Anesthesia for the adult congenital heart disease patient for non-cardiac surgery

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Disclosures

• None
Case examples

• How would you plan these anesthetics?
Laparoscopic Cholecystectomy

- 28 year old female
- 3 day history of fever and RUQ pain
- Nausea
- Radiographic confirmation of acute cholecystitis
- Patient has stage III palliation (Fontan) for HLHS
Laparoscopic Appendicetomy

- 19 year old male
- 1 day history of nausea, pyrexia, and RLQ pain
- CT confirmation of acute appendicitis
- Patient has history of mBT shunt followed by transannular patch for Tetralogy of Fallot
Pregnancy: Delivery planning

- 25 year old female
- Term pregnancy
- Large unrepaired ASD
- Severe pulmonary hypertension & RV dilatation
Topics to be Discussed

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

- ~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
- 1/13 infant deaths due to CHD
CHD Fast Facts

- CHD is common
- 30% of congenital disease is cardiac
- Baby's risk of CHD increased 3x if parents/siblings have CHD
- 2x as many children die of CHD as all cancer combined
- 2009; 40,000 hospital stays for CHD
- Health care cost nearly $2B/yr in the US
ACHD/GUCH Fast Facts

• CHD mortality decreased 40% in past 20 years
• 85-95% survive to adulthood
• 50% would have died without intervention
• 10% of all CHD are first diagnosed in adulthood
Incidence

- VSD 30%
- PDA 9%
- ASD 7%
- PS 7%
- Other 25%

- CoA 6%
- TGA 5%
- TOF 5%
- AS 5%
- HLHS <1%

• US CHD population estimate: 2,425,000
  – 1,444,500 adults with CHD
  – 160,000 adults with severe disease
• 40% have gaps in medical care >3 years
• Not enough ACHD centers for care
Challenges for Pediatric Anesthesiologists

• They are not just big CHD patients
• ACHD patient develop acquired disease that pediatric providers are not familiar with:
  – Drug/Alcohol Use/Abuse
  – Pregnancy
  – Cardiac Comorbidities – Coronary Disease
• Tough patients with complex & multiple Medical Comorbidities
Challenges for Adult Anesthesiologists

• Transitional care
  – Significant psychosocial overlay with patient and families
  – Dealing with pediatric cardiologists and practitioners
  – Gaps in medical care
• Complex Anatomy
• Obscure Surgical Repairs
Long term sequelae

- **Cardiac**
  - Arrhythmias and conduction defects
  - PHTN
  - Ventricular dysfunction
  - Aneurysms
  - Valvular pathology
  - Endocarditis
  - Residual shunts
  - Hypertension
  - Atherosclerosis (esp. TXP)

- **Extra-cardiac**
  - Neuro & Psych
    - Psychological, CVA, Seizures
  - Other syndromes
  - Secondary erythrocytosis
    - Thrombotic risk
  - Cholelithiasis
  - Lung disease
  - Hepatic dysfunction & cirrhosis
  - Renal impairment
  - Endocrine
  - Cancers
Changing Mortality

Histogram bars depict the proportion of all deaths (x-axis) according to age at death (y-axis) in our cohort of patients with congenital heart disease in the first (1987 to 1988; left) and final (2004 to 2005; right) years of observation. *Bold black curves with diamonds* represent the corresponding age at death distribution in the general Quebec population during the same periods of observation.
ACHD Classification

AHA/ACC GUIDELINE

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease
A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

- Recommend using physiological variable in conjunction with Anatomic features for classification - ACHD-AP
ACHD-AP

- Physiologic
  - Aortopathy
  - Arrhythmia
  - Concomitant VHD
  - End-organ dysfunction
  - Exercise capacity
  - Hypoxia/cyanosis
  - NYHA functional class
  - PHTN
  - Shunt
  - Venous and arterial stenosis

- Anatomic
  - Simple
  - Moderate
  - Complex
    - Cyanotic CHD
    - Double outlet ventricle
    - Fontan
    - Interrupted aortic arch
    - MS
    - Single ventricle
    - Pulmonary atresia
    - TGA
    - Truncus Arteriosus
    - Other AV or VA connection anomalies eg. heterotaxy
Shunt Physiology in CHD

• Too much pulmonary blood flow
  – congestive heart failure
  – pulmonary vascular disease

• Too little pulmonary blood flow leads to cyanosis
Qp/Qs

- Normal = 1
- >1 = L to R shunt “Pink”
- <1 = R to L shunt “Blue”
Blood flow through Shunts

- Poiseuille's Law
  - Resistance $\alpha$ length/radius^4
- Size of orifice (or length of shunt)
- Pressure in the chambers on either side of shunt
- Resistances downstream (PVR and SVR)
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
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
Right to Left Shunts

- Chronic hypoxia leads to polycythemia & hyperviscosity
  - Worsened by dehydration
- Ramifications for all organ systems
- Care with opioids
- IV induction is faster (vein to brain)
- PVR/SVR balance
Tetralogy of Fallot

Normal heart

Tetralogy of Fallot

- Overriding aorta
- Pulmonic stenosis
- Ventricular septal defect
- Right ventricular hypertrophy
Adults with TOF

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

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

- Pulmonary insufficiency or stenosis
- RV dilatation & dysfunction, functional TR
- Prolonged QT
- Sustained V-tach
- Atrial arrhythmias
- Heart failure
- Sudden death ~ 8% at age > 35

Poor Prognosis in TOF

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

Anesthetic considerations

• Arrythmias
  – Treat & manage metabolic disturbances, Pacemakers, AI CD
• PI
  – maintain low PVR, high-normal HR, manage RV failure
• PS
  – Maintain systemic afterload to preserve coronary perfusion
• Unrepaired TOF
  – Maintain SVR, low PVR, avoid hypovolemia & sympathetic surges, be prepared to treat ‘Spells’
• Palliated TOF
  – Balance Qp:Qs
Should I feel comfortable?

• Yes
  – Repaired TOF with good function
    • Treat symptomatically ie; RV failure, manage PI/PS

• No
  – Palliated or unrepaired
  – Reduced function
The Systemic Right Ventricle

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

- Atrioventricular discordance
- Ventriculoarterial discordance
d-TGA; Complete Transposition of the Great Arteries

- 1. Aorta arising from the right ventricle.
- 2. Pulmonary artery arising form the left ventricle
- Ventriculoarterial discordance
Atrial Switch Procedures

Intra-atrial baffle (mustard or Senning procedure)

Aorta
Pulmonary Artery
Baffle
Sequelae of a systemic RV

- RV dysfunction & failure
- Tricuspid insufficiency
- Arrhythmias & AV block
- 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
Long term sequelae of Switches

- Baffle Leak or stenosis
- Atrial arrhythmias & complete heart block
- Tricuspid regurgitation
- RV failure
- Coronary stenosis
- PA or branch PA stenosis
- Neoaortic regurgitation

Anesthetic Management Outcomes

- 50 patients
  - 45 arterial switches
  - 5 atrial switches
- 4 adverse events (8%)
  - significant bradycardia
  - 2 failed extubations
  - postoperative bleeding/hematoma

Should I feel comfortable?

• Yes
  – If no failure, no problem.
  – RV failure; treat like systemic ventricular failure
  – May have conduction abnormalities

• have external defib/pacer pads ON
The Single Ventricle

- Single RV
  - HLHS
- Single LV
  - Tricuspid atresia, HRH
- Indeterminate Ventricle
  - Unbalanced AV canal
Tricuspid Atresia

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

Opening Between Atria
Closed Tricuspid Valve
Underdeveloped Right Ventricle

Oxygen-rich Blood
Oxygen-poor Blood
Mixed Blood
HLHS
Hemifontan
Fontan
Sequelae of a Single Ventricle

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

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
- 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.
Anesthetic considerations

• Premedication
• Avoid prolonged fasting times
• Euvolemia
• All pulmonary blood flow is passive
  – Maintain transpulmonary gradient
  – Low PVR
• Careful with laparoscopic procedures
• Bleeding/coagulopathy
Ventilation Strategy

- Spontaneous ventilation
  - Limited by hypoventilation and atelectasis
- Mechanical ventilation
- Avoid hypoxia
- Avoid hypercarbia
Should I feel comfortable?

• Probably not
  – These patients should be anesthetized by a Congenital Cardiac Anesthesiologists at a specialty center

• Probably OK
  – High functioning Fontan’s for minor procedures
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

• IV induction slower
• Inhalation induction same or slightly slower
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

• High risk cases (CHD + PHTN)
Eisenmenger’s Syndrome

- Reversal of the shunt to right to left
- Same sequelae as unrepaired TOF
- End stage irreversible pulmonary disease
- 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

Anesthetic principles

- Continue home pulmonary vasodilator therapy
- Minimize PVR & avoid increases
- Maintain SVR
- Pulmonary vasodilatation
  - iNO
  - Milrinone
- Regional
- Avoid sympathetic surges
Should I feel comfortable?

• NO
  – These patients should **ALL** be anesthetized by a Congenital Cardiac Anesthesiologists at a specialty center
Pregnancy and CHD

• More CHD patients are surviving to childbearing years
• CHD is 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

- 2nd and 3rd 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 or LVOT obstruction
- Marfans 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 25 days out
- Perinatal mortality is 28%
- Maternal mortality for pregnancy termination is 7%

Indications for C-Section in patients with CHD

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

Siu SC, Colman JM, Heart Disease and Pregnancy, Heart 2001, 85:710-715
Anesthetic considerations

- CHD specific
- Neuraxial techniques
  - Epidural
  - Spinal
    - Not advisable, sudden loss of afterload & preload
- Vaginal delivery with instrumental assist to minimize pushing
- GA
Should I feel comfortable?

- No
  - Complex congenital heart disease
    - Consult congenital cardiac anesthesiology

- Yes
  - Adult Disease (Aortic disease, CHF, PHTN)
    - May consider consulting adult cardiac anesthesia & cardiac surgery
Summary; who should manage

- **Anyone (with good function)**
  - Repaired TOF
  - Systemic RV
  - Double switch

- **A Pediatric Cardiac Anesthesiologist**
  - Fontan Physiology
  - Eisenmengers
  - Complex Cyanotic Heart Defects