Cardiology Review

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Session Summary

This presentation will provide an overview of cyanotic, acyanotic, obstructive, and other congenital heart defects. There will also be a brief discussion regarding tachy arrhythmias, brady arrhythmias, and pulseless arrests, as well as compensated, decompensated, and irreversible shock.

Session Objectives

Upon completion of this presentation, the participant will:

- understand fetal circulation;
- understand assessment of the cardiac system;
- be able to discuss tachy and brady arrhythmias
- be able to recognize congenital heart disease, including
  - acyanotic lesions
  - obstructive lesions
  - cyanotic lesions

Test Questions

1. Infants with Tetralogy of Fallot who experience “hypoxic tet spells” are placed in knee chest position in order to:
   a. Increase the left to right shunting
   b. Increase the systemic vascular resistance
   c. Decrease the systemic vascular resistance

2. A 3-month-old with Down syndrome exhibits poor weight gain, tachypnea and grade 2/6 murmur. CX reveals cardiomegaly. Of the following, which is the MOST likely diagnosis?
   a. Coarctation of the aorta
   b. Complete atrioventricular septal defect
   c. Perimembranous VSD
3. A pan systolic murmur is noted on exam and the infant also has bilateral ventricular dilatation on ECHO and increased pulmonary vascularity on CXR. The likely etiology is:
   a. Large PDA
   b. Large VSD
   c. Pulmonary stenosis

4. A 28-week old infant on DOL 5 has a symptomatic PDA, he may experience which of the following symptoms:
   a. Oliguria
   b. Hypertension
   c. Weak radial pulses

5. Pulmonary vascularity is decreased in all of the below except:
   a. Tetralogy of Fallot
   b. TAPVR
   c. Tricuspid atresia

References


Session Outline

See presentation handout on the following pages.
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Embryology
- Begins developing between the 3rd to 7th week gestation with completion at 10 weeks
- 1st organ to function in utero
- Fetal heartbeat can be heard at 6wks
- Starts as long structure with 2 tubes

Embryology
- Elongates & Twists to Right
- Separates the Atria and Ventricles
- Formation of Valves
  - Mitral & Tricuspid
  - Great Vessel Formation
  - Aorta, Pulmonary Arteries and Vein
The Heart

- The heart consists of four chambers
- Valves that open and close to allow blood to enter and leave these vessels and chambers.
- S1 = Closing of TV and MV (AV valves)
- S2 = Closing of AV and PV (semilunar valves)

Fetal Circulation

Things unique to Fetal Circulation
- Foramen Ovale
- Ductus Arteriosus
- Ductus Venosus
- Placenta
- Umbilical Vessels
- Dominant Right Heart – pumping 2/3 of combined ventricular output

Parallel Circulation

High pulmonary vascular resistance

Low resistance placenta
**Extrauterine Cardiovascular Changes**

- Pulmonary Vascular resistance (PVR)
- Systemic Vascular Resistance (SVR)
- Ductus Arteriosus closes
  - Oxygen and PGE2 lessens
- Ductus Venosus closes
- Foramen Ovale closes

**Cardiac Output**

Cardiac Output (CO)

- The volume of blood ejected by the heart in 1 minute
- CO = stroke volume x heart rate
  - 200 ml/kg/min
  - Neonates increase HR in response to low CO

**Stroke Volume (SV)** is the difference between the ventricular end diastolic volume and the end systolic volume (1.5ml/kg)

- SV is affected by preload, contractility and afterload

**Conduction System**

Preload and Afterload

- Preload: volume entering ventricles
- Afterload: resistance left ventricle must overcome to circulate blood

Contractility (inotropy)

- The speed of ventricular contraction
- Contractility is affected by Catecholamine - increase contractility
- Acidosis, hypoxia... decrease contractility
Blood pressure

- Measurement of the pressure on the walls of the vessels as blood is pumped
- Determined by
  - Peripheral vascular resistance
  - Cardiac output
- Systolic: end of each heart contraction
- Diastolic: immediately before each contraction.
- Pulse pressure
  - Widened= PDA (blood runs off into pulmonary artery during diastole)
  - Narrow= pericardial tamponade, intravascular depletion and ECMO pt

Shock

- State of inadequate circulatory blood volume
- Results in decreased perfusion and oxygenation to tissues → lactic acidosis → heart failure
- Hypovolemic: loss of volume
  - Acute blood loss, pleural effusion, skin disruption
- Cardiogenic
  - Heart fails due to tamponade, tension pneumothorax, CHD
- Distributive- sepsis, body release toxins

Assessment of Cardiac

- Physical Assessment & History
- Observation
- Auscultation
- Palpation : PMI
- Diagnostics
  - EKG
  - Chest XRAY
  - Hyperoxia Test
  - Pre and Post Ductal Saturations
  - Echocardiogram

Sounds

- S1: closure of MV/TV
- S2: closure of Ao/pulmonic valve. Should be split!
- S3: extra sound may be normal in newborn related to ventricle filling.
- S4: rare, myocardial disease
Murmur

- Turbulent blood flow
- Innocent versus pathologic murmurs
  - FT infant may have murmur @24-48hr due to PDA closing → benign
- Location
- Intensity (1-6)
- Radiation
- Timing
  - Continuous: pathologic
  - Systole: usually benign
  - Diastole: PATHOLOGIC

Murmur Types

<table>
<thead>
<tr>
<th>Condition</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>Harsh pansystolic LLSB</td>
</tr>
<tr>
<td>PDA</td>
<td>Continuous machinery</td>
</tr>
<tr>
<td>Truncus Arteriosus</td>
<td>Harsh systolic, single S2</td>
</tr>
<tr>
<td>Valvular Stenosis</td>
<td>Loud ejection click</td>
</tr>
<tr>
<td>PPS</td>
<td>Radiates to axilla and back</td>
</tr>
<tr>
<td>qTIA</td>
<td></td>
</tr>
</tbody>
</table>

Common Electrolyte Disturbances

- Hyperkalemia= peaked T waves
- Hypokalemia= prominent U waves
- Hypercalcemia= short Qt interval
- Hypocalcemia= prolonged Qt interval

Dysrhythmias

- Brady arrhythmias
  - Sinus Bradycardia
  - Heart Block
- Tachyarrhythmia
  - Sinus tachycardia
  - Supraventricular Tachycardia
Supraventricular tachycardia (SVT)

- Heart rate sustained at > 220 bpm
- Treatment
  - Ice
  - Vagal maneuver
  - Adenosine: rapid infusion 1-2 sec followed by NS
- Cardioversion may be needed

Congenital Heart Disease

- <1% of all newborns,
- Prenatal Dx in about 50-80% of the time
- 30% of patients with chromosomal anomalies have CHD
- Multifactorial causes (90% of cases)
- Biggest risk factor = Family History of CHD

Incidence of Defects

<table>
<thead>
<tr>
<th>Acyanotic Heart Disease</th>
<th>Obstructive Lesions</th>
<th>Cyanotic Heart Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect (ASD)</td>
<td>Aortic stenosis (AS)</td>
<td>Transposition of the great arteries (TGA)</td>
</tr>
<tr>
<td>Ventricular septal defect (VSD)</td>
<td>Pulmonary stenosis (PS)</td>
<td>Tetralogy of fallot (TOF)</td>
</tr>
<tr>
<td>Patent ductus arteriosus (PDA)</td>
<td>Coarctation of the aorta (CoA)</td>
<td>Total anomalous pulmonary venous return (TAPVR)</td>
</tr>
<tr>
<td>Atrioventricular Canal</td>
<td>Truncus arteriosus (TA)</td>
<td>Tricuspid atresia</td>
</tr>
<tr>
<td>Pulmonary Stenosis (5-6%)</td>
<td></td>
<td>Pulmonary atresia</td>
</tr>
<tr>
<td>Tetralogy of Fallot (8-16%)</td>
<td></td>
<td>Hypoplastic left heart (HLHS)</td>
</tr>
<tr>
<td>VSD 20-25%</td>
<td></td>
<td>Ebstein's anomaly</td>
</tr>
</tbody>
</table>

Surgical repair is now successful and routine, with an overall mortality of < 4% nationally.
I. Acyanotic Heart Defects

- Left to Right shunt
- Cardiomegaly
- Increased pulmonary vascular markings
- CHF when PVR drops
- Pulmonary over circulation

Patent Ductus Arteriosus

- Stealing effect from systemic circulation & the increased pulmonary blood flow

<table>
<thead>
<tr>
<th>Hypotension</th>
<th>Oliguria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peripheral vasoconstriction</td>
<td>Metabolic acidosis</td>
</tr>
<tr>
<td>Hyper dynamic precordium</td>
<td>Widened pulse pressure</td>
</tr>
<tr>
<td>Pulmonary edema; CHF</td>
<td>Respiratory distress</td>
</tr>
<tr>
<td>Continuous Loud machinery murmur</td>
<td></td>
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</tbody>
</table>

Management of PDA:

- Term Infant
- Coiling closure at 3 months
- Preterm infant
- Conservative: Fluid restriction & Diuretics
- Hemodynamically significant PDA
  - Indomethacin/ibuprofen
  - Surgical Ligation

ASD

Hemodynamics
- Oxygenated blood from the left atrium is shunted to the right atrium then into the right ventricle and back to the lungs
- Rarely get CHF
- Systolic ejection murmur
- The increased volume and work of the RV leads to RV hypertrophy

Management
- Treat CHF
- Intractable CHF: surgical repair is necessary
VSD

- Most common CHD
- FLOW: L→R shunting via ventricular septum causing increased pulmonary blood flow
- Harsh pan systolic (holosystolic) murmur
- Urgency depends on size of VSD
  - Small: usually resolves by itself
  - Large: causes CHF in 6-8 weeks

Management of VSD

Mild VSD
  - Fluid restriction, Diuretics, Digoxin
Moderate to severe VSD
  - pulmonary banding, suturing or patching the of the defect

Atrioventricular Canal

- Abnormal development of the endocardial cushion
- Common in Down syndrome

Complete AV Canal | Partial AV canal
--- | ---
1 valve | Mitral regurgitation
VSD

- Treatment
  - PA Banding
  - AD, VSD closure and reconstruction of valve

Congestive Heart Failure

- The heart no longer able to pump adequate amount of blood to meet the needs of the body
- Results in systemic and venous congestion
- Can be caused by such things as CHD, infection, severe anemia, birth asphyxia and dysrhythmias

- Tachycardia, tachypnea,
- sudden weight gain or poor weight gain
- Poor feeding
- Hepatomegaly
- Arrhythmias
- Cardiomegaly

Eisenmenger's Syndrome
II. Lesions Obstructing Blood Flow

- Pulmonary Stenosis (PS)
- Aortic Stenosis (AS)
- Coarctation of the Aorta (CoAo)

**Pulmonary Stenosis**

- Obstruction of blood flow to pulmonary bed
- May be valvular (90%), subvalvular, or supravalvular
- Usually associated with large VSD
- Sudden death is possible in more severe PS (Critical PS)
- Harsh Systolic ejection murmur

**Aortic Stenosis**

- Obstruction of Blood flow to body

**Types:**
- Valvular
- Supravalvular: usually associated with William’s Syndrome
- Subvalvular
- Peripheral pulses are weak and thready
- Narrow pulse pressure is present in severe AS
Coarctation of the Aorta

- Strong pulses in upper extremities compared to lower extremities
- Severe cases may have LV pressure overload
- Loud S3 gallop is usually present

<table>
<thead>
<tr>
<th>Mild</th>
<th>Headaches</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mod</td>
<td>CHF</td>
</tr>
<tr>
<td>Severe</td>
<td>Shock</td>
</tr>
</tbody>
</table>

Management

- Treat the heart failure (digoxin & Lasix)
- Prostin
- Surgical Intervention
  - Anastomosis
  - Grafting
  - Balloon angioplasty

III. Cyanotic Heart Lesions

There has to be a RIGHT to left shunt to cause CYANOSIS

ST’s
- Transposition of Great Arteries
- Tetralogy of Fallot
- Tricuspid Atresia
- Truncus Arteriosus
- TAPVR
- Ebstein’s Anomaly
- Single Ventricle
- Pulmonary Atresia

Transposition of Great Arteries

- The aorta arises from the RV and the PA arises from the LV
- Hyoxia and cyanosis
- Survival is dependent on the communication between the 2 “parallel” circuits
  - VSD, ASD, PDA
- The amount of blood flows into and out of the pulmonary circulation must be equal
- Egg on string CXR
- Most common cyanotic lesion in NEWBORN period
**TGA Management**

- Prostin dependent
- Balloon septostomy

**TGA - XRAY**

- Egg on a string

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**Tetralogy of Fallot**

- **MOST COMMON CYANOTIC HEAR DISEASE**
- Includes 4 abnormalities:
  1) RVOT obstruction
  2) RVH
  3) VSD
  4) overriding of the aorta
- Severity depends on pulmonary stenosis degree

**Management of TOF**

- Treat CHF
- Prostin
- Surgical
  - Blalock-Taussig Shunt
  - Total surgical correction 3-6 months

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**Cyanosis** | Sats 75-85%
---|---
Right shaped heart
Tet (Hypoxic) Spells | Knee chest, morphine, O2, beta blocker
Murmur SEM
**TOF- XRAY**

Booth shape heart on X-RAY

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**Total Anomalous Pulmonary Venous Return**

Excuse me, what atrium was I suppose to connect to????

The pulmonary veins drain oxygenated blood directly or indirectly into the right atrium instead of the left atrium

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**TAPVR**

- Obstructive $\rightarrow$ cyanosis due to R$\rightarrow$ L mixing at ASD level
- Nonobstructive $\rightarrow$ CHF
- XRAY: Snowman Heart
- Surgical Correction: The pulmonary veins are reconnected to the left atrium and the ASD is closed. Performed within the first weeks after the child’s birth

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**TAPVR XRAY**

- Snowman
Truncus Arteriosus

- Only a single arterial trunk leaves the heart – supplies pulmonary, systemic and coronary circulation
- Large VSD is always present
- Cyanosis varies and depends on the amount of Pulmonary blood flow
- Associated with DiGeorge syndrome

Truncus Arteriosus Management

- Rastelli Operation
- Conduit is placed from the Right Ventricle to the Pulmonary Artery

Tricuspid Atresia

- Tricuspid valve is absent, RV and PA are Hypoplastic with decreased PBF
- 1-2% of all CHD
- ASD, VSD, or PDA are necessary for survival
- Single S2

Management of Tricuspid Atresia

- B-T Shunt
- Glenn
- Fontan
Pulmonary Atresia

- Communication at the atrial level is necessary for life
- These patients are duct dependent
- Single S2

Ebstein’s Anomaly

- Extremely large heart
- Abnormal development of the tricuspid valve
- Weak TV\(\rightarrow\)PVR\(\rightarrow\) Cyanosis

Ebstein’s Anomaly

- Treatment
  - Prostin
  - Treat heart failure
  - Pulmonary artery banding
  - Surgery

Hypoplastic Left Heart Syndrome

- 1 – 2% of all CHD

<table>
<thead>
<tr>
<th>Left Heart Underdeveloped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. LV Hypoplastic</td>
</tr>
<tr>
<td>2. Aortic Valve atresia or stenosis</td>
</tr>
<tr>
<td>3. Mitral valve atresia or stenosis</td>
</tr>
<tr>
<td>4. Aortic arch Hypoplastic</td>
</tr>
</tbody>
</table>

- Must have PFO/ASD – allow LA to receive oxygenated blood
- PDA dependent to ensure systemic circulation
HLHS

Presentation- HLHS
Cyanosis
Cardiogenic shock with PDA closure
Signs an symptoms of CHF
Poor perfusion : pulmonary over-circulation
Sever metabolic acidosis

Hypoplastic Left Heart Syndrome

Medical Management:
• Compassionate Care
• PGE1 infusion
• Must balance circuit of pulmonary and systemic circulation
• Keep sats 75 to 85%
• Avoid excessive pulmonary vasodilation
  → PBF → CHF

Surgical Management:
• Norwood: rebuild the tiny ascending aorta
• Stage II: Glenn Operation
• State III: Fontan procedure
• Cardiac Transplant

Rule of 4’s in Cardiac Patient

• pH= should be 7.40
  • Acidosis= lactic acid build up= muscle fatigue= bad cardiac contractility and function

• CO2= in the 40’s
  • respiratory acidosis

• Hematocrit= at least 40
  • Need higher Oxygen carrying capacity

• Potassium= level in the 4 range
  • Na/K pump regulates influx of electrical impulses to regulate heart muscle contraction.
  • Hyperkalemia can create lethal arrythmias

Maternal Diabetes | Hypertrophic cardiomyopathy, TGA, VSD
Maternal Lupus | Heart Block
Maternal Alcohol Abuse | TGF
Maternal Rubella | PDA, PPS
Down’s syndrome | 40% have CHD, AVC, VSD most common
Turner syndrome | Coarctation of the aorta
DiGeorge Syndrome | Truncus arteriosus