Congenital Heart Defects – Critical Care

Objectives
- Cover 4 different cardiac physiologic conditions
- Discuss pre and perioperative management
  - Fluids
  - Saturations
  - Oxygen
  - Medication/drip
  - Ventilator settings
  - Other

Shunt Physiology
Ventricular Septal Defect

- Cyanosis/Systolic/Dyspnea
- Exsanguination/Monovascular
- Transient
- Membranous, paraseptal, or coronary
- Muscular
- 20% VSD
- Spontaneous closure common
Shunt Physiology
Ventricular Septal Defect

• Volume loading of the left heart.
• Shunting of blood from the left ventricle → across the VSD → across the pulmonary valve → the left heart

Shunt Physiology
Ventricular Septal Defect
Heart failure

Preoperative Management

• Fluids
• Saturations
• Oxygen
• Medications

Other

• 2/3 to ¾ maintenance IV fluids
• Normal
• Avoid
• Anticongestive meds
  • Lasix
  • Enalapril/Captopril
  • Aldactone
  • Digoxin
• NG feeds
• Intubation
Ventricular Septal Defect Surgery

- Approach from the right atrium
- Closure with prosthetic patch

Ventricular Septal Defect Post-operative Management

<table>
<thead>
<tr>
<th>Post-operative Bleeding</th>
<th>&gt; 10 ml/kg/hour</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>FFP, cryoprecipitate, platelets, Factor VII, and PRBC</td>
</tr>
<tr>
<td>Fluids</td>
<td>% maintenance to maintenance</td>
</tr>
<tr>
<td>Saturations</td>
<td>Normal</td>
</tr>
<tr>
<td>Oxygen</td>
<td>As needed</td>
</tr>
<tr>
<td>Medications/drips</td>
<td>Milrinone 0.5 to 0.75 mcg/kg/min</td>
</tr>
<tr>
<td></td>
<td>Epi 0.04 mcg/kg/min</td>
</tr>
<tr>
<td></td>
<td>Lasix drip</td>
</tr>
<tr>
<td></td>
<td>Dexmedetomidine drip</td>
</tr>
<tr>
<td>Ventilator settings</td>
<td>Extubated in the OR or soon in the ICU</td>
</tr>
<tr>
<td>Other</td>
<td>No additional anticoagulation</td>
</tr>
<tr>
<td></td>
<td>Junctional Ectopic Tachycardia (JET)</td>
</tr>
</tbody>
</table>

Ventricular Septal Defect Junctional Ectopic Tachycardia

- Narrow complex tachycardia with lack of a P wave
- Ectopic focus at the AV node
- Self limited but can cause issues with cardiac output
Junctional Ectopic Tachycardia Management

- Normothermia or patient cooling
- Correct acidosis
- Normalize electrolytes (potassium and magnesium)
- Overdrive pacing
- Anti-arrhythmics
  - Amiodarone

Ventricular Septal Defect

- Defect in the ventricular septum
- Left to right shunt abnormality
  - Volume loading of the left heart leading to congestive heart failure
- Pre-operative
  - Management of congestive heart failure symptoms
    - Fluids – IV vs oral vs NG
    - Medications
      - Take away the work of feeding
- Post-operative
  - Bleeding
  - JET
  - Extubate shortly after surgery

Shunt Physiology - Cyanotic Tetralogy of Fallot
Tetralogy of Fallot

Obligate right to left shunt at the level of the VSD
Cyanosis is usually present
Crying and feeding can worsen cyanosis

<table>
<thead>
<tr>
<th>Fluids</th>
<th>Maintenance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Saturations</td>
<td>Variable 60-90% on room air</td>
</tr>
<tr>
<td>Oxygen</td>
<td>Usually not helpful</td>
</tr>
<tr>
<td>Medications/drips</td>
<td>Prostaglandin E1 0.025 mcg/kg/min to 0.1 mcg/kg/min</td>
</tr>
<tr>
<td>Ventilator settings</td>
<td>Set spell</td>
</tr>
<tr>
<td>Other</td>
<td>Prevent introduction of air bubbles in the IV line</td>
</tr>
</tbody>
</table>

Tetralogy of Fallot
Pre-operative Management

Tetralogy of Fallot
Tet Spell

Children with Tetralogy of Fallot exhibit bluish skin during episodes of crying or feeding.

"Tet spell"
Tetralogy of Fallot
Tet Spell - management

- Calm the infant/child (calm the health care provider)
- Place infant/child in knee to chest position
  - increases the systemic vascular resistance
- Morphine
  - decrease the “spasm” of the RVOT and suppresses respiratory centers
- Oxygen
  - May reduce the PVR

Tetralogy of Fallot
Surgery – Modified BT shunt

- Patch closure of the ventricular septal defect
- Incision across the RVOT and the pulmonary valve annulus
- Patch augmentation of the RVOT and the pulmonary annulus
Tetralogy of Fallot Surgery - Homograft

- Patch closure of the ventricular septal defect (7)
- Incision made in the anterior wall of the right ventricle and a hood sewn over (5)
- Anastomosis of the homograft to the pulmonary artery and the RV hood (4)

Pulmonary homograft

Post-operative Management

<table>
<thead>
<tr>
<th>Post-operative bleeding</th>
<th>&lt;10 ml/kg/hour</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fluids</td>
<td>% maintenance to maintenance</td>
</tr>
<tr>
<td>Saturations</td>
<td>Normal</td>
</tr>
<tr>
<td>Oxygen</td>
<td>As needed</td>
</tr>
<tr>
<td>Medications/drips</td>
<td>Milrinone 0.5 to 0.75 mcg/kg/min</td>
</tr>
<tr>
<td></td>
<td>Epi 0.04 mcg/kg/min</td>
</tr>
<tr>
<td></td>
<td>Lasix drip</td>
</tr>
<tr>
<td></td>
<td>Fentanyl and Versed drip</td>
</tr>
<tr>
<td>Ventilator settings</td>
<td>Standard ventilator settings</td>
</tr>
<tr>
<td></td>
<td>On ventilator until the patient's fluid status has improved</td>
</tr>
<tr>
<td>Other</td>
<td>Anti-inflammatory medications for homografts (ibuprofen)</td>
</tr>
</tbody>
</table>
**Tetralogy of Fallot**

**Low Cardiac Output**
- The right ventricle becomes noncompliant
- Higher end diastolic pressure
- Patients will usually require a high central venous pressure to maintain forward flow and cardiac output
- Usually requires fluid bolus or higher maintenance fluid levels
- Patients will usually become fluid overloaded in the first 24-38 hours

**Pre-operative management**
- Management of cyanosis
  - Medications – prostaglandin E1
  - Tet spell
  - Surgical – BT shunt

**Post-operative management**
- Low cardiac output
  - Milrinone
  - Fluid bolus and liberal fluids
  - Longer intubation period, extubate POD 1 to 3

**Transposition of the Great Arteries**
Transposition of the Great Arteries

- Association with maternal diabetes
- Large infants
- Male predominance (60-70%)
- Severe cyanosis (saturations 40 -50%)

Transposition of the Great Arteries

Pre-operative management

<table>
<thead>
<tr>
<th>Fluids</th>
<th>Maintenance fluids</th>
</tr>
</thead>
<tbody>
<tr>
<td>Saturations</td>
<td>Severe cyanosis (saturations 40 – 50%)</td>
</tr>
<tr>
<td>Oxygen</td>
<td>Usually not helpful</td>
</tr>
<tr>
<td>Medications/Drips</td>
<td>Prostaglandin E1 0.025 - 0.1 mcg/kg/min</td>
</tr>
<tr>
<td>Ventilator settings</td>
<td>Usually not intubated but can be helpful in patients if the cyanosis is not improving</td>
</tr>
<tr>
<td>Other</td>
<td></td>
</tr>
</tbody>
</table>
Transposition of the Great Arteries
Balloon Atrial Septostomy

Surgery – Arterial Switch
Transposition of the Great Arteries
Post-operative Management

| Post-operative bleeding | > 10 ml/kg/hour  
FFP, cryoprecipitate, platelets, Factor VII |
| Fluids | % maintenance to maintenance |
| Saturations | Normal |
| Oxygen | As needed |
| Medications/drips | Milrinone 0.5 to 0.75 mcg/kg/min  
Ep 0.04 mcg/kg/min  
Dobutamine  
Lasix drip  
Sedation with fentanyl and versed |
| Ventilator settings |  
| Other | ECMO |

Open Chest ECMO

- Patient is edematous and the chest cannot be closed
- Patient need additional cardiac support
- Bypass cannulas are left in place
- ECMO for a few days until patient less edematous or the cardiac function has improved.

Transposition of the Great Arteries

- Severely cyanotic patients with no improvement in saturations with oxygen
- Patient on Prostaglandin E1 until after atrial septostomy
- Surgery performed within the first week of life
- Post-operative
  - Cardiac output
  - Post-op bleeding and fluid shifts
  - Open chest ECMO
Hypoplastic Left Heart Syndrome
Single Ventricle Physiology

- Failure of development of the left ventricle
- Hypoplasia of the ascending aorta
- Filling of the coronaries through retrograde flow in the ascending aorta
- The right ventricle is the sole pumping chamber

Hypoplastic Left Ventricle

- The right ventricle pumps blood in the main pulmonary artery
- The PDA allows for filling of the aorta (1)
- The patent foramen ovale allows left to right shunting (2)

Hypoplastic Left Heart Pre-operative Management

<table>
<thead>
<tr>
<th>Fluids</th>
<th>5% maintenance to maintenance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Saturations</td>
<td>80% on room air</td>
</tr>
<tr>
<td>Oxygen</td>
<td>Do not give</td>
</tr>
<tr>
<td>Medications/drips</td>
<td>Prostaglandin E1 0.025 to 0.1 mcg/kg/min</td>
</tr>
<tr>
<td>Ventilator settings</td>
<td>Goal of ventilation is to alter PVR to limit pulmonary blood flow</td>
</tr>
<tr>
<td></td>
<td>- FiO₂ 18%</td>
</tr>
<tr>
<td></td>
<td>- Intubate and add exogenous CO₂</td>
</tr>
<tr>
<td></td>
<td>- Ventilate with higher mean airway pressures</td>
</tr>
<tr>
<td>Other</td>
<td>Frequent labs to ensure the blood gas is normal</td>
</tr>
</tbody>
</table>
Hypoplastic Left Heart

FiO<sub>2</sub> of 18%
Goal saturation of 80%
ABG of 7.40/40/40

Hypoplastic Left Heart Norwood Procedure

- The main pulmonary artery is transected
- PDA is ligated and remaining ductal tissue is removed from the aorta

Hypoplastic Left Heart Norwood Procedure

- The diminutive aorta is filleted open to the descending aorta
- Pulmonary homograft tissue is cut to size to augment the aorta and create an anastomosis to the pulmonary artery
Hypoplastic Left Heart
Norwood Procedure

- A right Blalock Taussig shunt is connected from the innominate artery to the main pulmonary artery.

Hypoplastic Left Heart
Post-operative Management

<table>
<thead>
<tr>
<th>Bleeding</th>
<th>&gt; 10 ml/kg/min</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fluids</td>
<td>Maintenance</td>
</tr>
<tr>
<td>Saturations</td>
<td>80% on room air</td>
</tr>
<tr>
<td>Oxygen</td>
<td>Do not give</td>
</tr>
<tr>
<td>Medications/drips</td>
<td>Minimine 0.5 to 1 mcg/kg/min Epi Dobutamine Sedation</td>
</tr>
<tr>
<td>Ventilator settings</td>
<td>Goal of ventilation is to alter PVR to limit pulmonary blood flow - Higher mean airway pressures - High volume, low rates</td>
</tr>
<tr>
<td>Other</td>
<td>Frequent blood gas ECMO</td>
</tr>
</tbody>
</table>

Hypoplastic Left Heart
Post-operative

- Goal of post-operative management is delicate management of the ratio of pulmonary blood flow to the systemic blood flow.
- The high saturations is an indication of excessive pulmonary blood flow.
Hypoplastic Left Heart
Sano Modification

- Disconnection of the SVC from the heart
- End to side anastomosis of the SVC to the pulmonary artery
- Relies on passive pulmonary blood flow

Hypoplastic Left Heart
Bidirectional Glenn

- Disconnection of the SVC from the heart
- End to side anastomosis of the SVC to the pulmonary artery
- Relies on passive pulmonary blood flow
Hypoplastic Left Heart
Post-operative Glenn and Fontan

| Bleeding        | > 10 mL/kg/min  
|                | FFP, Cryoprecipitate, Platelets, Factor VII |
| Fluids         | Maintenance  
|                | Avoid dehydration |
| Saturations    | 88% on room air |
| Oxygen         | As needed  
| Medications/drips | Milrinone 0.5 to 1 mcg/kg/min |
| Ventilator settings | Extubate the patient shortly after surgery, negative pressure ventilation promotes pulmonary blood flow |
| Other          | Watch for chylothorax |

Hypoplastic Left Heart
Chylothorax

- Milky pleural fluid made up of triglyceride and lymphocytes
- Disruption of the thoracic duct with surgical placement of the Glenn and/or Fontan

Hypoplastic Left Heart
Chylothorax

- Milky pleural fluid made up of triglyceride and lymphocytes
- Disruption of the thoracic duct with surgical placement of the Glenn and/or Fontan
Hypoplastic Left Heart Chylothorax - Management

- NPO
- TPN
- Evacuation of the chylous fluid
- Diet of medium chain triglycerides
- Pleurodesis

Hypoplastic Left Heart

- Pre-operative
  - Adjustment of saturations
  - Medications – Prostaglandin, no additional oxygen
  - FiO₂ or pCO₂
  - Ventilator adjustment
  - Frequent blood gasses
- Post-operative - Norwood
  - Maintaining delicate balance between SVR and PVR
  - Maintaining cardiac output
  - ECMO
  - No oxygen

Hypoplastic Left Heart

- Post-operative – Glenn and Fontan
  - Prevent dehydration
  - Quick extubation
  - Oxygen as needed
  - Watch for chylothorax
Questions?