The Adult with Congenital Heart Disease: When to Worry

Joseph D Kay MD
Associate Professor of Medicine & Pediatrics
Colorado’s Adult & Teen Congenital Heart Disease (CATCH) Program Director
2/26/2014

Disclosures
None

Outline
• Why talk about ACHD at an Anesthesia Conference? Incidence and increased survival
• Historic Shunts and Late effects
• Physiologic effects of lesions and late repair
  – Pulmonary Hypertension
  – Residual abnormalities
• Single Ventricle Physiology
• Pregnancy in Congenital Heart Disease
• SBE Prophylaxis
• Closing Pearls

Cardiology
Epidemiology - Changing Picture Of Congenital Heart Disease

Changing Prevalence of Adult Congenital Heart Disease (AHCD)

Changing Mortality in Congenital Heart Disease (CHD)

Khairy et al., JACC 56(14) Sept 2010
Mortality rate in CONCOR patients and in the general Dutch population by decade in 2007.

Verheugt C L et al. Eur Heart J 2010;31:1220-1229

Causes of, and age at, death in CONCOR patients (n = 197).

Verheugt C L et al. Eur Heart J 2010;31:1220-1229

CATCH ACHD Patients Seen Per Calendar Year

Rough Estimates of ACHD in US & Colorado

- By 2013, estimated 1.3 - 1.5 million ACHD survivors in US
- Total CO population 2011 – 5,116,796
- % > 18 y/o = 76% = 3,888,764
- Incidence of ACHD is 0.4 – 0.8%
- Assuming the lowest (to offset early death), estimated that 15,558 ACHD survivors are in Colorado

Basic Physiologic Problems

1. Too Much pulmonary blood flow limit (ASD, VSD, CAVCD, PDA)
2. Too little pulmonary blood flood shunt. (TOF, PA, TA, PA IVS)
3. Valvular or vessel obstruction relieve obstruction (AS, MS, PS, CoA)
4. Valve leaks repair or replace
5. Blue blood & red blood mixing correct anatomic reason (TGA, Truncus)
6. Half a heart Staged palliation to single ventricle

To Little Pulmonary Blood (Shunts)

1. Tetralogy of Fallot
2. Pulmonary Atresia
   - With intact Vent. Septum
   - With VSD
3. Transposition with Pulm. Stenosis
4. Single Ventricle Physiology
**Classic Blalock-Taussig Shunt (Lateral Thoracotomy)**

- First performed in 1945
- Benefits:
  - Will grow with child
- Problems:
  - BP on ipsilateral arm not reflective of central aortic pressure
  - Prone to thrombosis/kinking
  - "Steal" phenomenon from vertebral artery
  - PA stenosis

**Waterston Shunt (Lateral Thoracotomy)**

- First performed in 1946
- Problems:
  - Not pressure restrictive
  - Can cause CHF and PAH
  - Causes distortion of RPA with late PA stenosis

**Potts Shunt (Lateral Thoracotomy)**

- First performed in 1946
- Problems:
  - Not pressure restrictive
  - Can cause CHF and PAH
  - Causes distortion of LPA with late stenosis
  - Hard to take down surgically

**Modified BT Shunt (Lateral Thoracotomy) – Current Approach**

- First performed in 1961
- Performed on side opposite aortic arch
- Benefits:
  - Pressure restrictive
  - Less prone to kinking
  - DOES NOT AFFECT BP IN IPSILATERAL ARM
- Problems:
  - Gore-Tex doesn’t grow

**Too Much Pulmonary Blood Flow (Left to right shunting)**

- Septal defects - most common

**Too Much Pulmonary Blood Flow (Left to right shunting)**

- **Pre – Tricuspid left to right shunts (low pressure)**
- **Ventricular Level Shunts (High Pressure)**
  - Membranous, Muscular, Outlet, Inlet (AVCD)
- **Aortic Level Shunts (High Pressure)**
  - PDA, Aortic to PA Window, Aortic RA/RV fistula
Types of ASD: ECHO View

- A - Sinus venosus defect (usually with PAPVR)
- B - Secundum ASD
- C - Primum ASD
- D - Unroofed Coronary sinus (not shown)

Types of Ventricular Septal defects

- Top - 4 segments of the normal right and left ventricles
- Bottom - 4 types of VSDs

AV Canal Defects

- Top
  - A.) Normal
  - B.) Partial ECD with clefts in TV & MV
  - C.) Complete ECD
- Bottom (repair)
  - A.) Complete ECD viewed through the right atrium
  - B.) Inlet VSD closure and MV repair
  - C.) Primum ASD closure
  - D.) Final Repair

Too Much Pulmonary Blood Flow

Physiologic Consequences

- Pulmonary Arterial Hypertension (PAH)
  - More common with Higher pressure shunts (ventricular and aortic level) – if not fixed to after 2nd year of life
  - 5-10% occurrence of PAH in pre-tricuspid level shunts
- Ventricular dysfunction
  - Valvular regurgitation

PA Band – Palliation for large VSD

First performed in 1952
Prevents excessive pulmonary blood flow
Problems:
- Too tight
- Too loose (Late PAH)
- Distortion of branch PA’s
- Restenosis of PA

Coarctation
Coarctation Repairs

END TO END REPAIR

SUBCLAVIAN FLAP

DACRON PATCH

Coarctation - Facts

- Associated with diffuse arteriopathy of varying degree
  - Abnormal vascular reactivity in many
  - Medial necrosis of aorta in many (leading to Aortic aneurysms)
  - Intracranial Berry aneurysms incidence 3-6 x normal population
    (3 studies suggesting incidence of 10%)
  - Increased arterial and central aortic stiffness in majority

- Older age of repair associate with increase HTN late (> 5 y/o)
- Young age (< 12 m/o) associated with increased risk of renarrowing

Complications with Both Approaches to Coarctation

Pseudoaneurysm After Stenting

Restenosis and Aneurysm After surgery

Forbes TJ. JACC; 58(25) 2011:2664-74

Incidence of Intracranial Aneurysms (ICA)

- OSU Screened 43 Adults with repaired coarctation CT /MR
- Found 5 patients (11%) with ICA
- Only risk in this group was age
  - Avg 45.6 in ICA pts –vs- 30.89 in no ICA (p = 0.0003)
  - Trend with HTN but not significant (p = 0.167)
Concordant with Previously published Research on Incidence late after coarctation repair


II. Aortic Aneurysms

High Increased Risk of Aortic Pseudoaneurysm with Gortex Patch Repair


Coarctation Pre-Operative Recommendations

- Assess 4 limb BP’s with every encounter (particularly both arms to know which to monitor during anesthesia) – Subclavian flap with low left arm BP

- Important to have pre-operative assessment for aneurysms (particularly when older than 40)
Single Ventricle and Fontan Palliated Patients

ACHD population with the largest proportional increase in patients
>80% mortality of these lesions prior to 1970s

Evolution of the Fontan

Fontan - Long Term Complications

- Pulmonary atriovenous malformations (AVM’s) – Hypoxemia (air filters on IVS, accept lower saturations)
- Protein Losing Enteropathy (up to 10% in some studies) – 50% 5 year mortality (Hypercoagulable, low intravascular oncotic pressure)
- Thromboembolism (High central venous pressure)
- Arrhythmias – Greater than 50%, increases with age
- Congestive Heart Failure / Poor exercise tolerance
- Cirrhosis
- Coronary issues
- Progression of cyanosis (veno-atrial collaterals)
- Health and life insurance difficulties (now all eligible for Medicare)

Fenestrated Extra Cardiac Fontan & Bidirectional Glenn

Veno – Atrial Collaterals
Fontan – Important Considerations
1. Very Pre-load sensitive – Drop in BP in dehydrated adult (NPO); Consider pre-operative hydration
2. Air filters for IVs in any patient with resting saturations lower than 93% (or lower than 90% with activity) - Risk of air embolism to Brain
3. Caution with surgery that increases Intra-abdominal pressure (laproscopic) – Decreases Cardiac output
4. Caution with prolonged positive pressure ventilation – Decreases Cardiac Output
5. SBE prophylaxis if cyanotic

PREGNANCY AND THE HEART
Normal Hemodynamics Changes During Pregnancy
- Peripheral resistance ↓
  - Uterine blood flow
- Blood volume ↑40-45%
- Heart rate ↑10-20%
- Blood pressure → or ↓
- Pulmonary vascular resistance ↓
- Venous pressure in lower extremities ↑

Cardiac output ↑30% to 40%

Respiratory Changes with Pregnancy
- Minute Ventilation
- O2 Consumption
- PaO2
- PaCO2
- Functional Residual Capacity
- Inspiratory Capacity
- Total Volume
- Airway resistance
- Total Lung Capacity Unchanged

Hematological Changes in Pregnancy
- Plasma Volume
- Red Cell Mass
- Hematocrit
- Hemoglobin
- Mean Corpuscular Hgb
- Red Blood Cells
- Leukocytes
- Platelets
- Clotting Factors

HEMODYNAMIC CHANGES DURING LABOR AND DELIVERY
Uterine contractions - CO ↑ 50 - 80%
- Increase is blunted to 30% ↑ with C/S and epidural or GA
- Loss of 300 - 400 cc in normal vaginal delivery
- Cesarean section : 500 - 800 cc
- Hemodynamics return to baseline about 6wk after delivery
Risk Factors for Adverse Maternal Outcomes with Pregnancy

1. Previous CHF, TIA, CVA, or arrhythmia
2. Baseline NYHA > 2
3. Cyanosis (< 90%)
4. Left Heart Obstruction
   1. MVA < 2 cm²
   2. AVA < 1.5 cm²
   3. Peak LVOT gradient > 30 mmHg
5. Reduced Systemic Ventricular Function (<40%)

Cardiac Lesions at Higher Risk with Pregnancy

- Obstructive lesions
  - Aortic Valve stenosis
  - Mitral valve stenosis
  - Pulmonary or tricuspid valve stenosis
  - Coarctation of the Aorta (although frequently well tolerated with extensive collaterals)
- Poor systemic ventricular function (< 40%)
- Pulmonary Hypertension (Pulm pressures ≥ systemic pressure)
- Marfan Syndrome
  - Ascending aorta > 40 mm (45 in European guidelines)
- Cyanosis (< 90%)
  - Those Without Eisenmengers Syndrome
  - Cyanosis with Eisenmenger Syndrome

Low Risk Cardiac Lesions

1. Left to right shunts without pulmonary HTN
   - ASDs, PAPVR, small PDA, Small VSD
2. Regurgitant Lesions
   - Moderate to severe PI (although one study suggests increased risk of prematurity)
   - MR and AI (although slight increased risk of post partum CHF)
3. Mild Valvular stenosis

Heart Rate Response During Exercise and Pregnancy Outcome in Women With Congenital Heart Disease

Question: Does exercise testing predict pregnancy outcomes in women with ACHD?

- Retrospective assessment of outcome of pregnancy (maternal and fetal) in women with ACHD whom had CPX within 2 years of pregnancy
- 89 pregnancies in 83 women in 12 centers
- Heart rate reserve and chronotropic indexes not used in women with pacemaker implants

<table>
<thead>
<tr>
<th>Heart Rate Response During Exercise and Pregnancy Outcome in Women With Congenital Heart Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>All Pregnancies Excluding Patients With Pacemakers</strong></td>
</tr>
<tr>
<td>RER</td>
</tr>
<tr>
<td>Chronotropic index</td>
</tr>
<tr>
<td>Heart Rate Reserve</td>
</tr>
<tr>
<td>Percentage Age-predicted HR</td>
</tr>
<tr>
<td>Peak Heart Rate</td>
</tr>
<tr>
<td>Resting Heart Rate</td>
</tr>
<tr>
<td>Peak VO2 cc.KG/min</td>
</tr>
</tbody>
</table>

- Chronotropic index = [(peak HR - resting HR)/220 - age - resting HR]
- Heart Rate reserve = (peak HR - resting HR)
Heart Rate Response During Exercise and Pregnancy Outcome in Women With Congenital Heart Disease

- One or more adverse Cardiac events occurred in 18%
  - CHF in 14%
  - Sustained arrhythmia in 7%
  - 1 cardiac arrest and death
- Neonatal events occurred in 17 (20%)

<table>
<thead>
<tr>
<th>Predictors of Maternal Events</th>
<th>OR (95% CI)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>CRI</td>
<td>0.65 (0.47 – 0.9)</td>
<td>0.0089</td>
</tr>
<tr>
<td>Peak HR</td>
<td>0.71 (0.53 – 0.94)</td>
<td>0.0173</td>
</tr>
<tr>
<td>%MPHR</td>
<td>0.93 (0.88-0.98)</td>
<td>0.0106</td>
</tr>
<tr>
<td>HRR</td>
<td>0.78 (0.56-1.07)</td>
<td>0.121</td>
</tr>
<tr>
<td>PVO2</td>
<td>0.91 (0.81-1.03)</td>
<td>0.125</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Predictors of Neonatal event</th>
<th>OR (95% CI)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>CRI</td>
<td>0.74 (0.69-0.98)</td>
<td>0.0274</td>
</tr>
<tr>
<td>Peak HR</td>
<td>0.75 (0.60-0.94)</td>
<td>0.036</td>
</tr>
<tr>
<td>%MPHR</td>
<td>0.94 (0.85-0.99)</td>
<td>0.024</td>
</tr>
<tr>
<td>HRR</td>
<td>0.8 (0.64-0.99)</td>
<td>0.041</td>
</tr>
<tr>
<td>PVO2</td>
<td>0.91 (0.82-1.0)</td>
<td>0.06</td>
</tr>
</tbody>
</table>

• Neonatal events occurred in 17 (20%)

Pre-pregnancy (Pre-anesthesia) Visit (Women with moderate/Severe CHD)

- Careful History and physical
- Echo
- EKG
- Cardiopulmonary Exercise Testing

Discuss carefully risks of Pregnancy
- To Patient
- To Fetus (including risk of Cong Heart Def.)
- Risks of various contraceptive choices

2008 ACHD / AHA Guidelines

- Evaluate patients before dental procedures that involve manipulation of gingival tissue or the periapical region, or both. Patients with a history of endocarditis are considered to have CHD with the highest risk for adverse outcomes from IE, including those with the following indications:
  - Prosthetic cardiac valve or prosthetic material used for cardiac valve repair (Level of Evidence: A)
  - Previous IE (Level of Evidence: A)
  - Unrepaired or palliated cyanotic CHD, including surgically constructed palliative shunts and conduits (Level of Evidence: B)
- Prophylaxis is reasonable in patients with CHD with the highest risk for adverse outcomes from IE, including those with the following indications:
  - Prosthetic cardiac valve or prosthetic material used for cardiac valve repair (Level of Evidence: B)
- Prophylaxis is not recommended for non-dental procedures (such as esophagogastroduodenoscopy or colonoscopy) in the absence of active infection.

- Dental Procedures with Artificial valves or residual shunt
- Not recommended for native Valve problems, or intracardiac shunts closed > 6 months ago

PEARLS

1. Carefully exam the ACHD patient before surgery
   - 4 limb BP of lateral thoracotomy
2. Ensure pt has had pre-op cardiac exam with ACHD expert if elective surgery
3. Call your local ACHD expert with any questions or concerns (We don’t mind, as we tend to be over protective)
4. Air filters on IV’s for cyanosis (saturations < 90%)

ACHD SBE Recommendations

Thank You For Your Attention

Joseph.kay@ucdenver.edu