Goals

• Overview of fetal circulation
• Categories of CHD and most common lesions
• Cardiac surgery and neuroprotection
• Neurodevelopment in CHD
Adult vs Fetal circulation
Adult vs Fetal circulation
Fetal oxygen saturation
Abnormal fetal circulation

TGA

HLHS
Categories of CHD

Isolated
- Pulm stenosis
- Aortic stenosis
- PDA
- VSD
- ASD

Complex
- Atrioventricular septal def
- Tetralogy of Fallot
- Transposition of the GA
- Double outlet RV
- Truncus arteriousus

Single Ventricle
- Hypoplastic left heart snd
- Heterotaxy
- Tricuspid atresia
- Pulm atresia / IVS

Acyanotic temporary cyanosis prolonged cyanosis
DD and CHD categories

![Bar chart showing prevalence of neurodevelopmental impairment across different categories: Mild, Moderate, Severe, Palliated Neonate, Syndromic. The chart indicates the percentage of severe impairment, mild or combined disabilities, and no disabilities for each category. Prematurity is indicated by an arrow pointing to the Mild category.]
Ventricular Septal Defect

- Most common form of CHD (15-20% of all CHD)
- Many spontaneously resolve (35%)
- Large VSD
  - Prototype of L->R shunt
  - Varying degrees of CHF, pulmonary overcirculation, FTT and poor feeding develop by 6-8 weeks
- Surgical closure by 6m of age to prevent pulm HTN
Aortic Stenosis

- 4:1 male:female
- Most common bicuspid Ao valve
- Large range from asymptomatic to a spectrum of left ventricular hypoplasia
- Often associated with coarctation of the aorta
- Most commonly managed in infancy with balloon valvuloplasty
Cyanotic heart disease

- Tetralogy of Fallot
- Transposition of the Great Arteries
- Tricuspid Atresia
- Truncus Arteriosus
- Total Anomalous Pulmonary Venous Return
- Pulmonary Atresia
- Ebsteins
Spectrum of Tetralogy of Fallot

“pink” TOF

“Blue” TOF

TOF with pulm valve atresia and MAPCAs
Physiology

What factors determine where this RBC will go?

Variable degrees of cyanosis depending on age, activity, anxiety, medications
Transposition of the Great Arteries

- 2:1 male:female
- Least common critical CHD to be prenatally diagnosed
- Often presents with severe cyanosis and possibly shock shortly after birth
  - Stabilization with creation of ASD prior to surgery
  - Arterial switch procedure in first week of life
Single Ventricles

- HLHS
- Norwood
- Glenn
- Fontan

Other conditions:
- heterotaxy
- Unbalanced CAVC
- Tricuspid Atresia
- Double Inlet LV
- Pulmonary atresia / IVS
- Complex TGA
HLHS

- Complex spectrum of hypoplasia of the left sided structures such that the left ventricle/aorta is not able to supply the cardiac output.
- Uniformly fatal until mid 1980s → now expected ~70% of infants born with HLHS will enter adulthood
- Most morbidity in the first 6 months
  - Oldest patients 25 years old
  - 2-3% of all congenital heart disease
  - ~1000 infants in the US / year
  - 70-80% fetal diagnosis
HLHS physiology

- Excessive pulmonary blood flow at the expense of systemic blood flow.
  - Shock, NEC, poor cerebral and coronary perfusion

Ultimate goals for palliation for HLHS
- Separate the pulmonary and systemic circulations
- Normalize arterial O₂ saturations
- Normal ventricular volume load
Norwood procedure

1. Damus-Kay-Stansel (DKS) anastomosis
   • Y together the MPA and ascending aorta
   • Stable coronary circulation

2. Arch reconstruction with MPA, native aorta, homograft material
   • Unobstructed systemic output

3. Resect the atrial septum
   • Unrestrictive atrial communication

4. RV-PA shunt (4mm Gore-tex "Sano"
   • Alternative is BT shunt
   • Stable source of pulmonary blood flow

1st week of life
• 70% survival
• ~4 week hospitalization
• Delayed sternal closure
• ~10 days of ventilation
• 10-15% ECMO post-op
• 10-25% vocal cord dysfunction
• 10-15% NEC
Interstage period

• Time between discharge from Norwood (stage 1) and Glenn (stage 2)

• Increased period of vulnerability / unstable physiology

• Interstage mortality between 10-15% within the 3-4 month window
  – Post-mortem analysis shows concerning cardiac lesion in ¾ patients
  – combination with minor respiratory / GI illness
    • Often within 24h of symptoms or physician visit
Stage 2 palliation: Glenn

- Eliminate the high pressure source of pulmonary blood flow.
  - Reduce pressure and volume load on the RV
  - Improve ‘effective’ pulmonary blood flow
  - Improve sats?
  - Improve growth

- Obligate increase in the cerebral venous pressure 3-4mmHg → 12-14mmHg
Stage 3 palliation: Fontan

- 3-5 years of age
- Connect the IVC to the pulmonary circuit
- Obligate increase in the central venous pressure (GI, renal, hepatic)
Heterotaxy

• Abnormal L/R sidedness of visceral organs
• Often associated with complex arrangements of cardiac segments
  – Some (not all) will require single ventricle palliation
  – Frequent intracardiac anatomy is “AV canal” or “double outlet RV”
• High rate of need for pacemaker
Cardiac surgery

• Large topic that can’t be explored in 5 minutes

• Understanding techniques for cardiopulmonary bypass
Cardiopulmonary bypass: similar to VA ECMO → blood withdrawn from venous limb (IVC/SVC or RA), oxygenated and returned to aorta to maintain perfusion pressure, heparinized.  Typically performed at 32°C for neuroprotection.

Cross-Clamp: time when heart is ‘arrested,’ allowing intracardiac repair while bypass supports the circulation.
Deep hypothermic circulatory arrest

• Selective use for complex aortic arch reconstructions (Norwood, complex coarctation) and some additional operations that require bloodless field
• Patient cooled to 12-18°C, exsanguinated into reservoir, with no BP, breathing or brain activity
  – ‘clinical death’
• Maximum is 60min w inflection point at 40min for neurological injury
• Mostly replaced with “low flow cerebral perfusion,” under moderate hypothermia with 25ml/kg/min of flow.
Feeding dysfunction

- vocal cord paresis related to left recurrent laryngeal nerve injury
- perioperative neurologic injury
- pre- or postoperative cardiac dysfunction
- poor oromotor coordination related to left vagus nerve injury
- delayed oral feeding
- Genetic syndromes
### SCH Data: 1V Neonates S/P Norwood

#### Feeding at discharge

| N=27 | 6% All PO | 5% PO/NG | 35% NGT | 8% GT | 46% ND/NJ |

#### Feeding at BDG readmission

| N=25 | 16% All PO | 32% PO/NG | 40% NGT | 6% GT | 0% ND/NJ |

### Table: Feeding Patterns

<table>
<thead>
<tr>
<th>Author/Institution</th>
<th>Period</th>
<th>Cases</th>
<th>Feeding at d/c</th>
<th>% All PO at BDG</th>
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</thead>
<tbody>
<tr>
<td>Anderson 2010 (Cincinnati)</td>
<td>2001-2007</td>
<td>102 (survivors to BDG only)</td>
<td>All PO: 25% NGT: 71% GT: 4%</td>
<td>69%</td>
</tr>
<tr>
<td>Lambert 2014 (15 centers, PHN/SVRT)</td>
<td>2005-2008</td>
<td>432</td>
<td>All PO: 32% NGT: 54% GT: 13%</td>
<td></td>
</tr>
<tr>
<td>Hill 2014 (47 centers, NPC-QIC)</td>
<td>2008-2012</td>
<td>465 (survivors to BDG only)</td>
<td>All PO: 44% NGT: 38% GT: 18%</td>
<td>62%</td>
</tr>
</tbody>
</table>
Emerging concepts for ND disability in CHD patients

- Growing body of literature points to multiple, minor developmental, cognitive, emotional and psychological deficits in patients with repaired CHD.
  - Specific phenotype developmental delay
- *Developmental deficit is likely the thing that affects the majority of our patient’s life, not physical deficit.*
- Early identification and treatment is effective and feasible yet not being done.
- The responsibility lies within the realm of cardiology and cardiac surgery.
Structural brain defects

- Altered maturation
- Brain maturation of term neonates with CHD
- Acquired injury
  - White matter injury, stroke, hemorrhage in 15-40% of patients
Brain injury pre and post- cardiac surgery

- Largest study to date- MRI pre / post / 3 month post of 153 children undergoing surgery before 8 weeks
  - Patients in 4 categories: 2V, 2V +arch repair, 1V and 1V +arch repair
  - 20% with white matter injury pre and 47% with new white matter injury post
    - univariate analysis --New injury associated with gestational age, BW, brain maturity score, diagnostic category, DHCA, lactate and delayed sternal closure
    - Multivariate analysis – gestational age, BW and brain maturity score

New White Matter Brain Injury After Infant Heart Surgery Is Associated With Diagnostic Group and the Use of Circulatory Arrest

*Circulation. 2013;127:971-979*
New White Matter injury

CPB

DHCA
Boston Circulatory Arrest Trial

• Reported at 1, 4, 8 and 16 years follow-up

• Mild treatment effect between DHCA vs LFBP

  – DHCA associated with worse fine, gross or oromotor function and visual-spatial skills. LFBP more impulsive

  – At 8 years, still within normal range for IQ, but on average ~1 standard deviation from the norm.
    • 1/3 remedial academic support, 10% repeated a grade
    • Lower than expected academic achievement, fine motor skills, visual spacial skills and social cognition.
Questions?