Neonatal Cardiology Review

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Session Summary

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

Session Objectives

Upon completion of this presentation, the participant will be able to:

▪ understand fetal circulation;
▪ understand assessment of the cardiac system;
▪ discuss tachy and brady arrhythmias;
  - 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


Cardiology Review
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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

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

Cardiac Output

Cardiac Output (CO)
- The volume of blood ejected by the heart in 1 minute
- \( CO = \text{stroke volume} \times \text{heart rate} \)
- 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

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: increases contractility
Acidosis, hypoxia: decreases contractility

Conduction System
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)
  - Narrowed: 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

<table>
<thead>
<tr>
<th>Compensated</th>
<th>Uncompensated</th>
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<tbody>
<tr>
<td>BP still normal</td>
<td>Hypotensive</td>
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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 and Heart Sounds

<table>
<thead>
<tr>
<th>VSD</th>
<th>Harsh pansystolic LLSB</th>
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<tbody>
<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 aorta and back</td>
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</tbody>
</table>
Common Electrolyte Disturbances
- Hyperkalemia: peaked T waves, widened QRS
- Hypokalemia: prominent U waves
- Hypercalcemia: short QT interval
- Hypocalcemia: prolonged QT interval

Dysrhythmias
- Brady arrhythmias
  - Sinus Bradycardia
  - Heart Block
- Tachyarrhythmia
  - Sinus tachycardia
  - Supraventricular Tachycardia

SVT
Supraventricular tachycardia (SVT)
- Heart rate sustained at > 220bpm
- Dual AV node pathway
- 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
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>
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<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>
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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

- 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 or ASD
- 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

Pulmonary Stenosis

- Usually normal heart size.
- Severe pulmonary stenosis will result in decreased pulmonary blood flow
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

| Mild headaches |
| Mod | CHF |
| severe | Shock |

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

5T's
- Transposition of Great Arteries
- Tetralogy of Fallot
- Tricuspid Atresia
- Truncus Arteriosus
- TAPVR
- Ebsteins Anomaly
- Single Ventricle
- Pulmonary Atresia

Transposition of Great Arteries

- The aorta arises from the RV and the PA arises from the LV
- Hypoxia 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: PDA is needed for systemic perfusion
- Balloon septostomy

Arterial switch

great vessels are switched

TGA - XRAY

- Egg on a string
**Tetralogy of Fallot**

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

<table>
<thead>
<tr>
<th>Cyanosis</th>
<th>Sats 75-85%</th>
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<tbody>
<tr>
<td>Right to left shunting of blood at the VSD</td>
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</table>

**Management of TOF**

- Treat CHF
- Prostin
- Surgical
  - Blalock-Taussig Shunt: subclavian to pulmonary artery
  - Total surgical correction 3-6 months

**Total Anomalous Pulmonary Venous Return**

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

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

**TAPVR**

- Obstructive → cyanosis due to R→L mixing at atrial level
- Nonobstructive → 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

**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

Need to get blood into pulmonary artery circulation

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 → PVR → Cyanosis
Ebstein's Anomaly

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

Hypoplastic Left Heart Syndrome

• 1 – 2% of all CHD
  1. LV Hypoplastic
  2. Aortic Valve atresia or stenosis
  3. Mitral valve atresia or stenosis
  4. Aortic arch Hypoplastic

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

HLHS

Presentation- HLHS
Cyanosis
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
  \( \rightarrow PBF \rightarrow CHF \)

Surgical Management:
• Norwood: rebuild the tiny ascending aorta
• Stage II: Glenn Operation
• Stage 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
• \( CO_2 = \) 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 arrhythmias

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