ADULT CONGENITAL HEART SURGERY

Chelsea C. Capone, CCP, FPP
Lead Pediatric Perfusionist
UCSF Benioff Children's Hospitals

OBJECTIVES

• Review prevalence of ACHD
• Address CPB issues specific to ACHD patients
• VAD/transplant experience in ACHD population
• Review ACHD experience at Benioff Children's Hospitals

WHY?

• 1 in 100 babies are born with CHD.
• Greater than 90% of patients surviving initial surgery and living into adult years with good to excellent quality of life.
• Operative mortality for congenital heart disease is less than 2%.
• Early mortality less than 5%.
• Greater than 1 million in US alone
Nationwide Hospitalization Trends in Adult Congenital Heart Disease Across 2003–2012

Shikhar Aggarwal, MD, MPH; Karen Sud, MD; Venu Manca, MD

PREPARE FOR GUCH!
CPB CONSIDERATIONS

- High-risk re-entry strategies
- Anatomic problems
- Cannulation strategies
- Equipment considerations
- Collaterals
- Staffing
- Polycythemia
- Temperature strategies
HIGH RISK RE-ENTRY

- Planning is key
  - CT, 3D Models, Thoracoscopy
  - MRI, Catheterization
- Dissection
  - Oscillating saw
  - Limited dissection
- Alternative access sites
- Pleural Space

ANATOMIC

- RA
  - N-TGA
  - TGA
  - Fontan
- RV
  - Alice’s
  - TOF
  - TGA
- LV
  - Injured
  - Others
- Coronary distribution/blood

AIR EMBOLISM

- Aortic/Left heart injury/laceration
  - DHCA
- Right heart injury/laceration
  - Left to right shunts/Pulmonary CVP
- Negative venous pressure/Intraocular shunt=AIR

HIGH RISK RE-ENTRY

Risk Factors and Early Outcomes of Multiple Reoperations in Adults With Congenital Heart Disease

- Independent risk factors for early death:
  - Urgency
  - Increased CVP
  - Single-ventricle diagnosis
- Increased risk of cardiac injury
- Increased number of prior sternotomies

Strategies for High-Risk Reoperations in Congenital Heart Disease

- Inadvertent cardiectomy with significant hemorrhage during redo sternotomy increases mortality from 4.5 to 25%
- Reoperation between 6-12 months associated with worse outcomes and more difficult dissection.
**CANNULATION STRATEGIES**

- Discuss with surgeons prior to incision...COMMUNICATION!!!
- Alternative cannulation strategies
- Anatomic concerns
- Bypass strategies
  - Off-pump
  - CPB for open
  - CPB from beginning to end
  - Circulatory Arrest

**ALTERNATIVE CANNULATION**

- Femoral artery or vein
- Iliac artery or vein
- Axillary artery
- Internal JF
- Abdominal aorta
- Thoracotomy (right or left)
- Right axil
- Ascending aorta
- PA
- Pre-op assessment of femoral and neck cannulation sites
  - Possible stroke from cannulation of R carotid and RJ
  - Femoral vessels may be scarred or stenotic from multiple surgeries and catheterizations.
  - Previously repaired vessels (s/p ECMO) though patent may be narrow.

**CANNULAS**

- Medtronic Bio-Medicus
- NextGen Cannula
- Edwards Arterial/Inferior Vena Cava
- Edwards Venous
- Medtronic ECM Arterial
- Edwards OptiSiteArterial Cannula

- Bicaval venous cannulation for defects with intracardiac communication
- Bilateral SVC's
- Be prepared to cannulate with multiple venous cannulae and Y venous line
- Air entrainment via L-R shunt=venous air lock (made worse with VAVD)
- ID shunts via TEE prior to surgery

- s/p Arch repairs and/or prior aortic cannulation
- Site issue on aorta=risk of aortic dissection and/or dislodgement of emboli from calcified homograft=Can cause more dissection
- s/p Coarctation
- Assurant site of repair
- Spontaneous dissection possible in ascending aorta, especially when paired with bicuspidAV
- s/p PDA ligation
- Cannula may migrate into indentation in aorta at take off site
DISPOSABLES/EQUIPMENT

- Circuit Selection
- Arterial and Venous T
- Suckers
- Cell saver
- CO2 at field
- VAD
- ECMO circuit
- VADs

COLLATERALS

- When MAP is lower than expected despite full CPB support, look for L-R shunt
- Shunts to lungs will flood surgical field
- Aggressive pump suckers and/or cell saver
- Vent
- Washout of CDPG

SVO2

- Inaccurate with L-R shunts
- Acceptable SVO2 levels doesn’t equate to adequate perfusion
- Monitor lactate!
- Monitor cerebral NIRS

MYOCARDIAL PROTECTION

- AI
- Higher CDPG flow rates to close AV
- Ostial and/or retrograde
- Collaterals
- Sinusoids
- Abnormal coronary anatomy
- Hyperkalemia may occur with extra CDPG

- CHD with Chronic hypoxemia
- Less able to tolerate ischemia
- Reperfusion injury with sudden increase in partial P of oxygen. Can redevelop coronary reperfusion as well as post CPB pulse HTN and AV valve

OTHER POTENTIAL COMPLICATIONS...

- AI
- Unrecognized LV distention
- Subsequent myocardial injury without decompression
- Ventricular Rhythm
- Left atrial distention
- Lower extremity compartment syndromes
- Inadequate arterial perfusion (shunt)
- Inadequate venous drainage of legs due to cannula obstruction
AND MORE …

- Anatomical shunts
  - Left to right:
    - Lower systemic blood flow
    - Pulmonary edema
    - Air embolism
    - Massive air embolism
  - Right to left:
    - Massive air embolism

AND MORE …

- Filling while weaning
- Arrhythmias
- Coagulopathy
- Polycythemia
- Thrombocytopenia
- Reduced von Willebrand factor
- Chronic Renal Hypoxia
- Gout, hyperuricemia

STAFFING

BEST APPROACH!

- PLAN
- COMMUNICATE
- TEAMWORK
SPECIFIC PATIENT POPULATIONS

PREGNANCY IN ACHD

PREGNANCY AND ACHD

- Pregnancy-induced cardiovascular stress can cause:
  - Arrhythmias
  - Heart failure
  - Thromboembolism
  - MI
  - Aortic dissection
- Pre-eclampsia increases risk of heart failure
- More likely to experience adverse events after pregnancy as well
- Fetal risks:
  - Prematurity
  - Low birth weight
  - Recurrence of CHD
PREGNANCY

- Solutions:
  - Multi-disciplinary delivery plan
  - ECMO Standby
  - Follow by cardiologists
  - Intervention during 2nd trimester
  - Try to delay open heart surgery and CPB

HEART FAILURE

- HF definition from Heart Failure Society of America:
  - "HF is a syndrome characterized by either or both pulmonary and systemic venous congestion and/or inadequate peripheral oxygen delivery."
  - HF is leading cause of death in adults with CHD
  - Many interventions with CHD is palliative rather than curative.
  - Patients develop complications...HF!

HEART FAILURE IN ACHD

- HF definition from Heart Failure Society of America:
  - "HF is a syndrome characterized by either or both pulmonary and systemic venous congestion and/or inadequate peripheral oxygen delivery."
  - HF is leading cause of death in adults with CHD
  - Many interventions with CHD is palliative rather than curative.
  - Patients develop complications...HF!
**FAILING FONTAN**

- RA dilation
- Baffle thrombus
- PE
- Atrial arrhythmias
- Failure of systemic RV
- Restrictive physiology
- High PVR
- Cyanotic L-R shunt

- Systolic or diastolic ventricular dysfunction (resulting in increased ventricular filling pressures)
- Protein-losing enteropathy
- Multiple long-term complications exacerbate HR symptoms
- Increased CVP and low CO
- Chronic congestive hepatopathy with hepatic dysfunction, cirrhosis, ascites, varices, and hepatorenal syndrome
- Low flow through aortopulmonary connection increases risk of thrombosis and embolism which can lead to increased PVR and CHF
- Arrhythmias
- Cyanosis

**CCTGA**

- L-T develops HF by 5+ decade
- RV is systemic ventricle
- Develops cyanosis HF due to limitations of RV to support systemic circulation
- TV failure facing systemic pressures
- RCA cannot meet myocardial perfusion demands of systemic ventricle leading to myocardial ischemia
- Cardiac transplantation is current definitive solution for HF in these patients
TRANSPLANT IN CHD

- No special listing for patients with CHD.
- Less likely to receive ICD or VAD as bridge to transplantation.
- Some not amenable to VAD and/or inotropes
- Lower listing status
- Even with VAD, CHD patients have longer wait list times and higher wait list mortality.
- CHD patients statistically wait longer on list than non-CHD.
- Anatomical, pulmonary HTN, renal and liver disease, anti-human leukocyte antigen.

OTHER OPTIONS?

- Late failing Fontan or CCTGA with organ failure
- PLE
- Cirrhosis
- Renal failure
- Plastic bronchitis
- NOT A TRANSPLANT CANDIDATE

- All ACHD VADs and TAH between June 2006 and January 2016
- Characteristics of ACHD:
  - Younger (42 vs. 56)
  - More BiVADS and TAH (11/10% vs. 5/2%)
  - More RV dysfunction (47 vs. 32%)
  - Same % of INTERMACS 1 profile
  - LVADs
  - ACHD patients similar outcomes to non-ACHD patients
  - Survival for ACHD = Survival for Non-ACHD
  - 12 mos after MCS: 73% vs 80%

Inflow cannula occlusion with systemic RV predisposes to

- Hypertrophied trabeculations in the RV
- Extensive muscle resection necessary
- RV inflow cannula superior to atrial cannulation
- Mustard or Senning baffles cause obstruction to inflow cannula

Outcomes of ventricular assist device implantation in children and young adults: the Melbourne experience

- Global outcomes after ventricular assist device implantation in adults with simple congenital heart disease
- Survival at 6 months: 73% vs 80%
- Significant number of patients with INTERMACS 5
- LVADs are underutilized in ACHD patients with end stage HF

Ventricular assist devices for the failing univentricular circulation

- Chronic Heart Failure in Congenital Heart Disease
  - Systemic RV predominates inflow cannula occlusion
  - Inoperable atrioventricular lesions in the RV
  - Excessive atrioventricular valve regurgitation
  - RV inflow cannula superior to aortic cannulation
  - Pulmonary or systemic cause obstruction to inflow cannula

CCTGA CASE REPORTS

- Systemic RV inflow cannula failure:
  - Inoperable atrioventricular valve lesions
  - Pulmonary cause obstruction to inflow cannula

CCTGA AND VAD'S

- Systemic RV predominates inflow cannula occlusion
  - Inoperable atrioventricular lesions in the RV
  - Excessive atrioventricular valve regurgitation
  - RV inflow cannula superior to aortic cannulation
  - Pulmonary cause obstruction to inflow cannula
WHERE TO PERFORM ACH SURGERY!

- DVT
- CAD
- PVD
- Emphysema
- Renal-acute and chronic renal failure
- GI-irritations of liver, pancreatitis, viral hepatitis
- Psychiatric-depression, anxiety, panic disorders, substance abuse
- Endocrine-hypothyroidism, diabetes
- Gout
- Obstructive sleep apnea

COMORBIDITIES IN ACH

- DVT
- CAD
- PVD
- Emphysema
- Renal-acute and chronic renal failure
- GI-irritations of liver, pancreatitis, viral hepatitis
- Psychiatric-depression, anxiety, panic disorders, substance abuse
- Endocrine-hypothyroidism, diabetes
- Gout
- Obstructive sleep apnea
- Alzheimer's

Adult congenital heart disease: the challenges of a lifetime

"The development of an adult CHD program requires collaboration..."

"It is necessary to build an interdisciplinary team to care for patients' complex needs including in other medical disciplines... particularly to treat the most common complications of arrhythmias and heart failure."
Cardiac surgery in adults performed at children’s hospitals: Trends and outcomes

William T. Mahle, MD, Paul M. Kirshenfeld, MD, Kirk R. Kantor, MD, and Brian M. Koger, MD

- Adults with CHD undergoing surgery at a children’s hospital have hospital survival rates of more than 98%.
- Similar to other studies, hospital mortality was 1.6%.

- Adult vs. Pediatric Hospital
- Adult vs. Pediatric Heart Surgeon
- Canada and UK have dedicated adult centers for adult congenital heart surgery
- Mortality was similar at adult and pediatric hospitals (4.3 vs. 5.2%)
- Mortality was higher in adult surgeon group vs. pediatric surgeon group (15.2 vs. 2.7%)

Conclusions:
- Numerous coexisting medical problems are best facilitated in adult hospital setting.
- ACHD patients best served by congenital heart surgeon.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Pediatric Hospital n = 118 (Mean ± SD)</th>
<th>Adult Hospital n = 181 (Mean ± SD)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient factors</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at surgery</td>
<td>22 ± 6.6</td>
<td>37 ± 12.9</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Congenital medical problems</td>
<td>23 (19%)</td>
<td>17 (14%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>STS database defined risk factors</td>
<td>16 (13%)</td>
<td>43 (37%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Previous operations</td>
<td>36 (30%)</td>
<td>51 (44%)</td>
<td>0.124</td>
</tr>
<tr>
<td>Operative factors</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>BAV/3 valve</td>
<td>3.80 ± 0.9</td>
<td>4.17 ± 0.7</td>
<td>0.078</td>
</tr>
<tr>
<td>Outcomes</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mortality</td>
<td>6 (5.1%)</td>
<td>64 (3.7%)</td>
<td>0.155</td>
</tr>
</tbody>
</table>

Table 6: Comparison Between the Pediatric and Adult Hospital and Pediatric and Adult Surgeons

National Practice Patterns for Management of Adult Congenital Heart Disease

Operation By Pediatric Heart Surgeons Decreases In-Hospital Death

Tara Kamalton, MD; Brian S. Dipp, PhD; Thomas Peshon, MD; Ross M. Ungelker, MD, MBA; Carl F. Wolfe, MD, MS

- Mortality for CHD and total hospital charges significantly lower when operations performed by pediatric hearts surgeon vs non-pediatric heart surgeon (1.87% vs. 4.84%)
- Increased experience with pediatric heart surgery resulted in lower mortality
- Referral of GUCH patients to centers with specialized pediatric heart surgeons will reduce in-hospital death rates and improve the value of delivered care.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Pediatric Hospital n = 275 (Mean ± SD)</th>
<th>Adult Hospital n = 40 (Mean ± SD)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient factors</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at surgery</td>
<td>28 ± 15.4</td>
<td>42 ± 16.4</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Congenital medical problems</td>
<td>38 (14%)</td>
<td>36 (13%)</td>
<td>0.298</td>
</tr>
<tr>
<td>STS database defined risk factors</td>
<td>18 (13%)</td>
<td>35 (13%)</td>
<td>0.158</td>
</tr>
<tr>
<td>Previous operations</td>
<td>67 (25%)</td>
<td>39 (25%)</td>
<td>0.632</td>
</tr>
<tr>
<td>Operative factors</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>BAV/3 valve</td>
<td>3.80 ± 0.8</td>
<td>4.17 ± 0.7</td>
<td>0.032</td>
</tr>
<tr>
<td>Outcomes</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mortality</td>
<td>7 (2.7%)</td>
<td>7 (17.5)</td>
<td>0.0008</td>
</tr>
</tbody>
</table>

GUCH = adult congenital heart surgery; SD = standard deviation; STS = Society of Thoracic Surgeons.
• GUCH patients still offer unique physiologic and anatomic challenges consistent with congenital rather than adult acquired heart disease.
• Comorbidities: renal failure, PVD, COPD, DVT, substance abuse
• GUCH fare best when treated by congenital heart surgeons in a children’s hospital.
• In-hospital mortality reduced fourfold
• Once again emphasizes mortality decreased in this patient population due to refined processes of care at specialized pediatric centers in conjunction with experienced pediatric surgeons.

WHERE TO PERFORM ACH SURGERY!

OUR EXPERIENCE AT BENIOFF CHILDREN’S HOSPITALS

WHERE TO PERFORM ACH SURGERY!

OUR EXPERIENCE AT BENIOFF CHILDREN’S HOSPITALS
THANK YOU!
Overall mortality low

Admission characteristics predictive for in-hospital mortality:
- Older ACH patients
- Male sex
- Surgical complexity
- Government sponsored insurance

Pediatric hospitals with high ACH volume had the lowest mortality rates, when compared to pediatric hospitals with low adult volume.