Born Blue

Anesthesia and CHD

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Disclosures

- None to Report
Objectives

- Review all congenital defects in which the patient is “blue”
- Describe physiology of the single ventricle patient
- Identify the 3 stages of repair for the single ventricle patient
- Describe the anesthesia implications of each stage
Congenital Heart Disease (CHD)

- Most common birth defect
- Occurs in 1 out of 115 – 150 live births
- 40,000 births per year
  - 4,800 babies with CCHD (Critical Congenital Heart Disease)
- 1/13 infant deaths due to CHD
- CHD mortality decreased 33% from 1996 – 2006

March of dimes website, 2014
American Heart Association
Adult Congenital Heart Disease

• 1.3 million patients alive today with CHD
  – 2-3 million estimated by the adult congenital heart association

American Heart Association website, 2014
Adult Congenital heart association website, 2014
Health Care Admissions

Figure 1: Annual Number of ACHD Admissions in the U.S. Categorized by Level of Defect Complexity

Opotowsky, 2009
Health Care Costs

- Costs increased 127% (1998 to 2005)
- Individual admissions $19,000 to $43,000
- Estimated national costs increased from $691 million to $3.16 billion

Opotowsky, 2009
Causes of CHD

• Mom with History
  – Diabetes
  – Lupus
  – Rubella
  – Obesity
  – Phenylketonuria (PKU)

• Other contributing factors
  – Smoking
  – Alcohol

March of dimes website, 2014
Causes of CHD

- 30% of CHD patients also have a chromosomal defect
  - Downs
  - Turners
  - Noonan
  - Velocardiocraniofacial syndrome (VCF)
  - Allagille syndrome

March of dimes website, 2014
Genetics and CHD

• Chromosomal abnormalities 10%
  – 2/3-trisomy 21
  – 1/3-others
    • Trisomy 13
    • Trisomy 18
    • Turners syndrome
    • 22q11.2 deletion-conotruncal defects (TOF, aortic arch, truncus, VSD)

CDC website
Genetics and CHD

- Other 90% no specific genetic association
  - Environmental
    - Rubella, ethanol, lithium, maternal diabetes, folate deficiency, obesity, smoking
Most Common Types of CHD

- Ventricular Septal Defect (14 -16%)
- Atrial Septal Defect (4 -10%)
- Tetralogy of Fallot (9 -14%)
- Coarctation of the Aorta (8 -11%)
- Transposition of the Great Vessels (10 -11%)
- Single ventricle (4 -8%)
Critical Congenital Heart Disease

- Hypoplastic Left Heart Syndrome (HLHS)
- Pulmonary Atresia (PA)
- Tetrology of Fallot (TOF)
- Total Anomalous Pulmonary Venous Return (TAPV, TAPVR)
- Transposition of the Great Arteries (TGA)
- Tricuspid Atresia (TA)
- Truncus Arteriosus

March of dimes website, 2014
## Classification of Congenital Heart Disease (CHD) for Anesthesiologists

<table>
<thead>
<tr>
<th>Category</th>
<th>Examples</th>
<th>Characteristics</th>
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<tr>
<td>Left-to-right shunt lesions: two ventricles</td>
<td>VSD, ASD, PDA; aortopulmonary window; partial atrioventricular canal; partial anomalous pulmonary venous return</td>
<td>Acyanotic</td>
</tr>
<tr>
<td>Right-to-left shunt lesions: two ventricles</td>
<td>Tetralogy of Fallot; pulmonary atresia with VSD, pulmonary atresia with intact ventricular septum; double-outlet right ventricle; Ebstein’s anomaly</td>
<td>Cyanotic</td>
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<tr>
<td>Complete-mixing two-ventricle lesions</td>
<td>Dextrotransposition of the great arteries; total anomalous pulmonary venous return; truncus arteriosus; complete atrioventricular canal</td>
<td>Cyanotic; level of cyanosis depends on communications at atrial, ventricular, and great vessel levels</td>
</tr>
<tr>
<td>Complete-mixing single-ventricle lesions</td>
<td>HLHS, tricuspid atresia, other forms of univentricular heart</td>
<td>Cyanotic; complete mixing of pulmonary and systemic venous return; level of cyanosis depends on systemic/pulmonary blood flow ratio and degree of mixing</td>
</tr>
<tr>
<td>Obstructive lesions without shunting</td>
<td>Aortic stenosis, mitral stenosis, pulmonic stenosis; coarctation of aorta; interrupted aortic arch; cor triatriatum; hypertrophic cardiomyopathy</td>
<td></td>
</tr>
<tr>
<td>Regurgitant lesions without shunting</td>
<td>Aortic insufficiency, mitral insufficiency, pulmonic insufficiency, tricuspid insufficiency</td>
<td></td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td>Dilated cardiomyopathy, myocarditis, anomalous origin of left coronary artery from pulmonary artery; post-cardiac transplant coronary artery vasculopathy</td>
<td>Anatomically “normal” with decreased ventricular function</td>
</tr>
</tbody>
</table>
Left to Right Shunts

- VSD, ASD, AP window, partial AV canal, PAPVR
- Acyanotic

Shunts

- Left-to-right

- \( \downarrow \) Systemic perfusion
- Low cardiac output
- Hypotension
- LV failure
- LV volume load

- Avoid increase in SVR
- Avoid decrease in PVR
- \( \uparrow \) \( \text{FiO}_2 \) (hypoxic gas mixtures if needed)
- Avoid hyperventilation

Anesthesia and Uncommon Diseases, 2012
Right to Left Shunts

- TOF, PA with VSD, PA with IVS, DORV, Ebsteins’ anomaly
- Cyanotic
Complete Mixing Lesions

- DTGA, TAPVR, TA, Complete AV canal
- Cyanotic, level depends of communications

**Hemodynamic consequences**
- Hypoxemia (varying degrees)
- Qp/Qs very dependent on PVR and SVR
- Elevated hematocrit and hyperviscosity

**Hemodynamic goals**
- Adjust PVR/SVR for optimal Qp/Qs and oxygen saturations
  - ↓Fio2 and ↑Paco2 if Qp/Qs is high
- Improve mixed venous saturation
- Optimize tissue O2 delivery
Obstructive Lesions without shunting

- AS, MS, PS, Coarctation, IAA, Cortriatrium, hypertrophic cardiomyopathy

Hemodynamic consequences
- ↓ Pulmonary flow
- Hypoxemia
- RV hypertrophy
- RV dysfunction
- Tricuspid regurgitation

Hemodynamic goals
- Avoid increase in PVR
- Avoid decrease in SVR
- Hyperoxia
- Avoid hypoventilation
- Maintain preload
- Maintain ductal patency*
  (L → R shunt)

Obstructive lesions

Right-sided
- ↓ Systemic perfusion
- Low cardiac output
- Hypotension
- LV failure
- ↓ Coronary perfusion

Left-sided
- Avoid decrease in SVR
- Avoid decrease in PVR
- Maintain preload
- Maintain ductal patency*
  (R → L shunt)

* In ductus arteriosus-dependent CHD lesions

Anesthesia and Uncommon Diseases, 2012
Regurgitant lesions without shunting

- AI, MI, PI, TI

Hemodynamic consequences
- RV volume load
- RV enlargement
- RV dysfunction
- Elevated CVP
- Potential for R → L atrial shunt with high CVP/RAP

Hemodynamic goals
- Decrease RV afterload/lower PVR
  - Hyperoxia
  - Avoid hypoventilation
  - Increase heart rate
  - Enhance RV function

Right-sided
- LV volume load
- LV enlargement
- LV dysfunction
- Low systemic perfusion
- Pulmonary hypertension
- Elevated LAP, PCWP

Left-sided
- Decreased LV afterload/lower SVR
- Increase heart rate
- Enhance LV function
Cardiomyopathy

- Dilated cardiomyopathy, myocarditis, anomalous LCA from pulmonary artery, post transplant cardiomyopathy

- Anatomically “normal” with decreased function
HLHS

- Fourth most common congenital heart defect
- 7.5% of newborns
- Some genetic inheritance?
Variations of Single Left Ventricle (HRHS)

- Tricuspid atresia
- Double inlet left ventricle
- Malaligned complete AV canal
- Pulmonary atresia with intact ventricular septum
Variations of Single Right Ventricle (HLHS)

– Mitral valve atresia
  • Hypoplastic left heart syndrome (HLHS)
  • Double outlet right ventricle

– Aortic valve atresia
  – HLHS
  – Large VSD and normal LV
  – Malaligned complete AV canal
  – Heterotaxy syndromes-pulmonary stenosis or atresia
Two ventricles with potential single ventricle physiology

- Tetrology of Fallot with pulmonary atresia
- Truncus arteriosus
- Total anomalous pulmonary venous connection (TAPVR)
HLHS
History of CHD surgery

• 1939: Robert Gross ligates a PDA, Boston Children's
• 1944: Alfred Blalock performs BT shunt for TOF, Johns Hopkins
• 1953: John Gibbon uses CPB to close an ASD, University of Pennsylvania
• 1960: Dwight Harken replaces Aortic Valve, Brigham and Women's Hospital
History of CHD surgery

• 1967: Christian Barnard performs first heart transplant, South Africa
• 1975: Adib Domingos Jatene performs arterial switch for d-TGA, Brazil
• 1977: William Norwood performs Norwood for HLHS, CHOP
• 1985: LL Bailey performs first successful neonatal heart transplant, Loma Linda, CA
History CHD at Mott

- Bove arrived 1987-began work on HLHS
- 850 CHD operations per year (about 50 Norwoods)
- Overall 95% survival rate
- 2nd largest Congenital Heart Center in the US
HLHS
Treatment Options

- Hospice care at home
- Transplant
- 3 stage Repair
Stages of Single Ventricle Repair

- Norwood
  - Hybrid
- Bidirectional Glenn or Hemi-Fontan
- Fontan
Classic Norwood
Norwood
Norwood BTS AP
Sano Shunt Variation
Sano Shunt Variation
Sano AP view
Sano lateral view
MRI Pictures

Sano conduit

BT shunt
Central AP
Central lateral
Fig 2. The hybrid stage 1 palliation. Branch pulmonary artery bands and a stent across the patent ductus arteriosus are placed at one procedure, while the balloon atrial septostomy is performed as a separate procedure.
Bidirectional Glenn Anatomy

Andropoulos, 467
Bidirectional Glenn Anatomy

Reconstructed outflow from right ventricle to aortic pathways

Superior cardiopulmonary shunt

Anesthesia and Uncommon Diseases, pg 123
BDG AP
BDG lateral
Left Glenn AP
Left Glenn lateral
Hemi-Fontan Anatomy

Field guide 264
Hemi-Fontan Anatomy
Hemi-Fontan Anatomy
Hemi-Fontan Anatomy
Hemi-Fontan Anatomy
Hemi AP
Hemi lateral
Fontan Anatomy

Superior vena cava
Right pulmonary artery
Right atrium
Lateral tunnel
Systemic ventricle
Inferior vena cava
Aorta
Fontan (lateral tunnel)
Fontan Anatomy
Fontan AP view
Fontan lateral
HLHS Physiology

Balancing the parallel circulations

Goals

• good cardiac output
• normal end-organ function
• normal systemic oxygen delivery
• normal blood flow
Normal or “Series”

Normal or series circulation

Right atrium → Right ventricle → Lungs → Left atrium → Left ventricle → Aorta
Parallel or “Balanced” Norwood and Hemifontan Stage

Blood flow to lungs/body depends on balance of PVR/SVR

Right and left atrium | Right and left ventricle | Aorta

PVR, pulmonary vascular resistance; SVR, systemic vascular resistance
HLHS (after Fontan completion)

Single ventricle circulation (e.g. total cavopulmonary circulation)

SVC

Pulmonary artery

Lungs

Common atrium

Common ventricle

Aorta

IVC, inferior vena cava; SVC, superior vena cava
Single ventricle

Complete mixing of systemic and pulmonary venous blood at atrial and/or ventricular levels

Oxygen delivery affected by

- Vasodilators
- Vasoconstrictors
- Hemoglobin (viscosity)
- pH & P\textsubscript{CO\textsubscript{2}}
- Pulmonary venous obstruction
- Lung volume

HR  Contractility

Preload  Afterload

SVR  PVR  Cardiac output

Excess pulmonary blood flow (↑ PVR ↑ SVR)
- Oxygen saturation > 85%
- Decreased lung compliance
- Hepatomegaly
- Ventricular dilation
- AV valve regurgitation may produce low cardiac output

Inadequate pulmonary blood flow (↑ PVR ↓ SVR)
- Oxygen saturation > 75%
- Cyanosis
- Myocardial hypoxemia may produce low cardiac output

High cardiac output
- High mixed venous oxygen content
- Oxygen saturation > 80%
- Good perfusion without hepatomegaly, ventricular dilation, or lactic acidemia

Low cardiac output
- Low mixed venous oxygen content
- Oxygen saturation < 80%
- Poor perfusion
- Lactic acidemia
Single ventricle

Complete mixing of systemic and pulmonary venous blood at the atrial and/or ventricular levels

Oxygen delivery affected by

Vasodilators
Hemoglobin (viscosity)
PH & PCO₂
PO₂
Pulmonary venous obstruction
Lung volume

SVR

PVR

Cardiac output

HR
Preload
Afterload
Contractility

University of Michigan
C.S. Mott Children's Hospital

Andropoulos, 458
Excess pulmonary blood flow (↓PVR ↑SVR)
- Oxygen saturation > 85%
- Decreased lung compliance
- Hepatomegaly
- Ventricular dilation
- AV valve regurgitation may produce low cardiac output

Inadequate pulmonary blood flow (↑PVR ↓SVR)
- Oxygen saturation < 75%
- Cyanosis
- Myocardial hypoxemia may produce low cardiac output

High cardiac output
- High mixed venous oxygen content
- Oxygen saturation > 80%
- Good perfusion without hepatomegaly, ventricular dilation, or lactic acidemia

Low cardiac output
- Low mixed venous oxygen content
- Oxygen saturation < 80%
- Poor perfusion
- Lactic acidemia
Anesthetic Planning

• Understand hemodynamic consequences of patient's lesion and state of repair.
• Construct a set of hemodynamic goals for each patient.
Anesthetic Planning

- Plan anesthetic agents and techniques, ventilatory management, and inotropic/vasoactive drug support based on these goals.

- Although no anesthetic agent or technique is contraindicated, avoid agents or doses counter to hemodynamic goals, and use agents that promote these goals.
Preoperative Assessment

- **History**
  - Cardiac lesion
  - Cyanotic
  - One or two ventricles
  - Septated, any intra atrial or ventricular communication
  - Surgery or cath lab procedures
  - Palliated vs corrected
  - Ventricular function
Preoperative Assessment

• History (cont)
  – Coronary anatomy
  – Outflow tract obstruction
  – Exercise tolerance, feeding
  – Medical therapy
Preoperative Assessment

• Physical
  – General appearance
  – BP
  – Cyanosis, clubbing
  – Tachypnea, retractions
  – Peripheral pulses, perfusion
  – Precordium, heart sounds, murmurs
  – Hepatomegaly, JVD
  – Diaphoresis
  – Adequacy of veins, arterial pulses
Preoperative Assessment

- Chest X-ray
  - Heart size and configuration
  - Pulmonary vasculature
  - Pulmonary parenchymal disease
Preoperative Assessment

- EKG
  - Rhythm
  - Rate
  - ST segments
  - Axis deviation
Preoperative Assessment

• Hemoglobin
  – Normal, low, high for age and gender
Preoperative Assessment

- Oxygen saturation
  - What is normal for this patient
  - Any recent changes
  - On home O2
Preoperative Assessment

• **Echo**
  – Cardiac anatomy, residual defects, ventricular function
  – Outflow tract obstruction
  – Valvar regurgitation
  – Atrial/ventricular communication
Preoperative Assessment

- CT
  - Anatomy of extra cardiac structures
    - Aorta, pulmonary, arteries, veins
Preoperative Assessment

- **Cardiac MRI**
  - Anatomy of intra and extra cardiac structures
  - Ventricular function
  - Qp/Qs
Preoperative Assessment

- **Cardiac Cath**
  - Detailed anatomy
  - Hemodynamics
  - SVR/PVR, Qp/Qs
  - PVR reactivity
BOX 3-22  □ HYPOPLASTIC LEFT HEART SYNDROME (HLHS)

HLHS results in aortic atresia or severe stenosis; systemic, brain, and coronary blood flow depends on PDA. Parallel circulation results in HLHS, with Qp/Qs highly variable in the unrepaird state. Excessive Fio₂ and hyperventilation with lower PVR increase Qp/Qs and result in systemic and coronary steal and cardiovascular collapse. Neonatal palliation consists of aortic reconstruction with systemic-PA shunt, RV-PA shunt, or hybrid procedure. HLHS patients typically present for noncardiac surgery and continue to have unstable physiology after stage I palliation.
Norwood Anesthesia

- Preserve balance SVR and PVR
- Minimize myocardial depression
- Extreme caution hypotension
- HCT >40-45
- Narcotic, inhalational or combination

• NO List
  - Sat greater than 75-85% (not good)
  - NO hyperventilate
  - NO 100% oxygen
## Factors Affecting Hemodynamics of the HLHS Patient

<table>
<thead>
<tr>
<th>Manipulations to Increase PVR</th>
<th>Manipulations to Decrease PVR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Decrease inspired concentration O₂</td>
<td>Increased inspired concentration O₂</td>
</tr>
<tr>
<td>Hypoventilation (PCO₂ 40-50)</td>
<td>Hyperventilation (PCO₂ 20-25)</td>
</tr>
<tr>
<td>PEEP</td>
<td>Inotropic support</td>
</tr>
</tbody>
</table>

Stokes, 2005
Bidirectional Glenn or Hemifontan Physiology

Reconstructed outflow from right ventricle to aortic pathways
Superior cardiopulmonary shunt

Anesthesia and Uncommon Diseases, pg 123
Hemi-Fontan Physiology

- Caution to NOT hyperventilate
  - Decreases cerebral flow
  - Decreases pulmonary flow
- NOT use 100% oxygen
- Narcotic, inhalational or combination
Hemi-Fontan and Anesthesia

- Baseline Sats
- Exercise tolerance (SOB, dusky with feeds?)
- Echo, EKG, labs, cath, chest x-ray
- Other congenital issues
- SVC syndrome
- IV access
BOX 3-23  ■ PHYSIOLOGY AFTER BIDIRECTIONAL CAVOPULMONARY ANASTOMOSIS (BCPA)

The SVC is connected to the right PA, resulting in a cerebral-pulmonary-cardiac circulation. Hyperventilation will decrease cerebral blood flow and SVC and PA flow and will lead to arterial desaturation. BCPA circulation is relatively stable, and elective noncardiac surgery is often performed between the BCPA and Fontan stages in single-ventricle patients.
Fontan Physiology
The Fontan operation completes the total cavopulmonary anastomosis. A single systemic ventricle, with no pulmonary ventricle, is the result. Blood flow to the lungs depends on negative intrathoracic pressure during spontaneous respiration. Positive-pressure ventilation significantly compromises systemic venous return to the lungs and should be minimized in Fontan patients. Fontan patients are intolerant of low preload, and CVP monitoring to achieve high-normal preload is important during major noncardiac surgery.
Fontan Physiology

- Adequate preload
- Low pulmonary vascular resistance to preserve pulmonary flow
- Maintaining sinus rhythm
- Minimizing myocardial depression
- Spontaneous ventilation
Fontan and Anesthesia

- Baseline Saturation
- Exercise tolerance
- Echo, EKG, labs, cath, chest x-ray
- Other congenital issues
- Premed
- Narcotic, inhalational or combination
Fontan Long-term Issues

- Cardiovascular
  - Arrhythmias
  - Heart failure
  - Thromboembolism

- Pulmonary
  - Plastic Bronchitis
Fontan Long-term issues

- Protein Losing Enteropathy
  - Hypoalbuminemia
  - Intestinal protein loss
  - Edema
  - Ascites
  - Immune deficiency
Fontan

- Optimal Fontan physiology
  - CVP of 10-15mmHg
  - Pulmonary artery pressure of 10-15mmHg
  - Left atrial (LA) pressure of 5-10 mmHg

- Transpulmonary gradient
Postoperative

- Pain control
- Need for overnight stay
- Need for special monitoring
- Need for ICU bed
# Problems in Adults with Congenital Heart Disease

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<th>Psychosocial</th>
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<td>Shunts</td>
<td>Employment</td>
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<td>Ventricular dysfunction</td>
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<td>Valve stenosis or regurgitation</td>
<td>Flying or driving recommendations</td>
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<td>Pulmonary hypertension</td>
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<td>Management of pregnancy</td>
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<td>Endocarditis prophylaxis</td>
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</table>
Summary

• Understand the patient’s anatomy/physiology including shunts (Draw it out!)

• Understand how anesthetics and vasoactive substances affect the patient’s cardiovascular physiology

• Understand the effects of ventilation and oxygenation on the patient’s cardiovascular physiology
Happy Belated Valentine’s Day !!

- [http://youtu.be/SyBNLon4EjA](http://youtu.be/SyBNLon4EjA)
References

• Anesthesia and Uncommon Diseases
• Anesthesia for Congenital Heart Disease
• CDC, AHA, ACHA, March of Dimes websites