Congenital Heart Disease in the Adult

Paul Reynolds, MD
Uma and Sujit Pandit Professor and Chief of Pediatric Anesthesiology
The University of Michigan
Department of Anesthesiology
Topics to be Discussed

- Incidence
- History
- Primer on CHD
- Specific heart defects
- Pregnancy
- Who should anesthetize
Congenital Heart Disease

- Occurs in 1/100 live births
- 40,000 births/year in the USA alone
- #1 birth defect, #1 cause of deaths (birth defects)
- 10% deaths prior to diagnosis
- Of children having surgery:
  - 50% < 1 year old
  - 25% < 1 month old
- 1/13 infant deaths due to CHD
Fast Facts

• 30% of congenital disease is cardiac
• CHD mortality decreased 40% in past 20 years
• 80% or more survive to adulthood
• 2 million people with CHD in the US today, 50% adults
• 50% would have died without intervention
• 10% of all CHD are first diagnosed in adulthood
• Baby's risk of CHD increased 3X if parents/siblings have CHD
Fast Facts

- CHD as common as Autism
- CHD 25 times more common than Cystic Fibrosis
- 2X as many children die of CHD as all cancer combined
- Funding for childhood cancer 5X higher than CHD
- 2009; 40,000 hospital stays for CHD
- Health care cost nearly $2B/yr in the US
- <10% of adults with CHDs in the U.S. who need care from specialty adult CHD centers are receiving this recommended care
## Incidence

<table>
<thead>
<tr>
<th>Condition</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>30%</td>
</tr>
<tr>
<td>PDA</td>
<td>9%</td>
</tr>
<tr>
<td>ASD</td>
<td>7%</td>
</tr>
<tr>
<td>PS</td>
<td>7%</td>
</tr>
<tr>
<td>CoA</td>
<td>6%</td>
</tr>
<tr>
<td>TGA</td>
<td>5%</td>
</tr>
<tr>
<td>TOF</td>
<td>5%</td>
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<tr>
<td>AS</td>
<td>5%</td>
</tr>
<tr>
<td>HLHS</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>Other</td>
<td>25%</td>
</tr>
</tbody>
</table>

History of Cardiac Surgery

1938-1948: closed heart surgery
1952-1955: open heart surgery for CHD
1960-1970: open heart surgery for acquired disease
1970-1980: improved CPB, cold cardioplegia, DHCA
1980-1990: transplantation, neonatal surgery, IMAs, aprotinin
1990-1999: fast tracking, minimally invasive surgery, off bypass CABG, NO, 3 stage HLH repair
2000-present: VAD, Transmyocardial Laser Revascularization, hybrid procedures, Melody valves, TAVR
Surgical Mortality According To Age And Years

- <1Y
- 1-5 Y
- 6-12Y
- >12Y

1954-64
1990-2000
Qp/Qs

- Normal = 1
- $<1 = \text{R to L shunt “Blue”}$
- $>1 = \text{L to R shunt “Pink”}$
Shunt Physiology in CHD

– Too much pulmonary blood flow leads to congestive heart failure and eventually pulmonary vascular disease
– Too little pulmonary blood flow leads to cyanosis and systemic hypoxemia
Pulmonary Vascular Resistance

Increased by:
- hypoxia
- hypercarbia
- acidosis
- alpha agonists
- atelectasis
- hypovolemia
- hypothermia
- high airway pressures (PEEP)

Decreased by:
- oxygen
- hyperventilation
- alkalosis
- alpha antagonists
- PGE/prostaglandins
- vasodilators (SNP, NO)
- amrinone/milrinone
- isoproterenol
Blood flow through Shunts

- Poiseuille's Law:
  \[ \text{Resistance} \propto \frac{\text{length}}{\text{radius}^4} \]
- Size of orifice (or length of shunt)
- Pressure in the chambers on either side of shunt
- Resistances downstream (PVR and SVR)
Right to Left Shunts

• Tetralogy of Fallot (TOF)
• d-Transposition of Great Arteries (d-TGA)
• Tricuspid Atresia
• Truncus Arteriosus
• Pulmonary Atresia/Intact Ventricular Septum (PA/IVS)
• Eisenmenger’s Syndrome
Tetralogy of Fallot

1. Pulmonary stenosis (thickened, narrow pulmonary outflow tract)
2. Thickened right ventricle wall
3. Ventricular septal defect
4. Aorta overrides septal defect
Adults with TOF

- Un-repaired
- Palliated (aorto-pulmonary shunt)
- Complete repair
Life Expectancy in Unrepaired TOF


66% live to one year
49% to 3 years
24% to 10 years
9% beyond 30 years
Sequelae of Uncorrected TOF

- Chronic hypoxemia
- Cyanosis, clubbing, SOB
- Compensatory polycythemia
- “Tet spells”, squatting only seen in children
- CVA, cerebral abscess
- Thrombocytopenia
- Risk of paradoxical embolic events
- Venous stasis
Palliative Systemic to Pulmonary Shunts

- Waterston shunt; ascending AO to RPA

- Potts shunt; descending AO to LPA

- Blalock-Tausig shunt; subclavian to PA
Sequelae of Palliative Correction

- Pulmonary hypertension
- LV volume overload
- Branch PA stenosis

Long-term Survival in Corrected TOF

Sequelae of Complete Surgical Repair

- Prolonged QT
- Sustained V-tach
- Atrial arrhythmias
- Pulmonary insufficiency or stenosis
- Sudden death

Poor Prognosis in TOF

- Older patient at the time of repair
- Elevated right ventricular pressure
- Pulmonary regurgitation

Should I feel comfortable?

Yes:

    Repaired TOF
    Treat symptomatically ie; RV failure

No:

    Palliated or unrepaired
The Systemic Right Ventricle

• Congenital Heart Disease with Systemic Right Ventricle:
  – Congenital Corrected Transposition (l-TGA)
  – Complete Transposition of the Great Arteries (d-TGA) following atrial switch procedure
    • Senning Procedure
    • Mustard Procedure
I-TGA; Congenitally Corrected Transposition
d-TGA; Complete Transposition of the Great Arteries

1. Aorta arising from the right ventricle.
2. Pulmonary artery arising from the left ventricle.
Atrial Switch Procedures

Intra-atrial baffle
(mustard or Senning procedure)

Aorta

Pulmonary Artery

Baffle
Sequelae of a systemic RV

- RV dysfunction
- Tricuspid insufficiency
- Arrhythmias
- Pulmonary Hypertension

Gatzoulis et al; Late arrhythmia in adults with the Mustard procedure for transposition of great arteries: a surrogate marker for right ventricular dysfunction? Heart 2000,84(4) 409-415
Anesthetic Management Outcomes

- 71/2 year period all anesthetics at U of M
- 5/45 patients had atrial switches
- All 5 had arrhythmias/significant ongoing cardiac dysfunction
- 3/5 had arterial lines placed
- All had non pediatric anesthesiologists
- No complications

Should I feel comfortable?

Yes:
If no failure, no problem. RV failure; treat like systemic ventricular failure
May have conduction abnormalities
The Single Ventricle

- HLHS
- Tricuspid atresia, HRH
- Unbalanced AV canal
Tricuspid Atresia
Norwood
The Norwood Procedure

By Tom Diab
Hemifontan
Hemi-Fontan

By: Tom Diab
Fontan
Fontan

By: Tom Diab
Sequelae of a Single Ventricle

- Right atrial enlargement, hepatic dysfunction
- Systemic venous collateralization
- Atrial arrhythmias
- Venous stasis
- Protein losing enteropathy (PLE)
- Cyanosis

Driscoll DJ, Long-Term Results of the Fontan Operation. Pediatric Cardiol 2007, 28:438-442.
Differential Dx of Cyanosis in Fontan Patients

- Patent Surgical Fenestration
- Baffle Leak (deoxygenated blood from systemic veins contaminates left atrial pulmonary venous blood)
- Systemic venous collateralization to left side
- Pulmonary AVM’s
- Hepatic veins to Coronary sinus or LA
- Intrinsic pulmonary pathology
- Diaphragm paralysis

Driscoll DJ, Long-Term Results of the Fontan Operation. Pediatric Cardiol 2007, 28:438-442
Should I feel comfortable?

Probably not:
These patients should be anesthetized by a Pediatric Cardiac Anesthesiologists at a specialty center. (my opinion)
Left to Right Shunts

- Ventricular Septal Defect (VSD)
- Atrial Septal Defect (ASD)
- Patent Ductus Arteriosus (PDA)
- AV Canal (AVSD)
- Total Anomalous Pulmonary Venous Return (TAPVR)
**Left to Right Shunts**

- volume overloaded ventricles
- decreased cardiac reserve
- pulmonary venous congestion
- reduced lung compliance and increased airway resistance
- pulmonary vascular obstructive disease
Eisenmenger’s Syndrome

Over time left to right shunting causes

- Increased pulmonary blood flow
- Pulmonary vascular disease
- Acquired pulmonary hypertension
- Pulmonary pressures may exceed systemic pressures
Eisenmenger’s Syndrome

- Reversal of the shunt to right to left
- Same sequelae as unrepaired TOF
- End stage irreversible pulmonary disease
- \( \text{Qp/Qs} < 0.7 \)
- Poor prognosis (transplantation may be an option)
Poor Prognosis in Eisenmenger

- Syncope
- Hemoptysis
- NYHA Class III or IV
- Complex Congenital Heart Disease
- Sat < 85%
- RV dysfunction
- RVH on ECG
- Down Syndrome

Should I feel comfortable?

NO:
These patients should **ALL** be anesthetized by a Pediatric Cardiac Anesthesiologists at a specialty center.
Pregnancy and CHD

- As success of medical and surgical treatment of CHD improves more patients are surviving to childbearing years
- CHD is now the most common heart problem in women during pregnancy
- Anesthesiologist must understand how the physiologic changes of pregnancy affect the pathophysiology of the CHD
Pregnancy and CHD

• Common symptoms of late pregnancy can be similar to those for CHF
  – Dyspnea
  – Fatigue
  – Peripheral edema
Increased Cardiac Output in Pregnancy

- 2\textsuperscript{nd} and 3\textsuperscript{rd} trimester
  - Inc blood volume
  - Inc red cell mass
  - Inc heart rate
- Labor and delivery
  - Pain
  - Uterine contractions
- Immediately post partum
  - Relief of IVC compression
  - Auto transfusion from uterus

Siu SC, Colman JM, Heart Disease and Pregnancy, Heart 2001,85:710-715
Low Risk Pregnancies; Mortality < 1 %

- Small L to R shunts
- Repaired lesions without dysfunction
- Isolated MVP without regurgitation
- Bicuspid Aortic valve without stenosis
- Mild to moderate PS
- Valvar regurgitation with normal ventricular systolic function

Siu SC, Colman JM, Heart Disease and Pregnancy, Heart 2001,85:710-715
Moderate Risk Pregnancies; 1-5 % Mortality

- Unrepaired or palliated cyanotic CHD
- Large L to R shunt
- Uncorrected coarctation of the aorta
- Mitral or aortic stenosis
- Mechanical prosthetic valves
- Severe PS
- Moderate to severe systemic ventricular dysfunction
- Peripartum cardiomyopathy without ventricular dysfunction

Siu SC, Colman JM, Heart Disease and Pregnancy, Heart 2001,85:710-715
High Risk Pregnancies; 15-50% Mortality

• Severe pulmonary hypertension
• NYHA Class III or IV symptoms
• Severe aortic stenosis
• Marfan syndrome with aortic involvement
• Peripartum cardiomyopathy with ventricular dysfunction

Siu SC, Colman JM, Heart Disease and Pregnancy, Heart 2001,85:710-715
Pregnancy in TOF

- Increased risk of fetal loss
- Children more likely to have congenital anomalies than offspring in the general population
- Adverse maternal events, although rare, may be associated with left ventricular dysfunction, severe pulmonary HTN, and severe PR with RV dysfunction.

Pregnancy and Eisenmenger’s

- Pregnancy is contraindicated
- Maternal mortality approaches 50%
  - Thromboembolism (44%)
  - Hypovolemic (26%)
- Maternal deaths occur postpartum; as long as 7 - 25 days out
- Perinatal mortality is 28%
- Maternal mortality for pregnancy termination is 7%

Indications for C-Section in Patients with CHD

- Obstetrical concerns
- Aortic dissection
- R → L shunt with hypoxia and fetal distress
- Marfan’s syndrome with dilated aortic root
- Intractable arrhythmias
- Failure to switch from Coumadin to heparin 2 weeks prior to C-section

Siu SC, Colman JM, Heart Disease and Pregnancy, Heart 2001,85:710-715
Should I feel comfortable?

- Complex congenital heart disease
  Deliver in C&W, consult pediatric cardiac anesthesiology

- Adult Disease (Aortic disease, CHF, PPHtn)
  Deliver at UH/CVC with adult cardiac surgery and adult cardiac anesthesia backup
Summary; who should manage

– Anyone
  • Repaired TOF
  • Systemic RV

– A Pediatric Cardiac Anesthesiologist
  • Fontan Physiology
  • Eisenmengers
  • Complex Cyanotic Heart Defects
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