CONGENITAL HEART EMERGENCIES: KEEPING IT SIMPLE
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Objectives

• Focus on hypoplastic left heart
• Overview of the 3 stage palliation surgery
• Understand the complications that can occur at each stage and how to manage

Case 1

• 5 month old CC: Cyanosis
• HR: 110, RR: 42, BP: 84/Doppler, SpO2: 60%, Temp: 36.7 C
• HLHS, s/p BT shunt with LPA augmentation and PDA ligation
But what does that mean?

Breaking it all down

- Where do they get their pulmonary blood flow?

- Where do they get their systemic blood flow?

- 1V or 2V?
Hypoplastic Left Heart Syndrome: HLHS

- 960 babies each year
- Unknown cause
- The *left ventricle* is underdeveloped and too small.
- The *mitral valves* is not formed or is very small.
- The *aortic valve* is not formed or is very small.
- The ascending portion of the *aorta* is underdeveloped or is too small.
- May have ASD.

Hypoplastic Left Heart Syndrome (HLHS)
3 stage palliation surgery

1. Norwood

2. Bidirectional Glenn

3. Fontan
**Norwood (BT Shunt) – sats 75-85%**

**Post-operative changes**
- Uncontrolled PBF
- Re-constructed aortic outflow tract
- Fluid balance sensitive
- Widened pulse pressures
- Tenuous coronary circulation
- Single ventricle for all circulation

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**Norwood (Sano Shunt)**
Norwood (Sano Shunt)

- Post-operative changes
  - Direct PA communication with RV
  - Uncontrolled PBF
  - Neo-aortic reconstruction
  - Higher diastolic pressures
  - Better coronary perfusion

Case 1: Exam

- Cyanotic, no acute distress.
- Comfortable work of breathing, good air entry bilaterally.
- Right ventricular heave. Irregular rhythm with a normal S1 and single S2. There is a grade I/VI continuous murmur at the LUSB
- Abdomen is soft, nontender, and non-distended. The liver is palpated 1.5 cm below the right costal margin.
- No clubbing or edema. Warm and well perfused with 3+ pulses.
ED Presentation

Differential Diagnosis

- Respiratory infection
  - Low PVO2
- Sepsis
  - Low SVR
  - Low SVO2
  - Increased demand
- Shunt stenosis
- Anemia
Hypoxic

- Look for underlying cause
  - Pulmonary edema or infiltrate
  - Decreased pulmonary blood flow
  - Mixed venous desaturation

Chest X-ray

Pulmonary vascular congestion  Decreased pulmonary blood flow
What’s happening?

- BT Shunt narrowing
- At risk for shunt occlusion
- Can occur at any time, though more at risk when older and outgrowing shunt

What to do for Shunt Failure?

- Impending cardiac emergency, call on arrival
- Oxygen
  - Goal oxygen saturations >70%
  - Supplement oxygen to assist in maintaining oxygen saturation
- Heparin may be considered
What about oxygen therapy?

- Limit O2 therapy for cyanosis
- Maintain sats 75-85%
  - Use blended O2 with range of up to FiO2 0.4

- If sats >85% - O2 is vasodilator
  - ↓PVR → ↑PBF → Pulmonary edema and circulatory shock

What to do for resuscitation?

- Limit oxygen (remember: relative uncontrolled PBF)
- Hemoglobin
- Auscultate for murmur:
  - Continuous murmur at RUSB (? BT shunt)
  - Systolic murmur at RLSB/ LUSB (Sano shunt)
- Fluid balance:
  - Palpate liver
  - +/- rales and CXR to evaluate for CHF
  - Reverse dehydration
- Reverse acidosis
• PVR affects flow in the BT shunt
• Forward flow in both systole and diastole = continuous murmur
• Low PVR results in more flow through the shunt rather than to the body AND CORONARIES
• Higher alveolar pO2 = less flow to body
• ALWAYS TALK TO A CARDIOLOGIST BEFORE OXYGEN ADMINISTRATION TO A CHILD WITH A BT SHUNT

• PVR does not affect flow through a RV to PA shunt (aka Sano).
• Lower baseline sats but less volatile cardiac output & coronary flow
  – 2007 multi center trial with improved survival to 12 months
  – Fewer “death” spells
• Still talk to a cardiologist about oxygen

Stage 2: Bidirectional Glenn
Stage 2: Hemi Fontan

Case 2

- 10 year old CC: Diarrhea, fever, abdominal pain
- HR: 110, RR: 24, BP: 95/52, SpO2: 96%, Temp: 37.9 C
- HLHS s/p Fontan
Stage 3: Fontan

Hemi Fontan  Fontan

Case 2:

- **HEENT:** *Mucous membranes are dry.* No tonsillar exudate. Oropharynx is clear. No adenopathy.
- **Cardiovascular:** Normal rate, regular rhythm and S1 normal. S2 single. No murmur.
- **Pulmonary/Chest:** No respiratory distress. Good air entry.
- **Abdominal:** Full and soft. He exhibits no distension. Bowel sounds are increased. There is tenderness.
- **Skin:** Skin is warm. Capillary refill takes *3 to 5 seconds.*
Chest X-ray

No infiltrates
Sternotomy wires
Pacer

Labs

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Fontan Post-op

- Pulmonary blood flow is passive
- Cyanosis
  - Increase in PVR
  - Decrease in SVR
- Dehydration
  - Non fenestrated: decrease perfusion
  - Fenestrated: more cyanotic due to shunting

Fontan Other Considerations

- Fluid resuscitation
- Anasarca due to protein losing enteropathy
- Increase risk of arrhythmias and thromboembolic events.
A dry Fontain is a dead Fontan

- Be worried when Fontan’s are at risk for dehydration
- When dehydrated, boluses of 20mL/kg and repeated are ok

What information does cardiology require?

- 4 extremity BP’s, weight %iles
- H&P
  - Murmurs
  - Organomegaly
  - Pulses
  - ECG
  - Labs, CXR findings, saturations
Take home pearls

- Oxygen can kill
- A dry Fontan is a dead Fontan

How do you reach Peds Cardiology?

REFERRALS & CONSULTATIONS
M-LINE
800-962-3555
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- Stacey Noel, MD
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Thank you