CRASH Vail, Colorado 2018
Colorado Review of Anesthesia for SurgiCenters and Hospitals
and Ski Holiday!

Anesthetic considerations in Adults with Congenital Heart Disease

Dr. Mark Twite MA MB BChir FRCP
Director of Congenital Cardiac Anesthesiology
Associate Professor
Department of Anesthesiology
University of Colorado, Anschutz Medical Campus & Children’s Hospital Colorado

CRASH Vail, Colorado 2018
Colorado Review of Anesthesia for SurgiCenters and Hospitals

No Financial Disclosures

Objectives
Understand and Discuss the:
1. Changing epidemiology of congenital heart disease (CHD)
2. Current outcomes for patients with CHD
3. Perioperative anesthetic approach to adults with CHD undergoing non-cardiac surgery

Case
• 37yr old female in Atrial Flutter presents for TEE, EP study and ablation

• Tricuspid Atresia s/p Fontan
  - Non-alcoholic cirrhosis (elevated LFT & low albumin)
  - Protein losing enteropathy
  - Hypoalbuminemia 57.5-59% in air. Polycythemia Hct 65
  - Pacemaker Dual Chamber DDD at 70bpm

• Medications
  - Apixaban, Aspirin
  - L-Arginine
  - Digoxin, Quinapril, Sotalol
  - Furosemide, Spironolactone
  - Melatonin, Valium, Ambien, Tramadol

Case
• 37yr old female in Atrial Flutter presents for TEE, EP study and ablation

• Echocardiogram 9/2016 TTE
  1. History of Tricuspid atresia, status post initial class (RA to PA) Fontan, with subsequent Fontan conversion to extra-annular lateral tunnel Fontan through the RA body
  2. Subjectively there remains normal LV systolic function.
  3. Dilated IVC without respiratory collapse. Low velocity flow within IVC suggests IVC pressure is greater than 15 mm Hg.
  4. The Glenn anastomosis is not well visualized, though there is normal respiratory variation in SVC flow suggestive of no obstruction within the Fontan pathway.
  5. Trivial aortic regurgitation, mild mitral regurgitation (mean gradient 5.6 mmHg).
  6. Hypoalbuminemic liver disease with subjectively normal function. Hypoplastic right ventricle is not well seen.
  7. No pericardial effusion.
Case

• 37-year-old female in Atrial Flutter presents for TEE, EP study and ablation

• Cardiac Cath
  CATH (Denver, 2010) no fenestration, unobstructed PA's, Fontan 11-12 mmHg, no change with volume challenge; small angiographic right to left shunt seen likely from the stent time of the lateral tunnel.
  
  CATH (Denver, 2013) Fontan pressures 18 mmHg, PCWP 14 mmHg, small veno-venous collaterals off left innominate embolized, 100% FIO2 and improved Fontan pressure to 11 mmHg
  
  CATH (Denver, 2016) hepatic wedge 10 mmHg, hepatic vein 15 mmHg (transhepatic gradient 4 mmHg), Fontan 13, RPA 11, LPA 12, PCWP 9. Fick CO 5.06. S/P 10ml/kg bolus PA increased to 17, PCWP to 14. Fick CO 5.74. With 100% O2, PA pressure decreased to 13, PCWP to 12. Fick CO 5.69, with 40 ppm I NO PA pressure 14 , PCWP 13, CO 5.64

Congenital Heart Disease

• Most common congenital disorder of newborns
  - 1% of live births
  - Leading cause of infant deaths in the USA
  - Accounts for more than half of all deaths from congenital anomalies worldwide

• Estimated 1.5 million adults living in USA with CHD

• NIH funded CHD research from 2005 – 2015
  - 663 CHD research projects for a total cost of $991 million
  - 70% Basic science (Cardiac developmental biology most common)
  - 27% Clinical
  - 3% Both

Changing Epidemiology: Adults and Children in Quebec

<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>20</td>
</tr>
</tbody>
</table>

Changing Epidemiology: Mortality

Classification of CHD Complexity

<table>
<thead>
<tr>
<th>Simple</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect</td>
<td>Anomalous pulmonary venous drainage</td>
<td>Single ventricle palliation</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>Anterior ventricular canal defect</td>
<td>Transposition of the great arteries</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>Coarctation of the aorta</td>
<td>Truncus arteriosus</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Tricuspid atresia</td>
<td>Pulmonary atresia</td>
</tr>
<tr>
<td>Eisenmenger syndrome</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Simple defects have a favorable natural history unless they are unrepaired with a significant L to R shunt which may then develop Eisenmenger syndrome.
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>Moderate</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>Anomalouos pulmonary venous drainage</td>
<td>Single ventricle palliation</td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>Aortoventricular canal defect</td>
<td>Transposition of the great arteries</td>
<td></td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>Coarctation of the aorta</td>
<td>Truncus arteriosus</td>
<td>Pulmonary atresia</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Tricuspid atresia</td>
<td>Pulmonary atresia</td>
<td>Eisenmenger syndrome</td>
</tr>
</tbody>
</table>

Long term survival > 20 years

CHD Pathophysiology

- Atrial stretch, connections, synchrony, residual lesions
- Ventricular function, rhythm, geometry, residual lesions
- Pulmonary hypertension
- Cyanosis & Bleeding
- Valve stenosis & regurgitation
- Heart Failure
- Arrhythmias
- Outflow stenosis, dilatation, compression
- Endocarditis prophylaxis

COMORBIDITIES

- Congenital & Acquired
- Tetralogy of Fallot
- PI, RV Dilation

Lesion Specific

1. Single Ventricle
   - Left vs Right, PLE
   - Thromboembolic
   - Liver Dysfunction
2. Tetralogy of Fallot
3. TGA
   - Baffle stenosis, coronary

Special Situations

- Pregnancy, laparoscopy, regional & neuraxial

Endocarditis prophylaxis

- Valve stenosis, dilatation, compression

Psycho social

Changing Surgical techniques: TGA

1960 Senning-Mustard

1975 Jatene Arterial Switch Operation

Changing Surgical techniques: TA Stage 1 BT Shunt

Changing Surgical techniques: TA Stage 2 Glenn Shunt
Changing Surgical techniques: TA Stage 3 Fontan

- AtrioPulmonary
- Lateral Tunnel
- Extracardiac

Effects of anesthetic agents

<table>
<thead>
<tr>
<th>Agent</th>
<th>Effect on Contractility</th>
<th>Effect on MAP</th>
<th>Effect on SVR</th>
<th>Effect on PAP</th>
<th>Effect on VR</th>
<th>Effect on HR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Norepinephrine</td>
<td>↑↑</td>
<td>↑↑</td>
<td>↑↑</td>
<td>↑</td>
<td>↑↑</td>
<td>↑↑</td>
</tr>
<tr>
<td>Epinephrine</td>
<td>↑</td>
<td>↑</td>
<td>↑</td>
<td>↑↑</td>
<td>↑↑</td>
<td>↑↑</td>
</tr>
<tr>
<td>Dopamine</td>
<td>↑↑</td>
<td>↑↑</td>
<td>↑↑</td>
<td>↑↑</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Procainamide</td>
<td>↑</td>
<td>↑</td>
<td>↑</td>
<td>↑</td>
<td>↑↑</td>
<td>↑↑</td>
</tr>
<tr>
<td>Ketamine</td>
<td>↑↑</td>
<td>↑</td>
<td>↑</td>
<td>↑↑</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Propofol</td>
<td>↑↑</td>
<td>↑</td>
<td>↑</td>
<td>↑↑</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Remifentanil</td>
<td>↑</td>
<td>↑</td>
<td>↑</td>
<td>↑↑</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Dexmedetomidine</td>
<td>↑↑</td>
<td>↑</td>
<td>↑</td>
<td>↑↑</td>
<td>↑</td>
<td>↑</td>
</tr>
</tbody>
</table>

Consensus Statements

**AHA SCIENTIFIC STATEMENT** Circulation, October 2017

Diagnosis and Management of Noncardiac Complications in Adults With Congenital Heart Disease

**AHA SCIENTIFIC STATEMENT** Circulation, January 2017

Management of Pregnancy in Patients With Complex Congenital Heart Disease

A Scientific Statement for Healthcare Professionals From the American Heart Association

Atrial Arrhythmias

Studied 482 adults with CHD from 12 centers in the USA

- Most common presenting arrhythmia:
  - Intra-atrial re-entrant tachycardia (IART) 61.6% (increased with CHD complexity)
  - Atrial fibrillation (AF) 28.8%, increased with age to surpass IART at 50yrs of age
  - Focal atrial tachycardia 9.5%

Time-series analysis: referral to specialized ACHD centers and ACHD patient mortality

Atrial Arrhythmias

Risk factors for developing atrial arrhythmias:

- Single ventricle
- Previous intracardiac repair
- Systemic right ventricle
- Pulmonary hypertension
Adults with CHD and atrial arrhythmias:
• x 4 increase of heart failure
• x 2 increase in death

Atrial arrhythmias frequent cause of sudden cardiac death (43%)
Increasing incidence with complexity of CHD
- Eisenmenger
  - Transposition of great arteries (atrial switch)
  - Single ventricles lesions palliated to Fontan circulation

Single Center study in USA of Fontan patients
• 10, 20 and 30 year freedom from arrhythmias of 71%, 42% and 24%
• SCD 52 of 1052 Fontan patients (5%) with 65% due to arrhythmias

Risk estimates for arrhythmias across CHD

Majority of SCD victims unrecognized with an area under the curve of 0.6 for the discriminative ability of current guidelines
Critical clinical reasoning essential when deciding on ICD placement in adult CHD patients
**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

**Heart Liver Transplant**

UNOS Database 1987 – 2015
- 61437 Heart Tx
- 190 CHLT
- 41 had CHD
- 30 day, 1 and 5 year survival 95%, 86% and 83%
- CHLT with and without CHD comparable
- Trend towards better survival for CHLT compared with isolated Heart Tx for CHD

**CHD Pathophysiology**

- **Atrial stenosis**, connections, synchrony, residual lesions
- **Heart Failure**
- **Arrhythmias**
- **Outflow stenosis**, dilatation, compression
- **COMORBITIES**
  - Congenital & Acquired
  - Pulmonary Hypertension
  - Cyanosis & Bleeding
  - Ventricular function, rhythm, geometry, residual lesions

**Endocarditis prophylaxis**

- **Lesion Specific**
  - 1. Single Ventricle
  - Left vs Right, PLE
  - Thromboembolic
- **Liver Dysfunction**
  - **TGA**
  - Baffle stenosis, cyanosis

**Special Situations**

- Pregnancy, laparoscopy, regional & neuraxial