary
- References

Learning Objectives

- Define congenital heart disease and discuss the prevalence, etiology, and diagnosis
- Demonstrate a basic knowledge of the anatomy and physiology of a normal heart
- Identify common and complex types of congenital heart defects (CHDs)
- Discuss the anatomy and physiology of 5 CHDs
- Discuss the medical/surgical interventions of each
- Discuss the nutrition management of CHD and apply knowledge in a hypothetical pediatric case

Definition & Prevalence

- Congenital Heart Disease (CHD)
  - Congenital = _____________________________
  - Definition: problem with the heart’s structure and function that is present at birth
  - Congenital heart “__________________” more accurate than “disease”
    - Defects occur when the heart or blood vessels near the heart do not develop normally before birth
- Most common birth defect in the United States (U.S.) and leading cause of infant death
- ~1% or ________________ infants affected each year
- 1 out of every 120 infants is born with a CHD in the U.S.
- ~1 million infants are born with CHD worldwide
Etiology

- 80% Unknown
  - Multifactorial inheritance with association of both _____________ & _____________ contributors
  - Genetic Factors:
    - Heredity – rare that two children in the same family might have a defect
  - Environmental Factors:
    - Maternal Conditions
      - Smoking, drugs, and alcohol
      - Obesity
      - ______________________
      - Viral infections (i.e. rubella)

- 20% Known
  - Chromosome abnormalities (_____________%)
    - Down syndrome (40-50%)
    - Trisomy 18
    - Williams syndrome
  - Mendelian Syndromes (3-5%)
    - Noonan syndrome (_____________%)
  - Non-syndromal single gene disorders
    - 30 genes have been linked to CHDs
    - _______________ factor genes most common
    - NKKX2-5: TOF, HLHS, TGA, __________, ASD, DORV

CHD Risk Factor: Maternal Diabetes

- _______________ increased risk of CHDs in infants from mothers with pregestational diabetes
- Maternal glucose control affects cardiogenesis at a very early stage of _______________ development (_____________ gestation)

Diagnosis

- Prenatally
  - Fetal _______________
- Postnatally (shortly after birth)
  - Physical exam
    - Signs & Symptoms: Heart murmur, _______________, shortness of breath (SOB), rapid breathing, failure to thrive (FTT), signs of heart failure
      - _______________
  - Electrocardiogram (EKG)
  - Chest X-Ray
Healthy Heart Anatomy & Function

Child & Adult Heart

Fetal Heart

- **Oval foramen**
  - Allows oxygenated blood from the placenta circulation to bypass lungs and go straight to left side of the heart

- **Ductus arteriosus**
  - Allows mixed blood to go back to placenta for oxygenated blood (fetal lungs not developed)
After Birth

- When the baby is born and the umbilical cord is cut, the lungs are now needed to supply oxygen. The lungs expand, their blood vessels relax to accept more flow
- Foramen ovale & ductus arteriosus begin to close
- Shunts sometimes remain ____________ or can be kept open with medication (patent)
  - BAD: if infant otherwise healthy
  - GOOD: if infant has CHD, as it can allow mixed blood to circulate until defect repaired

Common Types of CHDs

<table>
<thead>
<tr>
<th>Defect Category</th>
<th>Defect Name</th>
</tr>
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<tbody>
<tr>
<td>right-to-left cardiac shunting (abnormal blood flow)</td>
<td>• Ventricular septal defect (VSD)</td>
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<tr>
<td></td>
<td>• Atrial septal defect (ASD)</td>
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<tr>
<td></td>
<td>• Patent ductus arteriosus (PDA)</td>
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<tr>
<td></td>
<td>• Atrioventricular septal defect (AVSD)</td>
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<tr>
<td>left-to-right cardiac shunting</td>
<td>• Tetralogy of Fallot (TOF)</td>
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<tr>
<td></td>
<td>• Pulmonary stenosis (PS)</td>
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<tr>
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<td>• Aortic stenosis (AS)</td>
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<td>• Interrupted aortic arch (IAA)</td>
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<td>• Coarctation of the aorta (CoA)</td>
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<td></td>
<td>• Transposition of the great arteries (TGA)</td>
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<tr>
<td></td>
<td>• Total anomalous pulmonary venous return (TAPVR)</td>
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<tr>
<td></td>
<td>• Double outlet right ventricle (DORV)</td>
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<tr>
<td></td>
<td>• Double inlet left ventricle (DILV)</td>
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<tr>
<td></td>
<td>• Pulmonary atresia (PA)</td>
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<td></td>
<td>• Tricuspid atresia (TA)</td>
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<tr>
<td></td>
<td>• Hypoplastic left heart syndrome (HLHS)</td>
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Ventricular Septal Defect (VSD)

- Most common form of CHD
  - _________________ of all CHD cases
- Description
  - Hole in the wall (_____________) separating the two lower chambers of the heart
- Allows blood to mix
- Red blood from high pressured left side to low pressured right side
- Extra blood pumped into pulmonary arteries ______ work of heart & lungs, can lead to congested lungs
- Holes can vary in size
  - Small VSD- ____________________________
    with normal growth & development
  - Large VSD- pulmonary HTN & CHF with __________ growth & development

**VSD: Medical Management**

- Small VSD
  - Observation only
  - Often___________ on their own
  - Infective (______________________) endocarditis prophylaxis
    - ____________________________ used in prevention of an infection caused by bacteria that enter the bloodstream and settle in the ________________ of the heart, heart ______________, or blood vessel
- Large VSD
  - Diuretics
  - ______________ feeding of caloric ______________ formulas
  - Infective endocarditis prophylaxis

**VSD: Surgical Repair**

- Small VSD:
  - Surgery usually ______ indicated unless another defect is involved
- Moderate to large VSD:
  - Surgery indicated if __________ or ______________ cannot be improved with medical management
  - Hole should be closed surgically within first _______________ of age
- **Temporary repair**
  - Child very small or very sick
  - Pulmonary artery ________ is placed around the pulmonary artery
    - Restricts the extra blood flow to the lungs
  - Hole closed when the child is older

- **Permanent repair**
  - Closure of defect with stitches or a patch

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**Tetrology of Fallot (TOF)**

- _________ of all CHD cases
- Four Defects:
  1. ________________ Septal Defect: Abnormal opening or hole that allows blood to pass from the RV to the LV without going through the lungs
  2. Pulmonary Stenosis: Narrowing at or beneath the pulmonary valve that partially blocks the flow of blood
from the right side of the heart to the lungs

3. Overriding ________: aorta lies directly over the VSD

4. Right ventricular ________________: right ventricle is more muscular than normal and often enlarged
   - Most common form of CHD that produces ________________
   - Growth retardation can occur

TOF: Medical Management

- Hypercyanotic spells
  - Severe cyanosis + paleness, ____________, altered level of consciousness
- Occur after __________ or after feeding
- Treatment:
  - ________________ positioning
  - Morphine sulfate
  - Oxygen
  - IVF
  - Sodium bicarbonate
  - Phenylephrine
  - Propranolol
- Infective endocarditis prophylaxis
- **____________________ indicated for all patients

TOF: Surgical Repair

- Blalock-Taussig Shunt (BT-Shunt)
  - ________________
  - Performed if infant ____________ or pulmonary arteries ____________ for corrective procedure
  - Small tube from subclavian artery into pulmonary artery
    - Allows blood to enter lungs

- Complete Repair
  - ________________ of age
• Infants who had palliative shunt may have complete repair ___________ months of age
  o VSD closed with patch
    ▪ Allows oxygen poor blood to flow into lungs
  o Incision across pulmonary annulus + enlargement with wide patch
    ▪ Relieves pulmonary narrowing

Transposition of the Great Arteries (TGA)

• ___________of all CHD cases
• Description:
  o Pulmonary artery & aorta are in opposite positions
    ▪ Aorta carrying oxygen ___________blood to body
  o Pulmonary artery carrying oxygen ___________blood to lungs
• ASD, VSD, & PDA necessary for survival
• Moderate to severe cyanosis

TGA: Medical & Surgical Management

• Medical
  o Prostaglandin E1 infusion to keep PDA open
  o Correction of metabolic acidosis
  o Diuretics (i.e. Aldactone, Lasix)
  o Frequent feedings of caloric dense formula
• Surgical
  o Arterial Switch
    ▪ Aorta and pulmonary artery divided
    ▪ Aorta and pulmonary artery _________ to proper position
  o Coronary arteries
    ▪ Removed from aortic trunk & attached to pulmonary trunk
    ▪ Supply heart with _____________ blood
  o Hole between VSD _____________ with a patch or stitches
  o PDA tied off and/or cut

Arterial Switch Operation Step 1

Arterial Switch Operation Step 2

Arterial Switch Operation Step 3
**Complex Single Ventricular Defects**

**Hypoplastic Left Heart Syndrome (HLHS)**

- _______ of all CHD cases
- Most common cause of ______________ from CHD in 1st month of life
- Description:
  - ____________ ventricle underdevelopment (left ventricular hypoplasia)
  - Mitral valve not formed (mitral atresia) or undeveloped (mitral stenosis)
  - ____________ valve not formed (aortic atresia) or underdeveloped (aortic stenosis)
  - Ascending aorta underdeveloped & narrowed (CoA)
  - ASD
- Become extremely ill within 1st few ___________ of life

**HLHS: Medical Management**

- ________________ infusion to keep PDA open
- Correction of metabolic acidosis
- Infective endocarditis prophylaxis
- Frequent feedings of caloric dense formula

**Tricuspid Atresia**

- _________ of all CHD cases
- Description:
  - No tricuspid valve
  - Small right ventricle
  - ASD & VSD
  - PS
- Allows for _______________ in the right atrium to flow through ASD→ left atrium. The ____________ blood→left ventricle→aorta→body
- Amounts of blood flows through VSD → right ventricle → pulmonary artery → lungs (or PDA)

**Tricuspid Atresia: Medical Management**
- Prostaglandin E1 may be used to keep PDA open until surgical repair
- Frequent feedings of caloric dense formula

**HLHS & Tricuspid Atresia: Surgical Repair**

- **Stage I: Norwood**
  - After _______ of life
    - Shunt between aorta and right pulmonary artery
    - Sano modification (RV-PA conduit)
      - Shunt between right ventricle and main pulmonary artery
    - Pulmonary artery & small aorta fashioned together to make new larger aorta
    - PDA is _______
    - Atrial septum removed
    - Narrowing aorta _______
  - _______

- **Stage II: Bidirectional Glenn Procedure**
  - _______ of age
    - Shunt from Stage I removed
    - _______ from head, neck, and upper body directed to the right pulmonary artery through superior vena cava

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**Stage I - Norwood**

- Blalock-Taussig Shunt
- Surgically Enlarged Atrial Septal Defect
- Homograft Patch (To Reconstruct Aorta)
- Left Atrium (LA)
- Right Atrium (RA)
- Left Ventricle (LV)
- Right Ventricle (RV)

**Stage II - Bidirectional Glenn**

- Divided Blalock-Taussig Shunt
- Superior Vena Cava Attached to Right PA
- Left PA
- Right PA

- Oxygen-rich Blood
- Oxygen-poor Blood
- Mixed Blood

LA: Left Atrium   RA: Right Atrium   LV: Left Ventricle   RV: Right Ventricle
• **Stage III: Fontan Procedure**
  2-4 years of age
  - End to end anastomosis of inferior vena cava to right pulmonary artery through ____________
    - Allows oxygen poor blood to be sent to lungs
  - Tube placed through right atrium & connected to right pulmonary artery
    - Allows rerouting of oxygen ___________ blood to the lungs
    - Heart remains available to receive ___________ blood from lungs & pump to body
  - Holes (______________) placed in tube
    - Allow heart and body to adjust

**Nutrition Management of CHD**

**Nutrition Indication**

- _________________& FTT have long been recognized as common consequences of CHDs
- Infants with cyanotic, including the complex single ventricular physiology defects are put at a _______________ risk for malnutrition
  - Especially during 1<sup>st</sup> and 2<sup>nd</sup> stage repairs
- Adequate nutrition is _________________ for growth, wound healing, and immune function

**Poor Nutrition & Growth**

- Cause is multi-factorial
- Mostly related to imbalance between:
  - Energy intake and energy expenditure
    - Energy deficit
    - Catabolism
- Increased severity of defect = increased energy needs
  - Needs may increase for catch-up growth
  - Median enteral intake < 100 kcal/kg/day of 100 consecutive infants (52 single ventricle physiology) post-operatively
o Measured oxygen consumption with respiratory mass spectrometry
o Found that hypermetabolic response is present within the first 72 hours after Norwood operation

**Causes of Malnutrition in CHD Patients**
- Inadequate ____________
  - Fatigue while feeding, fluid restriction, delayed gastric emptying or motility causing early satiety, increased work of breathing, feeding difficulty (vocal cord injury, uncoordinated suck & swallow)
- ____________ energy needs
  - Increased metabolic expenditure related to increase in cardiac and respiratory work
  - Decreased fat stores and increased lean body mass
- Inefficient nutrient utilization/absorption
  - When combined with poor nutritional intake and increased metabolic rate

**Nutrition Assessment**
- Weight
  - Consider pre-operative weight (euvoletic weight)
- Length
- HC
- Wt-for-length
- Assess for trends on appropriate growth chart
  - Criteria for determination of malnutrition in CHD patients
    - Wt-for-age < 3rd %-ile
    - Ht-for-age < 3rd %-ile
    - Wt-for-length < 3rd % ile
    - Reduction of > 2 %-iles for wt-for-age, ht-for-age, and/or wt-for-length

**Determination of Estimated Needs**

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Energy (kcal/kg)</th>
<th>Protein (g/kg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-0.5</td>
<td>120-150</td>
<td>2.2-3.5</td>
</tr>
<tr>
<td>0.5-1</td>
<td>110-140</td>
<td>1.5-2.5</td>
</tr>
<tr>
<td>1-3</td>
<td>100-120</td>
<td>1.2-2</td>
</tr>
<tr>
<td>4-6</td>
<td>80-100</td>
<td>1.2-1.5</td>
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<td>7-10</td>
<td>60-90</td>
<td>1.0-1.5</td>
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<tr>
<td>11-14</td>
<td>Males: 55-60</td>
<td>1.0-1.5</td>
</tr>
<tr>
<td></td>
<td>Female: 45-60</td>
<td></td>
</tr>
<tr>
<td>15-18</td>
<td>45-55</td>
<td>1.0-1.5</td>
</tr>
</tbody>
</table>
• If catch-up growth indicated, used catch-up growth equation with appropriate wt-age recommendations
• Potassium: 2-5 mEq/kg
• Sodium: at least 2-3 mEq/kg (even if on sodium restriction)
• Supplement iron & vitamin D
  o Breast fed infants
  o Low formula intake

Nutrition Interventions

• Parenteral nutrition
  o Limited because of fluid restriction & medication drips in ICU
  o Concentrate medication drips to maximize fluid availability
  o Consider extra zinc
    ▪ Additional 100 mcg/kg for infants <2.5 kg or 50 mcg/kg infants with open chest
• Enteral nutrition
  o Trophic feeds working up to 24 hour continuous
  o Increase formula caloric density
    ▪ 24-27 kcal/oz: via formula concentration
    ▪ 30-35 kcal/oz (if necessary): via polycose and microlipid
    ▪ Breast milk can be used if fortified to appropriate caloric density
  o Continuous to bolus or oral intake + nocturnal (50% estimated needs over 12 hours) – Acute Setting
    • Or PO with NG-tube back-up
  o Intermittent feeds
    ▪ Every 3-4 hours
• Oral nutrition
  o Transition when clinically feasible after extubation in ICU
  o Approval by SLP
• Home nutrition
  o Ensure family knows recipe & appropriate mixing procedures
  o Written intake & weight gain goals provided with mixing instructions
  o Ensure adequate formula supply (WIC?)
  o Nutrition-related home monitoring criteria includes:
    ▪ Weight loss of 30 g in one day
    ▪ Failure to gain 20 g/day for 3 days
    ▪ 2 episodes of vomiting
    ▪ Loose stools

Nutrition-Related Goals

• Pre-operative period:
  o Provide adequate nutrition to meet patient’s needs until surgery
• Post-operative critical care setting:
  o Initiate nutrition support as soon as possible
Avoid overfeeding

- **Step-down/acute care setting:**
  - Provide adequate nutrition to meet needs & correct deficiencies
  - Transition from PN to EN 100%
  - Transition EN to oral, or combination oral/supplemental EN
  - Age-appropriate or catch-up weight gain & growth
  - Caregivers are receptive in understanding nutrition discharge plan
- **Post-discharge/home setting:**
  - Age appropriate or catch-up weight gain & growth
  - Age appropriate feeding behaviors

**Barriers to Nutrition**

- Fluid restriction
- Hyperglycemia
- NPO (extubation, other procedures)
- Impaired renal function
- Hemodynamically unstable
- Electrolyte imbalances

**Special Considerations & Nutrition-Related Complications**

- **ECMO =** Extracorporeal membranous oxygenation
  - Dialysis for the lungs
  - Removes the blood from the ___________ and oxygenates it before putting it back into the body
  - May be needed after major heart surgery to allow the heart to become stronger
- **Chylothorax:** accumulation of ____________ which transports long-chain triglycerides in the lymphatic system after damage to thoracic duct
  - Injury to thoracic duct during surgery
  - Elevated central venous pressure post-surgery

**Nutrition Management**

- **Infants:** high MCT formula (Enfaport)
- **Older children:** very low-fat (< 10 g/day) diet
  - ___________ only in extreme cases
- **Necrotizing enterocolitis (NEC):** Bowel compromise and potential necrosis in neonatal period
  - Poor gut _________________ due to diastolic flow reversal (single ventricle defects, i.e. HLHS, tricuspid atresia)
  - Deep hypothermia during cardiopulmonary bypass (CPB)
  - Ischemia/reperfusion injury associated with CPB
  - _________________ response to CHD and CPB

**Nutrition Management**

- Adequate TPN if feeds are held
- EBM or hypoallergenic standard infant formula when feeds restarted; advance conservatively
**Case Study**

- 4 month old, full-term male with VSD who presents with SOB, FTT, and feeding difficulties.

**Nutrition Assessment:**

- **Growth Parameters**
  1. Wt: 4.5 kg (<2\textsuperscript{nd} %-
  ile)
  2. Length: 59 cm (<2\textsuperscript{nd} %-
  ile)
  3. HC: 40.5 cm (10-25\textsuperscript{th} %-
  ile)
  4. Wt-for-ht: 5.5 kg
  5. Wt-age: 1 month old

- **Intake History**
  1. Breast feeds PO ad lib 3-4 x per day; hold feeds if respiratory rate exceeds 50
  2. Supplementation with Enfamil 20 kcal/oz: 1.5-2 oz/feed q 3-4 hours
  3. Mom reports infant feeds very slowly and for long periods of time (takes > 30
     minutes to feed); and appears out of breath and tired at the end of feeding

- **Labs:** WNL
- **Meds:** Lasix, fentanyl, warfarin

**Estimated Nutritional Needs (Catch-Up Growth)**

- 1. Calories:
- 2. Protein:

**Recommendations:**

- 1. ________________________________
- 2. ________________________________
- 3. ________________________________

**Goals:**

- 1. ________________________________
- 2. ________________________________

**Monitor/Evaluation:**

- 1. ________________________________
- 2. ________________________________
- 3. ________________________________

**Summary**

- CHDs are the most common birth defect responsible for infant death, while still considered rare
- Defects range from simple to complex, with single ventricular physiology being the most complex
- CHD infants generally have higher energy & nutrient needs, and feeding difficulties & growth failure are life-long issues
- Nutrition management plays key role in optimizing patients outcomes and chances for survival post-operatively
  - Promote wound healing
  - Promote catch-up growth and development
  - Reduce risk of infection
- Many factors negatively impact their ability to consume, absorb, or utilize nutrient
References:

15. https://www.littlehearts.org/Content/CHD_Resources.asp
18. http://www.texasheartinstitute.org/HIC/Topics/Cond/CongenitalHeartDisease.cfm