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
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 = “existing at birth”
  - **Definition**: problem with the heart’s structure and function that is present at birth
  - Congenital heart “defect” 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 ~40,000 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
TRUE OR FALSE

CHDs are 60 times more prevalent than childhood cancer
Etiology

80% Unknown
Multifactorial inheritance with association of both genetic & environmental 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
    - Diabetes*
  - Viral infections (i.e. rubella)

20% Known

- **Chromosome abnormalities (8-10%)**
  - Down syndrome (40-50%)
  - Trisomy 18
  - Williams syndrome

- **Mendelian Syndromes (3-5%)**
  - Noonan syndrome (80-90%)

- **Non-syndromal single gene disorders**
  - > 30 genes have been linked to CHDs
  - Transcription factor genes most common
    - **NKX2-5:** TOF, HLHS, TGA, VSD, ASD, DORV
CHD Risk Factor: Maternal Diabetes

- Five-fold increased risk of CHDs in infants from mothers with pregestational diabetes
- Maternal glucose control affects cardiogenesis at a very early stage of fetal development (< 7 weeks gestation)

- Compared the incidence & pattern of CHD in infants of T1DM mothers (n=308) with infants of healthy non-diabetic mothers (n=1879)
  - Odds ratio for certain types of CHDs ranged from 2.52 (Anomalies of pulmonary artery) to 18.24 (Single ventricle physiology) (p < 0.05)
  - Increased incidence of TGA, truncus arterosus, visceral heterotaxia, and single ventricle defects in infants of T1DM mothers

- Compared T1DM mothers who had infants with CHD with T1DM mothers who had healthy infants
  - Average 1st trimester HgbA1C concentration was 9.79% vs. 8.14% (p < 0.05)
Diagnosis

- Prenatally
  - Fetal echocardiogram
- Postnatally (shortly after birth)
  - Physical exam
    - **Signs & Symptoms:** Heart murmur, cyanosis, shortness of breath, rapid breathing, failure to thrive (FTT), signs of heart failure
  - Echocardiography (Echo)*
  - Cardiac Catheterization*
  - Electrocardiogram (EKG)
  - Chest X-Ray
  - Pulse oximetry
TRUE OR FALSE

CHDs cause chest pain or other painful symptoms
Heart Anatomy & Physiology
Healthy Heart Anatomy & Function

RIGHT SIDE = LOWER PRESSURE

LEFT SIDE = HIGHER PRESSURE

http://www.youtube.com/watch?v=JA0Wb3gc4mE&feature=player_embedded
**Fetal Heart**

**Foramen ovale**
- 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…

- Foramen ovale & ductus arteriosus begin to close
- Shunts sometimes remain open or can be kept open (patent)
  - **BAD:** if infant otherwise healthy
  - **GOOD:** if infant has CHD, as it can allow mixed blood to circulate until defect repaired
    - Prostoglandin E1
Types of Defects: Medical & Surgical Management
## Common Types of CHDs

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

Most common form of CHD
- 15-20% of all CHD cases

Description
- Hole in the wall (septum) separating the two lower chambers of the heart
  - Allows blood to mix
  - Red blood from high pressured left side → low pressured right side
  - Extra blood pumped into pulmonary arteries increases work of heart & lungs → congested lungs
- Holes can vary in size
  - Small VSD- asymptomatic with normal growth & development
  - Large VSD- pulmonary HTN & CHF with delayed growth & development
VSD: Medical Management

**Small VSD**
- Observation only
- Often close on their own
- Infective (bacterial) endocarditis prophylaxis
  - Antibiotics used in prevention of an infection caused by bacteria that enter the bloodstream and settle in the lining of the heart, heart valve, or blood vessel

**Large VSD**
- Diuretics
- Frequent feedings of caloric dense formulas
- Infective endocarditis prophylaxis
VSD: Surgical Repair

- **Small VSD:**
  - Surgery usually not indicated unless another defect is involved

- **Moderate to large VSD:**
  - Surgery indicated if CHF or growth failure cannot be improved with medical management
  - Hole should be closed surgically within first 6 months of age

- **Temporary repair**
  - Child very small or very sick
  - Pulmonary artery band 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
VSD: Surgical Management

Temporary Repair

Permanent Repair

VSD Patch (To Close Opening Between Ventricles)
Tetrology of Fallot (TOF)

8-10% of all CHD cases

Four Defects:
1. Ventricular Septal Defect
2. Pulmonary Stenosis
3. Overriding aorta
4. Right ventricular hypertrophy

- Most common form of CHD that produces cyanosis
- Growth retardation can occur
TOF: Medical Management

- **Hypercyanotic spells**
  - Severe cyanosis + paleness, tachypnea, altered level of consciousness
  - Occur after crying or after feeding
- **Treatment:**
  - Knee-chest positioning
  - Morphine sulfate
  - Oxygen
  - IVF
  - Sodium bicarbonate
  - Phenylephrine
  - Propranolol
- **Infective endocarditis prophylaxis**
  **Surgical repair indicated for all patients**
TOF: Surgical Repair

**Blalock-Taussig Shunt (BT-Shunt)**
- Temporary
- Performed if infant too sick or pulmonary arteries too small for corrective procedure
- Small tube from subclavian artery into pulmonary artery
  - Allows blood to enter lungs
- *Something the Lord Made*

**Complete Repair**
- 1-6 months of age
  - Infants who had palliative shunt may have complete repair 6-12 months of age
- VSD closed with patch
  - Allows oxygen poor blood to flow into lungs
- Incision across pulmonary annulus + enlargement with wide patch
  - Relieves pulmonary narrowing
TOF: Surgical Management

Temporary Repair

- Modified Blalock-Taussig Shunt
- Pulmonary Stenosis (narrowing)
- Ventricular Septal Defect

Complete Repair

- Patch to Enlarge Narrowed Pathway from RV to PA
- VSD Patch (To Close Opening Between Ventricles)
Transposition of the Great Arteries (TGA)

5% of all CHD cases

Description:
- Pulmonary artery & aorta are in opposite positions
  - Aorta carrying oxygen poor blood to body
  - Pulmonary artery carrying oxygen rich blood to lungs
- ASD, VSD, & PDA necessary for survival
- Moderate to severe cyanosis
TGA: Medical & Surgical Management

**Medical**
- Prostaglandin E1 infusion to keep PDA open
- Correction of metabolic acidosis
- Diuretics
- Frequent feedings of caloric dense formula

**Surgical**
- Arterial Switch
  - Aorta and pulmonary artery divided
  - Aorta and pulmonary artery moved to proper position
- Coronary arteries
  - Removed from aortic trunk & attached to pulmonary trunk
  - Supply heart with oxygenated blood
- Hole between VSD closed with a patch or stitches
- PDA tied off and/or cut
TGA: Surgical Management Cont.

Arterial Switch Operation Step 3

LA: Left Atrium  RA: Right Atrium  LV: Left Ventricle  RV: Right Ventricle

- Oxygen-rich Blood
- Oxygen-poor Blood
Complex Single Ventricular Defects
Hypoplastic Left Heart Syndrome (HLHS)

- 1% of all CHD cases
- Most common cause of death from CHD in 1st month of life

**Description:**
- Left ventricle underdevelopment (left ventricular hypoplasia)
- Mitral valve not formed (mitral atresia) or undeveloped (mitral stenosis)
- Aortic valve not formed (aortic atresia) or underdeveloped (aortic stenosis)
- Ascending aorta underdeveloped & narrowed (CoA)
- ASD

Become extremely ill within 1st few hours-days of life
HLHS: Medical Management

• Prostaglandin E1 infusion to keep PDA open
• Correction of metabolic acidosis
• Infective endocarditis prophylaxis
• Frequent feedings of caloric dense formula
Tricuspid Atresia

1-3% of all CHD cases

Description:
- No tricuspid valve
- Small right ventricle
- ASD & VSD
- PS

Allows for blue blood in the right atrium to flow through ASD→ left atrium. The mixed blood→left ventricle→aorta→body

Smaller 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 3 days of life
• Blalock-Taussig Shunt (BT Shunt)
  • 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 removed
• Atrial septum removed
• Narrowing aorta opened

Stage II: Bidirectional Glenn Procedure
6 months of age
• Shunt from Stage I removed
• Blue blood from head, neck, and upper body directed to the right pulmonary artery through superior vena cava
Surgical Repair: Stage I & II

Stage I - Norwood
- Blalock-Taussig Shunt
- Surgically Enlarged Atrial Septal Defect
- Homograft Patch (To Reconstruct Aorta)
- Homograft Patch on PA

Stage II - Bidirectional Glenn
- Divided Blalock-Taussig Shunt
- Superior Vena Cava Attached to Right PA
- Left PA

LA: Left Atrium  RA: Right Atrium  LV: Left Ventricle  RV: Right Ventricle
- Oxygen-rich Blood
- Oxygen-poor Blood
- Mixed Blood
HLHS & Tricuspid Atresia: Surgical Repair

Stage III: Fontan Procedure

2-4 years of age

- End to end anastomosis of inferior vena cava to right pulmonary artery through tunnel
  - Allows oxygen poor blood to be sent to lungs
  - Tube placed through right atrium & connected to right pulmonary artery
    - Allows rerouting of oxygen poor blood to the lungs
    - Heart remains available to receive oxygenated blood from lungs & pump to body
  - Holes (fenestrations) placed in tube
    - Allow heart and body to adjust

![Diagram of Stage III Extracardiac Fenestrated Fontan](image)
Nutrition Management of CHD
Nutrition Indication

- Malnutrition & FTT have long been recognized as common consequences of CHDs
- Infants with cyanotic, including the complex single ventricular physiology defects are put at a higher risk for malnutrition
- Adequate nutrition is essential for growth, wound healing, and immune function
Poor Nutrition & Growth Failure

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

- Measured oxygen consumption with respiratory mass spectrometry
- Found that hypermetabolic response is present within the first 72 hours after Norwood operation
Causes Malnutrition in CHD Patients

- Inadequate intake
  - 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)

- Increased 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 (euvolemic weight)
- **Length**
- **HC**
- **Wt-for-length**
- **Assess for** for trends on appropriate growth chart
  - Wt-for-age <3\textsuperscript{rd} %-ile
  - Ht-for-age < 3\textsuperscript{rd} %-ile
  - Wt-for-length <3\textsuperscript{rd} % 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>
</tr>
<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
  - Breast fed infants
  - Low formula intake
Nutrition Interventions

• Parenteral nutrition
  • Limited because of fluid restriction & medication drips in ICU
  • Concentrate medication drips to maximize fluid availability
  • Consider extra zinc
    • Additional 100 mcg/kg for infants <2.5 kg or 50 mcg/kg infants with open chest

• Enteral nutrition
  • Trophic feeds working up to 24 hour continuous
  • 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
  • Continuous to bolus or oral intake + nocturnal (50% estimated needs over 12 hours) – Acute Setting
    • Or PO with NG-tube back-up
  • Intermittent feeds
    • Every 3-4 hours
• Oral nutrition
  • Transition when clinically feasible after extubation in ICU
  • Approval by SLP

• Home nutrition
  • Ensure family knows recipe & appropriate mixing procedures
  • Written intake & weight gain goals provided with mixing instructions
  • Ensure adequate formula supply (WIC?)
  • 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:
  • Provide adequate nutrition to meet patient’s needs until surgery

• Post-operative critical care setting:
  • 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 body 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 chyle 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
  - TPN only in extreme cases
• Necrotizing enterocolitis (NEC)
  • Bowel compromise and potential necrosis in neonatal period
    • Poor gut perfusion 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
    • Proinflammatory response to CHD and CPB

• Nutrition Management
  • Adequate TPN if feeds are held
  • Use expressed breast milk 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
  • Wt: 4.5 kg (<2\textsuperscript{nd} %-ile)
  • Length: 59 cm (<2\textsuperscript{nd} %-ile)
  • HC: 40.5 cm (10-25\textsuperscript{th} %-ile)
  • Wt-for-ht: 5.5 kg
  • Wt-age: 1 month old

• Intake History
  • Breast feeds PO ad lib 3-4 x per day; hold feeds if respiratory rate exceeds 50
  • Supplementation with Enfamil 20 kcal/oz: 1.5-2 oz/feeding q 3-4 hours
  • 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
Case Study Cont.

- Labs: WNL
- Medications: Lasix
- Estimated Nutritional Needs (Catch-up Growth):

  \[ \text{Kcal/kg} = \frac{\text{kcal/kg for wt-age x IBW}}{\text{current wt}} \quad \text{protein g/kg} = \frac{\text{kcal/kg} \times 0.08-0.1}{4 \text{ kcal/g}} \]

<table>
<thead>
<tr>
<th>Age</th>
<th>Energy (kcal/kg)</th>
<th>Protein (g/kg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-3 mos.</td>
<td>95-107</td>
<td>1.5-2</td>
</tr>
<tr>
<td>4-6 mos.</td>
<td>82-83</td>
<td>1.5-2</td>
</tr>
</tbody>
</table>

- Calorie: 120-130 kcal/kg
- Protein: 2.5-3 grams/kg
Intervention

• **Recommendations:**
  - Increase formula caloric density to 24 kcal/oz (13 oz Enfamil Concentrate + 9 oz water)
  - Limit infant to 5-10 minutes per breast, supplement with higher calorie formula
  - Feed q 3 hours with goal of 2.5 oz/feeding = provides 133 ml/kg, 107 kcal/kg, 2.3 g protein/kg
  - After 2-3 days, increase to 27 kcal/oz (13 oz Enfamil Concentrate + 6 oz water) = provides 133 ml/kg, 120 kcal/kg, 2.6 g protein/kg

• **Goals:**
  1. Nutritional intake of breast milk and concentrated formula to meet >90% of catch-up calorie needs within the next 48-72 hrs
  2. Weight gain of 25-35 grams/day

**Monitor & Evaluation**

• GI tolerance & adequacy of formula intake
• Appropriate weight gain
• Need for supplemental NG feedings
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 nutrients
References

• Chamberlain RS. *Surgical Repairs of Congenital Heart Defects*. 2002; Pritchett and Hull Associates, Inc.

  - [http://www.lpch.org/DiseaseHealthInfo/HealthLibrary/cardiac/chd.html](http://www.lpch.org/DiseaseHealthInfo/HealthLibrary/cardiac/chd.html)
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