How would you do this anesthetic?

• 25 year old male presents with 2 day history of fever, vomiting, and abdominal pain. On exam, he has RLQ tenderness. CT confirms appendicitis. To the OR for appendectomy.
• The patient has a history of Tetralogy of Fallot.
What this talk will NOT do.

Objectives

Participants will

- Recognize the changing epidemiology of congenital heart disease, and the challenges it presents for the adult patient.
- Consider the pathophysiology and long term consequences of congenital heart disease.
- Examine general strategies for providing anesthesia to adults with congenital heart disease.
- Discuss complex congenital heart disease and its impact on adult patients

Physiologic Classification

Congenital Heart Disease

- Acyanotic CHD
  - With Left-to-Right Shunt
  - Without Left-to-Right Shunt
- Cyanotic CHD
  - Ductal-dependent Pulmonary Blood Flow
  - Ductal-dependent Systemic Blood Flow
  - Mixing Lesions

Acyanotic CHD

With L-to-R shunt
- Physiology
  - Volume overload in at least 1 cardiac chamber
  - Increase Pulmonary Blood Flow can lead to PHTN
- ASD
- VSD
- PDA
- PAPVR

Without L-to-R shunt
- Physiology
  - Increase pressure load
  - Limited CO/blood flow
  - Can lead to significant cardiac hypertrophy
- Valve disease
- Coarctation of Aorta
Cyanotic CHD

- Right-to-Left shunt
  - Venous admixture is added to the systemic circulation
- Ductal-dependent PBF
  - Tetralogy of Fallot
  - Pulmonary Atresia
  - Tricuspid Atresia
  - Single Ventricle
- Ductal-dependent SBF
  - Critical Coarctation of Aorta
  - Critical Aortic Stenosis
  - Hypoplastic Left Heart Syndrome
- Mixing Lesions
  - AVSD
  - Double Outlet Right Ventricle
  - Transposition of the Great Arteries
  - Truncus Arteriosus

- Transition at birth
  - Lungs inflate
  - Increase in oxygenation
  - Placental removal
- Leads to:
  - Decrease PVR
  - Increase PBF
  - Closure of ductus venosus
  - Constriction of ductus arteriosus
  - Reversal of pressure gradients
  - Closure of PFO

Series vs. Parallel Circulation

Series vs Parallel

Hypoxemia
Volume overload

Adult Congenital Heart Disease

- Most common congenital disorder of newborns
  - 1% of live births
- Advances in cardiovascular medicine and surgery
  - 85-95% survive to adulthood
  - 15-25% survival of patients prior to treatment availability

Changing Epidemiology: Mortality

Distribution of Age at Death in Patients With Congenital Heart Disease in 1987 to 1988 and 2004 to 2005. In 1987 to 1988, a significant increase in median age of death (2 years vs. 23 years) was observed. These changes reflect improvements in medical care and surgical techniques. The bar graph illustrates the age-specific death rates with a significant decrease in mortality at younger ages.
**Long term Survival by complexity of CHD**

<table>
<thead>
<tr>
<th></th>
<th>95%</th>
<th>90%</th>
<th>80%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Simple</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect (repaired, small, no assoc)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PFO</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild PS</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Atrioventricular canal defect</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Coarctation of the aorta</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Single ventricle palliation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anomalous pulmonary venous drainage</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eisenmenger syndrome</td>
<td></td>
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</tbody>
</table>

Long term survival > 20 years

*Marelli, Circulation 2007 & 2014*  
*Warnes, J Am Coll Cardiol 2008, Connelly, Canadian Consensus on ACHD, 1996, Can J Cardiol, 1998*

**U.S Estimates of CHD**

- 1 million children + 1.4 million adults with CHD living in the US in 2010
- 300,000 with severe disease; 160,000 adults
- 63% increase in Adult CHD from 2000-2010
- ACHD patients require ongoing care and interventions
- Possible need >200 ACHD centers of care (currently 114)

*Gilboa, Circulation, 2016*

**ACHD: Health Services Utilization**

Nationwide Inpatient Sample 1998 to 2005

- ACHD Hospitalizations increased 102%  
  (36,000 in 1998 to 72,000 in 2005)
- Average patient age was 53.8 years
- Patients with > 2 comorbidities increased 23% to 33%
- Leading indications for admission
  - Heart Failure
  - Arrhythmias
  - Coronary artery disease
- Estimated total national cost of these hospitalizations increased 357% to $3.16 billion

*Opotowsky, J Am Coll Cardiol 2009*  
*Rodriguez, Congenit Heart Dis 2013*

**Challenges of Adult Patients**

- 42% patients in N. America had gaps of >3 years in their care; (mean age 19 yrs. +/- 9 mos.)
- Life events: independence, jobs, marriage, family
- Common reason for gaps:
  - Geography
  - “feeling well”
  - “changing or losing insurance”
  - “financial problems”
  - “lost track of time”
  - “decreased parental involvement”
- Increased risk of developing cardiac s/sx
  - 3x more likely to require intervention

*Gurvitz, J Am Coll Cardiol, 2013*  
*Yeung, Int J Cardiol, 2008*

**Long term sequelae**

<table>
<thead>
<tr>
<th>Cardiac</th>
<th>Non-cardiac</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary hypertension</td>
<td>Secondary erythrocytosis</td>
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<tr>
<td>Ventricular dysfunction</td>
<td>Cholelithiasis</td>
</tr>
<tr>
<td>Dysrhythmias and conduction defects</td>
<td>Nephrolithiasis</td>
</tr>
<tr>
<td>Residual shunts - Cyanosis</td>
<td>Neurodevelopment abnormalities</td>
</tr>
<tr>
<td>Valvular lesions</td>
<td>CNS disease</td>
</tr>
<tr>
<td>Stenosis</td>
<td>Secura</td>
</tr>
<tr>
<td>regurgitation</td>
<td>CVT (thromboembolism)</td>
</tr>
<tr>
<td>Hypertension</td>
<td>Hearing/vision loss</td>
</tr>
<tr>
<td>Aneurysms</td>
<td>Chronic lung disease</td>
</tr>
<tr>
<td></td>
<td>Hepatic dysfunction, Cirrhosis</td>
</tr>
<tr>
<td></td>
<td>Renal impairment</td>
</tr>
<tr>
<td></td>
<td>Coagulopathy</td>
</tr>
<tr>
<td></td>
<td>Cancers</td>
</tr>
</tbody>
</table>

*Schwartz, Lawrence, MD Anesthesia for Adults with Congenital Heart Disease*

**Changing Epidemiology: Adults and children in Quebec, Canada, with all (A) and severe (B) CHD**

<table>
<thead>
<tr>
<th>Year</th>
<th>Median Age (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1985</td>
<td>11</td>
</tr>
<tr>
<td>2000</td>
<td>17</td>
</tr>
<tr>
<td>2010</td>
<td>26</td>
</tr>
</tbody>
</table>

*Marelli, Circulation 2007 & 2014*
Chronic Cyanosis

- Insufficient pulmonary blood flow
  - Pulmonary Atresia
  - RVOTO
  - Pulmonary arteries
  - PVR

- Contamination of arterial blood by venous blood
  - Right-to-Left Shunt
  - Increased venous admixture

Physiology of chronic cyanosis

- Hypoxemia $\rightarrow$ Erythropoietin $\rightarrow$ Increase mass and stiffness of RBCs
  - Polycythemia is attempt to increase delivery of oxygen

- Increased hematocrit (50-75%) $\rightarrow$ Hyperviscosity
  - Increase shear stress at the vascular endothelium
  - Endothelial dysfunction

- All organ systems are affected

Cyanosis effects on organ systems

- CV
  - Myocardial ischemia risk (myocardial perfusion, micro-occlusion)
    - HCT>65% may decrease DO$_2$
    - Hyperviscosity increase myocardial work
  - Endothelial injury $\rightarrow$ imbalance of vasodilators/vasoconstrictors;(NO, Prostacycline) $\rightarrow$ BP, myocardial work, perfusion
  - Myocardial gene expression alterations
    - Up-regulation of genes assoc with apoptosis, remodeling
    - Down-regulation of genes assoc with myocardial contractility
  - Risk of systolic and diastolic dysfunction

- Coagulation
  - Bleeding risk
    - Decrease von Willebrand factor
    - Relative thrombocytopenia, decrease PLT life-time & function
  - Thrombosis risk – stiff RBC

- Neurologic
  - Cerebral abscess (R-HL shunt)

- Respiratory
  - Blunted hypoxic ventilatory drive
  - Hyperventilation
  - ETCO$_2$ may underestimate P$_{CO_2}$(R-to-L shunts)

- Renal
  - Chronic hypoxemia proliferative lesions in glomeruli
  - Thickening of basal membrane
  - Proteinuria, increase uric acid

Anesthetic Implications

- NPO strategies
  - Dehydration can worsen hyperviscosity
  - Avoid prolong fasting, consider IV fluids

- Opioid use can lead to profound hypoxemia
  - Careful post-op monitoring

- Phlebotomy
  - Not routine
  - Perhaps some benefit with HCT>65%

- Manage the shunt
  - SVR/PVR manipulation

- Maintain myocardial function
**Pulmonary Hypertension**

- Mean pulmonary artery pressure > 25 mmHg at rest
- Pulmonary vascular resistance index > 3 Wood units/m²
- Pulmonary venous congestion
  - Obstruction
  - High left atrial pressures
- Large, unrestricted L-to-R shunt
  - Increase flow through the pulmonary vasculature
  - Exposure of pulmonary vasculature to systemic pressures

---

**Vascular changes**

- Hypertrophy of arterial/arteriolar media
- Intimal cellular proliferation
- Smooth muscle cell migration to subendothelium
- Progressive fibrosis
- Obliteration of arterioles and small arteries

Heath & Edwards, Circulation, 1958

**Pulmonary Hypertension**

- Vascular changes may be reversible in early childhood
- Become progressive and permanent in adults
- Eisenmenger Syndrome
  - Severe, fixed PHTN, equalization of RV and LV pressures
  - L-to-R shunt can reverse to become R-to-L, with RV dysfunction and severe hypoxemia
  - Anesthesia is a bad idea in this patient – high mortality
  - Should become more rare

---

**Anesthesia & PHTN**

- Pulmonary HTN + CHD = High Risk Anesthetic
- Primary goal of anesthetic is to minimize PVR and maintain SVR
- Prevention and treatment of acute pulmonary hypertensive crisis

---

**Anesthetic principles**

- Continue home meds
  - Sildenafil, bosentan, prostacycline, etc...
- Avoid & correct respiratory or metabolic acidosis
- Avoid sympathetic nervous system stimulation, maintain appropriate pain control and depth of anesthesia
- Adequate oxygenation
- Maintain normothermia
- Inotrope/Vasopressor support as needed
- Pulmonary vasodilatation
  - Milrinone
- Inhaled Nitric Oxide availability
- Regional analgesia
  - Peripheral nerve blocks okay
  - Caution with neuroaxial blockade

---

**Ventilatory management & PHTN**

- Careful attention to lung volumes and intrathoracic pressure
- Want to be in the Goldilocks zone = FRC
Acute pulmonary hypertensive crisis

- Hyperventilation
- \( \text{FiO}_2 = 1.0 \)
- \( \text{NaHCO}_3 \)
- Adequate depth of anesthesia
- Systemic vasoconstriction
  - Shunt L-to-R
  - Maintain coronary perfusion
- Support Cardiac Output
  - Inotropic support
  - Maintain preload
- Pulmonary vasodilator therapy
  - ECMO

Treatment for PHTN crisis

Dysrhythmia

- Atrial and ventricular dysrhythmias are common
  - Repaired Tetralogy of Fallot
  - TGA
  - Single ventricle Fontan completion
  - Congenital conduction abnormalities
  - Supraventricular dysrhythmias occur in 20-45% of patients with previous atrial surgery
  - Intraatrial reentrant tachycardia is most common
  - Heart block occurs with VSD involvement
  - Arrhythmia may not be well tolerated in a patient with borderline or depressed function

Factors leading to arrhythmias in CHD

- Arrhythmia may not be well tolerated in a patient with borderline or depressed function
- Abnormal rhythms may be refractory to medication

Pre-op evaluation

ASA recommendations

- Type of device
  - Device information card
  - Single/dual chamber, mono/bi-polar
- Dependency on device / Escape rhythm
  - Symptomatic bradycardia, syncope, recent AVN ablation
  - % paced
- Device function
  - Battery life, impedance, thresholds,
  - Interrogation is never a bad idea

Many ACHD patients will require permanent pacemaker or AICD
Electromagnetic interference

- Sources of EMI
  - Electrocautery
  - Transthoracic defibrillation
  - Therapeutic radiation
  - RFA
  - Shock wave lithotripsy
  - MRI
  - Nerve stimulators
  - Fasciculations
  - Shivering
  - Large tidal volume ventilation

- Effects of EMI
  - Inappropriate triggering
  - Overpacing
  - Inhibition of triggering
  - Asynchronous pacing
  - ICDs
  - Inappropriate shock
  - Lead current
  - Electrical discharge to the myocardium
  - Arrhythmia, burns

Reducing EMI risk

- Bipolar diathermy > Monopolar diathermy
  - If monopolar must be used
    - Cutting > Coagulation (less power)
    - Short, intermittent, irregular (< 1 sec) bursts
  - Good skin contact with diathermy grounding pad
    - Keep the pad as far from the device as possible
    - But, don’t cross the streams

Magnet

- They are made for interrogation, not emergencies. But...
  - Magnet over the pacemaker will switch to asynchronous pacing – avoid oversensing the bovie & asystole
  - Manufacturer determined rate
    - May not be enough cardiac output for patient
    - May decrease with waning battery life
    - May stop pacing altogether
  - AICD
    - In general, will turn off anti-tachycardia therapy
    - Avoid oversensing the bovie and inappropriate shock

Anesthesia

- Commonly used anesthetic agents generally do not directly effect pacemaker function, but...
  - Elevated lead threshold and failure to capture
    - Metabolic or ischemic changes
      - Hyperkalemia
      - Acidosis
      - Alkalosis
      - Hypothermia
      - Hyperglycemia
      - Hypoxemia
      - Hypocarbia
    - Large fluid shifts, dehydration, bleeding

Pathophysiology of Heart Failure in CHD

- Increased catecholamines
- Chronic pressure overload
- Renin-angiotensin-aldosterone system
- Geometric changes
- Hypertrophic myocardium
- Abnormal perfusion
- Chronic volume overload
- Abnormal fetal/Neonatal development
- Chronic cyanosis
- Abnormal architecture
Additional Factors

- Lesion specific issues (RV systemic ventricle)
- Previous surgery
  - Ischemic injury
  - Early life cardiopulmonary bypass
  - Early coronary artery disease
  - Hypertension
  - Diabetes
  - EtOH Abuse

Ventricular Dysfunction in ACHD

- Very common, 25% of all CHD have heart failure
  - Single V 50%, TOF 50%, TGA (atrial switch) 33%

- Significant mortality and morbidity
  - Heart Failure was leading cause of death in 26% of > 8000 ACHD patients, 1998-2005
  - Hospital admissions increase 100%
  - Exercise intolerance reported by 50% ACHD

ACHD Heart Failure

- Single Ventricle palliated to Fontan
  - Single ventricle has both the systemic and pulmonary resistances in series

- Tetralogy of Fallot
  - Pulmonary regurgitation causes RV dilation and dysfunction, abnormal septal configuration, altered RV/LV interaction and LV dysfunction

Subsystemic RV

1. D-TGA palliated with a Mustard/Senning atrial switch
2. ccTGA

Preoperative considerations

- History
  - Current vs. Baseline clinical state
  - Exercise tolerance
  - Prior anesthetics
- Medical therapy
  - Common drugs: ACE Inhibitors, Diuretics, Beta-blockers
  - Can lead to significant hypotension when combined with anesthesia
- ECG, Imaging, Labs
- Cardiology “consult”
- Inpatient issue
  - Inotrope, mechanical ventilation

Anesthetic considerations

- Overall goal is a balanced anesthetic that maintains myocardial oxygen supply with myocardial oxygen consumption

\[ \text{DO}_2 = \text{VO}_2 \]
Supply and Demand

- Atrial and Ventricular end-diastolic pressures elevated
- CO is very sensitive to filling
- Systemic vascular resistance
  - Afterload reduction improves work and CO
- Positive pressure ventilatory support
  - Decrease VR, preload, CO
  - Afterload reduction improves transmural gradient
- Anesthetic Agents
  - Too deep → hypotension and worsening
  - Too light → increase SVR and myocardial oxygen consumption

Balance

- A balanced, carefully titrated anesthetic is the key to hemodynamic stability
  - There is no general recipe, or magic regimen
  - How to use the drugs >> Which drugs to use

- Monitoring
  - Arterial line, Central Venous Pressure, Pulmonary Arterial Catheter
  - Transesophageal echocardiography

- Hope for the best, prepare for the worst
  - Resuscitation, Inotropes
  - ECMO

Infective bacterial endocarditis

- The pathophysiology is multifactorial, but dependent on valvular or mural endocardial damage and bacteremia.
  - Endothelial injury
  - Platelet and fibrin deposition
  - Leads to nonbacterial thrombotic endocarditis.
  - A transient bacteremia provides the source for bacterial adherence and proliferation within the vegetation.
- Cyanotic heart disease, shunts, foreign materials implants

Infective endocarditis is associated with high morbidity and mortality
But it is exceedingly rare
More restrictive use of antibiotics

Table I. Agents for Dental Procedures

<table>
<thead>
<tr>
<th>Situation</th>
<th>Drug</th>
<th>Adult dose (mg/kg)</th>
<th>Pediatric dose (mg/kg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral</td>
<td>Penicillin</td>
<td>2 g</td>
<td>50 mg (0.5 mg/kg)</td>
</tr>
<tr>
<td>After surgery and medications</td>
<td>Ciprofloxacin</td>
<td>2 g</td>
<td>5 mg (0.5 mg/kg)</td>
</tr>
<tr>
<td>After surgery and medications</td>
<td>Clindamycin</td>
<td>900 mg</td>
<td>20 mg (mg/kg)</td>
</tr>
<tr>
<td>After surgery and medications</td>
<td>Azithromycin</td>
<td>500 mg</td>
<td>15 mg (0.5 mg/kg)</td>
</tr>
<tr>
<td>After surgery</td>
<td>Ciprofloxacin</td>
<td>1 g</td>
<td>5 mg (0.5 mg/kg)</td>
</tr>
<tr>
<td>After surgery and medications</td>
<td>Clindamycin</td>
<td>1 g</td>
<td>5 mg (0.5 mg/kg)</td>
</tr>
<tr>
<td>In Holter Electrocardiography</td>
<td>Clindamycin</td>
<td>900 mg</td>
<td>20 mg (0.5 mg/kg)</td>
</tr>
</tbody>
</table>

antibiotic recommendations

Infective endocarditis is associated with high morbidity and mortality
But it is exceedingly rare
More restrictive use of antibiotics

Cardiac Conditions Associated with the Highest Risk of Adverse Outcome From Endocarditis for Which Prophylaxis with Dental Procedures is Recommended

- Prosthetic cardiac valve
- Previous infective endocarditis
- Congenital heart disease (CHD)
  - Unrepaired cyanotic CHD, including those with palliative shunts and conduits
  - Completely repaired CHD with prosthetic material or device either by surgery or cellular intervention during the first 6 months after repair
  - Repaired CHD with residual defects at the site or adjacent to the site of a previous patch or prosthetic device which-related endocarditis
- Cardiac transplantation recipients who develop cardiac allograft rejection
- Cyanotic heart disease
- Shunts
- Foreign materials

Schwartz, Lawrence, MD
Anesthesia for Adults with Congenital Heart Disease

Tetralogy of Fallot
Transposition of the Great Arteries
Fontan Palliation for Univentricular Defects
Modified Blalock-Taussig Shunt

TOF is the most common form of Cyanotic CHD
10% of CHD

Untreated mortality
33% by 1 year
50% by 3 years
98% by 4th decade
Bertrandou, Am J Cardiol, 1976

Complete repair with Transannular Patch

TOF is the most common form of Cyanotic CHD
10% of CHD

Adults with TOF
• With current medical and surgical management, survival to adulthood is 85%
• Many live good quality of life
• But repair is not a cure
• Common long term sequelae of TOF:
  - Obstruction to pulmonary blood flow and/or
  - Pulmonary regurgitation → RV enlargement and dysfunction
  - Secondary tricuspid regurgitation
  - Heart failure with worsening exercise tolerance
  - Supraventricular & ventricular tachyarrhythmias
  - Sudden Death
Bertrandou, Am J Cardiol, 1976
Nollert, J am Coll Cardiol, 1997
Gatzoulis, Lancet, 2000

Adults with TOF and RV dysfunction
• Pulmonary Valve Replacement is indicated to maintain or improve quality of life, and lower risk for sudden death
• Risk of sudden death due to arrhythmia associated with older age of repair, incomplete correction, RV overload & dysfunction
• Many patients may be asymptomatic until RV dysfunction leads to LV dysfunction
• Timing of surgical or transcatheter PVR is essential
  - Too late → risk of permanent injury to the RV
  - Too early → on the clock for reoperation
  - MRI to evaluate RV size + end-diastolic volumes
Nollert, J am Coll Cardiol, 1997
Murphy, NEJM, 1993
Gatzoulis, Lancet, 2000

Sudden Death & Arrhythmia
• Long term prevalence of sudden death is 3.98%, reaching 8.3% at 35 years follow-up
• More likely to occur in patients with correction at older age, multiple interventions, and transannular patch
• QRS > 180 ms; increase QRS duration 3.5 ms/year
Nollert, J am Coll Cardiol, 1997
Murphy, NEJM, 1993
Gatzoulis, Lancet, 2000

Anesthetic Considerations
• Volume overload – chronic PR
  • Manage RV dysfunction
  • Maintain lower pulmonary vascular resistance
• Pressure overload – chronic RVOTO
  • Manage RV dysfunction
  • Maintain adequate systemic vascular resistance to ensure subendocardial perfusion
• Arrhythmias/AV Block
  • Manage pacemakers, AICDs
  • Avoid, manage metabolic derangements
• Uncorrected TOF – quite rare today
  • Maintain SVR, keep PVR low, maintain flow through RVOTO
  • Avoid and treat “Tet Spell”
  • Palliated TOF
  • Balance of parallel circulations (Qp:Qs)

D-Transposition of the Great Arteries
Mustard Repair
Jatene Arterial Switch Operation

Ventriculoarterial Discordance = Parallel circuit
Survival dependent on mixing (ASD, VSD)
Cannot survive to adulthood without surgery
**Atrial Switch – Mustard procedure**

- Intra-atrial baffle
- Systemic RV
- Fails in about 25 years
- Baffle obstruction
- Baffle leak
- Failing systemic RV
- Atrial tachyarrhythmia

**Jatene Arterial Switch Operation**

- Neonatal surgery
- Life expectancy 97% at 10 years
- Hemodynamics close to normal
- 25% with neo-Aortic valve regurgitation
- Myocardial ischemia due to coronary ostia issues

**Congenitally corrected TGA, L-TGA**

- Vентriculoarterial discordance
  AND Atrioventricular discordance
- Circulation is in series
- RV is systemic Ventricle
  - Pressure overload
  - RV failure in 3rd decade
  - VSD, AV-valve abnormalities
  - AV Block is common
  - May present in adulthood

**Definition of Single Ventricle**

- Anatomically heterogeneous group of defects
- Parallel circulation
- Universally fatal in infancy without intervention
- Physiologically, single ventricle is any defect that progresses to Fontan Palliation

**STAGE 1**
- Cavopulmonary Shunt
  - Modified Bidirectional Glenn
  - Low Qp Norwood Procedure
  - MBTS

**STAGE 2**
- Cavopulmonary Shunt

**STAGE 3**
- Total Cavopulmonary Shunt
  - Fontan Completion

- Extracardiac Conduit has become the preferred surgical technique
  - Improved flow hemodynamics
  - Less risk of atrial thromboembolism
  - Less atrial dysrhythmia, sinus node dysfunction
  - Smoother post-op course

But no real improvement in 10 year survival of ECC over LT

*Khairy, Circulation, 2012*
Goals of Univentricle Palliation

- Stabilize pulmonary and systemic blood flow
- Bring parallel circulation to a series circulation in 3 stages
- Unload the ventricle
- Correct hypoxemia
- No obstructions, regurgitations, (shunt)

Fontan Physiology

- The is no pulmonary ventricle
- All pulmonary blood flow is non-pulsatile, venous pressure
  - Decrease of venous capacitance
  - Increased systemic pressure
  - Increase PVR
  - Decrease endothelial NO

The entire cardiac output is dependent on pulmonary blood flow.

Transpulmonary gradient ideally 5-10 mmHg

Kelly, Am J Cardiol, 1995; Krishnan, Circulation, 2009; Bove, Ped Cardiol, 2007; Redington, Ped Cardiol, 2006

Palliation, not cure

- Ventricular Failure
- Atrial Arrhythmias
- Sinus/AV nodal dysfunction
- Cyanosis
  - Fenestration, A/M, collateral vasculature
- Obstruction
- AV/Valvulopathy
- Pulmonary hypertension
- Exercise intolerance
- Heart Transplantation
- Thromboembolic complications
- Protein Losing Enteropathy
- Varocies, GI ulcers
- Intrinsic lung disease
- Plastic Bronchitis
- Congestive Hepatopathy
- Renal insufficiency
- Psych-social disorders
- Adult co-morbidities
  - Diabetes, hypertension, obesity, hyperlipidemia

Anesthesia for Fontan Adult

- #1 Ensure an adequate trans-pulmonary pressure gradient
  - SVC or IVC obstruction
  - High pulmonary venous pressure due to high LVEDP,
  - AV valve stenosis or regurgitation, or loss of sinus rhythm
  - Pressure gradient decreases and there will be poor forward blood flow through the lungs.
  - Low CO associated with hypotension and hypoxia quickly ensues.

Preoperative Assessment

- History with focus on type of Fontan
  - Right vs. Left ventricle
  - Fenestration?
- Exercise tolerance
  - Reserve capacity < 50% age match controls
  - Recent illness URI
  - May increase airway resistance, PVR to pose unacceptable risk
- Exam
  - Oxygen saturation – not just about the lungs; CV function too
  - Signs of systemic venous congestion
- Coagulation studies
  - Hypercoagulable state vs. anticoagulation Rx
- Echocardiogram
- Catheterization

Anesthetic Management of Fontan

- Premedication is often desirable
  - This is a well experienced, anxious group of patients
  - Again, no specific agent or technique in recommended
  - Caution with neuroaxial blockade
    - Sympathectomy → reduction of preload
  - IV induction with attention to myocardial depression if indicated
    - Ketamine, Dexmedetomidine, Opioids, Etomidate
**Effect of mechanical ventilation**

- 40% increase in PBF with return of PPV
- Mean increase in PBF 38%

Fontan circulation prefers spontaneous ventilation/negative intrathoracic pressure. Positive pressure and PEEP can reduce, stagnate, or reverse Fontan flow.

**Ventilation strategy for adult Fontan**

- Spontaneous ventilation may be advantageous.
- But under anesthesia, hypoventilation with hypercarbia, hypoxia, & atelectasis can lead to increased PVR.
- Goal is to maintain adequate ventilation, while minimizing mean airway pressure.

**Cardiovascular Strategies**

- Maintain transpulmonary gradient, keep EDP low and maintain adequate preload.
- NPO, prolonged fasting, dehydration decrease CO.
- Avoid volume overloading in ventricle with diastolic dysfunction.
- Afterload reduction, but maintain coronary perfusion.
- Maintain sinus rhythm.
- Adverse effects of laparoscopy.
- Prepare for bleeding.
- Coagulopathy, ASA, Warfarin.
- Monitoring.
  - Arterial line, CVP pressures, SvO₂.
  - TEE – assessment of function and filling.

**Pregnancy with CHD**

Annual deliveries for women with CHD increased 35% from 1998-2007.

CHD is most common heart disease among pregnant women.

Most women with CHD will do well with pregnancy.

But certain risk factors do exist, particularly in complex CHD.
Normal CV changes in pregnancy

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Change</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blood Volume</td>
<td>Increases by 30%</td>
</tr>
<tr>
<td>Plasma Volume</td>
<td>Increases by 45%</td>
</tr>
<tr>
<td>Cardiac Output</td>
<td>Increases by 30-50%</td>
</tr>
<tr>
<td>Stroke Volume</td>
<td>Increases by 25%</td>
</tr>
<tr>
<td>Heart Rate</td>
<td>Increases by 15-25%</td>
</tr>
<tr>
<td>Peripheral Vascular Resistance</td>
<td>Decreases by 15-20%</td>
</tr>
<tr>
<td>CVP</td>
<td>Unchanged</td>
</tr>
</tbody>
</table>

Can lead to pulmonary overload in L→R shunts
Increased shunting & cyanosis in R→L shunts

Maternal Cardiac Event Risk

- Scoring
  - 0 predictors <10% risk
  - 1 predictor = 25-30% risk
  - 2+ predictors 75-100% risk

- In highest risk patients, pregnancy is discouraged
  - Severe LVOTO
  - Marfan’s Disease with aortic diameter > 44 mm
  - Systemic ventricular dysfunction, NYHA 3-4, EF<40%
  - Severe PHTN
  - High risk of mortality

Maternal Predictors of Primary Cardiac Event

<table>
<thead>
<tr>
<th>Complications</th>
<th>Predictor</th>
<th>OR (95% CI)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac</td>
<td>Prior cardiac event or aortic stenosis</td>
<td>8.0 (1.4-46)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Neonatal</td>
<td>NYHA class III or IV or cyanosis</td>
<td>3.1 (1.1-8.3)</td>
<td>0.036</td>
</tr>
<tr>
<td>Left heart obstruction</td>
<td>6.5 (1.9-21.6)</td>
<td>0.006</td>
<td></td>
</tr>
<tr>
<td>Systemic ventricular dysfunction</td>
<td>3.1 (1.1-5.9)</td>
<td>0.049</td>
<td></td>
</tr>
</tbody>
</table>

Maternal predictors of neonatal events

- Prematurity
- Low birth weight
- Respiratory Distress Syndrome
- Intraventricular Hemorrhage
- Infant mortality 4%

Labor & Delivery

- Lesion specific
  - Volume overloaded hearts do better than pressure loaded
  - Vaginal delivery preferred (with assist to minimize valsalva)
  - Carefully titrated epidural anesthetic with judicious use of crystalloids
  - Lowering SVR will favor regurgitant lesions; TOF with PR
  - Spinal anesthesia unsuitable for ACHD
    - Sudden hemodynamic change
      - Absolutely contraindicated for moderate/complex CHD
      - Reverse L-to-R shunts; worsen R-to-L shunts
  - Caesarian sections typically performed under GA
    - Ascending Ao >45mm; PTL with po anticoagulation; Symptomatic AS; Severe heart failure

Summary

- Improvements in medical and perioperative care have significantly improved outcomes in Congenital Heart Disease.
- 85% of patients with CHD survive to adulthood. This has created an Adult CHD population that is larger than the pediatric CHD population. And it is growing rapidly.
- Most patients will continue to need medical and surgical interventions throughout their lives. Patients with moderate to complex CHD may have serious long term complications from the CHD.
- Anesthesiologist must be familiar with the physiology of ACHD in order to safely care for these patients in the perioperative setting.
- There are no evidence-based guidelines for the anesthetic care of adults with CHD. However, it is known that ACHD patients are a higher risk for anesthesia complications. The clinical state of each patient must be considered carefully in order to provide a safe anesthetic plan.
Shaun White
2 time Olympic Gold Medal winner
15 X-Games Gold Medals
23 X-Games Medals
Tetralogy of Fallot

Think snow!