High Risk Cardiac Lesions for Non-Cardiac Surgery

- Williams, Pulm HTN, Sinusoids - Oh my!

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Nothing to disclose, no conflict of interest
OR schedule for Monday

8:30:
• 9 yo girl with Williams Syndrome for bilateral strabismus surgery

11:00:
• 12 yo with pulm HTN for Broviac insertion

14:00:
• 1 mo with PA/IVS s/p repair for lap G-tube
Objectives

• Describe the pathophysiology and anesthetic concerns for patients with Williams syndrome

• Discuss the preoperative evaluation and anesthetic management of patients with pulmonary HTN

• Understand the treatment strategies for patients with Sinusoids and the anesthetic implications for non-cardiac surgery
Back to our OR schedule for Monday ...

First Case:
• 9 yo girl with Williams Syndrome for bilateral strabismus surgery

“Elfin” facies

“Cocktail Party” Personality
Sudden cardiac death under anesthesia in pediatric patient with Williams syndrome: a case report and review of literature.

Gupta P1, Tobias JD, Goyal S, Miller MD, Melendez E, Noviski N, De Moor MM, Mehta V.

Anaesthesia-related haemodynamic complications in Williams syndrome patients: a review of one institution's experience.

Olsen M1, Fahy CJ, Costi DA, Kelly AJ, Burgoyne LL.
Williams-Beuren Syndrome

• First described in 1961
• Prevalence: ~1:10 000 – 1:20 000
• Microdeletion syndrome:
  Spontaneous deletion of 26-28 genes in a specific segment on chromosome 7
  Including the elastin gene (ELN)

  ⇒ Cardiovascular changes
  ⇒ Connective tissue abnormalities
  ⇒ Endocrine problems
  ⇒ Cognitive and behavioral issues
  ⇒ “premature aging”

<table>
<thead>
<tr>
<th>Organ System</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Auditory, ear, nose throat</td>
<td>Hyperacusis, recurrent otitis media, hearing loss later in life</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>Elastin arteriopathy, vascular stenosis: SVAS, PPS, CAD, hypertension, stroke</td>
</tr>
<tr>
<td>Development/Cognition</td>
<td>Global impairment, characteristic pattern: strong language skills, poor visuospatial</td>
</tr>
<tr>
<td>Dental</td>
<td>Small or unusual shaped teeth, malocclusion</td>
</tr>
<tr>
<td>Endocrine</td>
<td>Hypercalcemia, glucose intolerance, early onset of puberty, osteopenia, hypothyroidism</td>
</tr>
<tr>
<td>GI</td>
<td>Feeding intolerance, poor weight gain, GERD, constipation, diverticulitis</td>
</tr>
<tr>
<td>GU</td>
<td>Renal anomalies, bladder diverticula, nephrocalcinosis, delayed toilet training, UTIs</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>Short stature, scoliosis, joint contractures or laxity</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Hypotonia, hyperreflexia, poor balance and coordination, Type I Chiari malformation</td>
</tr>
<tr>
<td>Ophthalmologic</td>
<td>Strabismus, poor vision, narrow lacrimal duct</td>
</tr>
<tr>
<td>Personality</td>
<td>Friendly, “cocktail party”, ADHD, anxiety and phobias</td>
</tr>
<tr>
<td>Skin and integument</td>
<td>Soft skin, premature aging, premature graying of hair</td>
</tr>
</tbody>
</table>
Anesthesia Concerns

Generalized elastin arteriopathy

• Reduced amount of elastin in media of great vessels
• Hypertrophy of smooth muscle cells and collagen
  ⇒ “Stiff” arteries: Loss of windkessel effect, wide pulse pressure, hypertension
  ⇒ Impaired coronary perfusion (ostial occlusions or coronary artery stenosis)
  ⇒ 70% Supravalvar aortic stenosis (sinutubular junction, hourglass)
  ⇒ Peripheral pulmonary stenosis, occ. Central PS
  ⇒ Involvement of transverse, descending aorta, renal and mesenteric arteries

⇒ Bilateral outflow obstruction, biventricular hypertrophy, CAD, HTN
Anesthesia Concerns

Generalized elastin arteriopathy

- Cardiovascular complications are the major cause of death
- CV associated mortality 25-100 x higher
- Multiple case reports of sudden death under anesthesia

- “Stiff” arteries: Loss of windkessel effect, wide pulse pressure, hypertension
- Impaired coronary perfusion (ostial occlusions or coronary artery stenosis)
- 70% Supravalvar aortic stenosis (sinutubular junction, hourglass)
- Peripheral pulmonary stenosis, occ. Central PS
- Involvement of transverse, descending aorta, renal and mesenteric arteries

=> Bilateral outflow obstruction, biventricular hypertrophy, CAD, HTN
Burch TM et al:
A & A 2008
107(6):1848-54.
Preoperative Evaluation

• Careful risk/benefit evaluation of planned procedure

• History and Physical exam
  • CV symptoms, medications, h/o syncope, chest pain, arrhythmia etc. ?
  • Previous Anesthesia ?
  • Cooperation ? Behavioral issues ? ADHD/anxiety medications ?
  • Dental status, airway exam ?
  • Endocrine status ? Review of recent labs: renal, thyroid, electrolytes

• Review of diagnostic studies
  • ECG: evidence of ventricular hypertrophy, ST changes, arrhythmia
  • Echo: supravalvar stenosis, gradients, ventricular hypertrophy and function
  • MRI: function, gradients, status of coronaries
  • Cath: “gold standard”, location of obstruction, gradients, coronary angio, renal involvement

• Discussion with Cardiologist
Preoperative Evaluation

• Careful risk/benefit evaluation of planned procedure
• History and Physical exam

Unfortunately, no relationship between the degree of supravalvar obstruction and coronary involvement

=> Severe coronary stenosis is possible even in the absence of a significant gradient

• ECG: evidence of ventricular hypertrophy, ST changes, arrhythmia
• Echo: supravalvar stenosis, gradients, ventricular hypertrophy and function
• MRI: function, gradients, status of coronaries
• Cath: “gold standard”, location of obstruction, gradients, coronary angio, renal involvement

• Discussion with Cardiologist
Anesthetic Management

Optimal myocardial oxygen demand and supply ratio
Increased demand: hypertrophied myocardium with diastolic dysfunction
Decreased supply: impaired coronary blood and perfusion pressure

<table>
<thead>
<tr>
<th>Goals</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maintain age appropriate heart rate</td>
<td>Careful with vagolytic drugs (atropine, glycopyrrolate) or drugs with sympathomimetic activity (ketamine, pancuronium) Avoid excessive tachycardia during reversal with neostigmine &amp; atropine Low dose epinephrine(0.1-1 μg/kg) or ephedrine preferred for bradycardia</td>
</tr>
<tr>
<td>Maintain Sinus Rhythm</td>
<td>Aggressive treatment of SVT (cardioversion often preferable)</td>
</tr>
<tr>
<td>Maintain Preload</td>
<td>Short NPO times, careful titration of vasodilating anesthetic drugs, rapid fluid administration can cause pulmonary edema (diastolic dysfunction)</td>
</tr>
<tr>
<td>Maintain Contractility</td>
<td>Careful titration of all negative inotropic agents</td>
</tr>
<tr>
<td>Maintain SVR</td>
<td>Treatment of hypotension with pure agonists (phenylephrine)</td>
</tr>
<tr>
<td>Avoid Increases in PVR</td>
<td>Avoid hypoxia and hypercarbia, optimize ventilation strategy: low PIPs</td>
</tr>
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</table>
Preparation

• Choose the right venue!
  Not a dental office or remote satellite facility, expert help available

• Communicate with your team! High risk for CPR

• Equipment and medications ready
  • 5 lead ECG and ST monitoring, print “baseline strip” before induction
  • Defibrillator for potential cardioversion: at least close by and checked.
  • Phenylephrine, esmolol and epinephrine in appropriate concentrations

• Appropriate Recovery
  • Prolonged observation and monitoring, adequate PONV and pain treatment, usually not a fast track day surgery case
Anesthesia Plan

• Communicate with your team! High risk for CPR
• Good premedication!
• Induction: intravenous versus inhalation?
• Airway management: LMA vs ETT, spontaneous vs controlled
• Monitoring: Need for A-line or CVL?
• Mental exercise: Discussion of potential problems and treatment
  • Hypotension: phenylephrine
  • Tachycardia: deepen anesthesia, careful titration of esmolol
  • “Oculocardiac Reflex” - Bradycardia: stop stimulation, low dose epinephrine
Currently no evidence that Williams syndrome is associated with congenital prolonged QT syndromes and channelopathies!

Retrospective ECG review: 270 WS patient – 499 ECGs
QTc prolongation: WS 13.6% vs healthy children 2%

Retrospective ambulatory ECG review: 1980-2007
56 ECGs in 26 WS patients:
PVCs in 73% of ECGs, 81% of patients
VT in 9% of ECGs and 15% of patients, mean 3.6 beats
QTc correlated with age and total number of PVCs

Abnormalities of Cardiac Repolarization in Williams Syndrome

R. Thomas Collins II, MD\textsuperscript{a,b,*}, Peter F. Aziz, MD\textsuperscript{a,c}, Marie M. Gleason, MD\textsuperscript{a,c}, Paige B. Kaplan, MBCh\textsuperscript{b,c}, and Mauliy J. Shah, MBBS\textsuperscript{a,e}


Relation of Ventricular Ectopic Complexes to QTc Interval on Ambulatory Electrocardiograms in Williams Syndrome

R. Thomas Collins II, MD\textsuperscript{a,b,*}, Peter F. Aziz, MD\textsuperscript{a}, Christopher J. Swearingen, PhD\textsuperscript{a,b}, and Paige B. Kaplan, MBCh\textsuperscript{d,e}

Acquired Long QT Syndrome

- **Variety of Drugs**
- **Electrolyte Disturbances**
  - Hypokalemia
  - Hypocalcemia
  - Hypomagnesemia
- **Medical Conditions:**
  - **Bradycardia:** AV block, sick sinus etc
  - **Myocardial dysfunction:** CHF, myocarditis, cardiomyopathies
  - **Endocrinopathy:** hypothyroidism, hyperparathyroidism
  - **Neurologic:** encephalitis, head trauma, stroke, tumor
  - **Nutritional:** alcoholism, anorexia, starvation

[www.torsades.org](http://www.torsades.org)
[www.QTdrugs.org](http://www.QTdrugs.org)
Currently no evidence that Williams syndrome is associated with congenital prolonged QT syndromes and channelopathies!

No case reports of torsades!
Currently no evidence that Williams syndrome is associated with congenital prolonged QT syndromes and channelopathies.

No case reports of torsades.

Williams Syndrome

- Bilateral outflow obstructions
- Severe biventricular hypertrophy
- Unrecognized coronary stenosis
- Supra-systemic RV pressures
- Risk for ischemia and arrhythmia
Back to our OR schedule ...

Second Case:
- 12 yo girl with Pulmonary Hypertension for broviac insertion

“Another disaster waiting to happen”
Incidence of anesthesia related death: 0.98 per 10 000 anesthetics
Pulmonary Hypertension involved in 50%
(5 out of 10 deaths)

# 1: 15yo autistic girl with severe pulm. HTN, on max. medical therapy, for surgical iv line
Sedation with midazolam and remifentanil
=> Acute pulmonary hypertensive crisis in recovery => ICU => Floor => arrested at night

# 2: 4yo with restrictive CM and pulm. HTN for cardiac cath: ST changes during cath
=> Further ST changes at end of cath => V-fib => unable to resuscitate

# 3: 8yo girl with primary pulm. HTN for surgical IV line:
=> GETA with Fentanyl, Propofol and Isofl.: 60 min later bradycardia & arrest => CPR no success

#4: 1yo with new diagnosis of primary pulm. HTN for cardiac cath
=> Cardiac arrest after induction of anesthesia => unable to resuscitate

#5: 5 mo, Ex 29wk premie with Trisomy 21, CAVC, CLD and pulm. HTN for cardiac surgery
=> Loss of cardiac output during A-line insertion => CPR onto bypass => off onto ECMO
Pulmonary Hypertension

- Prevalence in general population: 15-50/million, f:m 1.7:1
  - 0.5 million people with PH in developed world
  - 35 million in developing countries (high altitude, infections like schistosomiasis)

- Estimated prevalence in children: < 10 cases per 1 million

- Pediatric Data from multicenter PAH registry:
  - At time of diagnosis: mPAP 56mmHg and PVR 17 Wood units/m²
  - 5y survival: 74± 6%
  - No significant difference between idiopathic PAH or PAH associated with CHD

### WHO-Classification of Pulmonary Hypertension (latest update 2009)

| Pulmonary Arterial Hypertension | - Idiopathic  
|                                | - Familial  
|                                | - Associated with:  
|                                |   - Collagen vascular disease  
|                                |   - Congenital systemic-to-pulmonary shunts  
|                                |   - Portal hypertension  
|                                |   - HIV infection  
|                                |   - Drugs and toxins (Cocaine, Methamphetamine…)  
|                                |   - Persistent pulmonary hypertension of the newborn  
|                                |   - Pulmonary veno-occlusive disease  
| With Left-Heart disease        | - Left-sided atrial or ventricular disease  
|                                | - Left-sided valvular heart disease  
| With Disorders of Respiratory System | - Chronic obstructive pulmonary disease  
|                                | - Interstitial lung disease  
|                                | - Sleep disordered breathing  
|                                | - Alveolar hypoventilation disorders  
|                                | - Chronic exposure to high altitude  
|                                | - Neonatal lung disease  
| Chronic Thrombotic or Acute Embolic Disease | - Thromboembolic obstruction proximal or distal PAs  
| Miscellaneous                  | e.g. Sarcoidosis, Histiocytosis, compression by tumors  |
Children are not just small adults...

Important pediatric considerations

• Fetal origins of vascular disease
• Perinatal maladaptation
• Early childhood maldevelopment
• Complex Heterogeneity:
  • Prematurity
  • CHD
  • Syndromes, chromosomal anomalies
  • Lung disease secondary to aspiration
  • Sleep disordered breathing
Panama Classification of Pediatric PHVD

Table 1: The broad schema of 10 basic categories of Pediatric Pulmonary Hypertensive Vascular Disease

<table>
<thead>
<tr>
<th>Category</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Prenatal or developmental pulmonary hypertensive vascular disease</td>
</tr>
<tr>
<td>2</td>
<td>Perinatal pulmonary vascular maladaptation</td>
</tr>
<tr>
<td>3</td>
<td>Pediatric cardiovascular disease</td>
</tr>
<tr>
<td>4</td>
<td>Bronchopulmonary dysplasia</td>
</tr>
<tr>
<td>5</td>
<td>Isolated pediatric pulmonary hypertensive vascular disease (isolated pediatric PAH)</td>
</tr>
<tr>
<td>6</td>
<td>Multifactorial pulmonary hypertensive vascular disease in congenital malformation syndromes</td>
</tr>
<tr>
<td>7</td>
<td>Pediatric lung disease</td>
</tr>
<tr>
<td>8</td>
<td>Pediatric thromboembolic disease</td>
</tr>
<tr>
<td>9</td>
<td>Pediatric hypobaric hypoxic exposure</td>
</tr>
<tr>
<td>10</td>
<td>Pediatric pulmonary vascular disease associated with other system disorders</td>
</tr>
</tbody>
</table>

Talking about complex heterogeneity...
New Definition: “Panama” Criteria

• **Biventricular Circulation:**
  \[ \text{mPAP} > 25\text{mmHg} + \text{PVR} > 3 \text{ Wood units/m}^2 \]

• **Univentricular Circulation:** *(s/p cavopulmonary anastomosis)*
  \[ \text{PVR} > 3 \text{ Wood Units/m}^2 \]
  or \[ \text{TPG} > 6\text{mmHg}, \text{ even if mPAP} < 25\text{mmHg} \]
Routine work up for PAH:

Potential Anesthesia involvement:

=> Prior to Diagnosis and treatment!

Mullen MP. Chap 10 in Nadas’s Pediatric Cardiology, 2nd ed, p 120
Pathophysiology and Treatment

- Vasoconstriction
- Smooth muscle cell proliferation
- Endothelial cell proliferation
- Thrombosis

Endothelial cell dysfunction

- Imbalance between vasodilators (prostacyclin, NO) and vasoconstrictors (Thromboxan A₂, Endothelin-1), growth inhibitors and mitogenic factors, anti- and pro-thrombogenic elements
# Medical Management

<table>
<thead>
<tr>
<th>Mechanism of Action</th>
<th>Medication</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calcium Channel Blocker</td>
<td>Nifedipine</td>
<td>If positive response to vasodilator testing, careful dose titration</td>
</tr>
<tr>
<td></td>
<td>Diltiazem and Amlodipine</td>
<td></td>
</tr>
<tr>
<td>Prostacyclin</td>
<td>Epoprostenol (Flolan): iv, short half life</td>
<td>Side effects: headache, diarrhea, jaw and leg pain, rash, nausea, flushing, syncope, catheter and pump cx</td>
</tr>
<tr>
<td>(=) vasodilation, platelet inhibition</td>
<td>Treprostinil (Remodulin): iv or sc, inhaled</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Iloprost (Ventavis): inhaled 6-9x/day</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Beroprost: oral 4-6x/day</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Endothelin Receptor Antagonists</td>
<td>Bosentan: oral BID</td>
<td>Hepatic toxicity, anemia, potential teratogen (contraception)</td>
</tr>
<tr>
<td></td>
<td>Sitaxsentan, Ambrisentan</td>
<td></td>
</tr>
<tr>
<td>Inhaled Nitric Oxide</td>
<td>NO: 20-40ppm, special delivery equipment</td>
<td>Rebound hypertension, Methemoglobinemia</td>
</tr>
<tr>
<td>(=) Selective pulmonary vasodilation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Phosphodiesterase type 5 inhibitors</td>
<td>Sildenafil</td>
<td>FDA warning: increase in mortality with long term therapy and high doses?</td>
</tr>
<tr>
<td></td>
<td>Tadalafil</td>
<td></td>
</tr>
<tr>
<td>Miscellaneous: Vasodilation, diuresis, anticoagulation</td>
<td>Oxygen, Diuretics, Coumadin</td>
<td></td>
</tr>
</tbody>
</table>
What’s so special about Pulmonary Hypertension?

Why are we so worried?
Right Ventricle ≠ Left Ventricle

- Triangular or crescent shape
- Inflow, apical and outflow area
- Thinner, more compliant, less contractile
- Muscle layers:
  - Circumferential superficial in continuity with LV
  - Longitudinal deep connected with septum
- RV contracts in peristaltic motion
- Ventricular Interdependence:
  - LV contraction augments RV CO by 40-50%
- Low impedance pulmonary vascular bed:
  - Short isovolemic contraction time
  - 60% of RV SV after peak pressure
  - RV stroke work only 25%
- Coronary blood flow throughout cycle

- Ellipsoidal shape
- Septum bows into RV throughout cycle
- Twisting/shortening motion
- High impedance systemic circulation
- Longer isovolumic contraction time
- Square shaped pressure volume loop
- Coronary blood flow only in diastole

Major differences in ventricular geometry and fiber orientation => Different performance
Ventricular Interdependence

Bronicki RA, Baden HP: Pathophysiology of Right Ventricular Failure in Pulmonary Hypertension, S15-22
## Manipulations of PVR

<table>
<thead>
<tr>
<th>Factors increasing PVR</th>
<th>Factors decreasing PVR</th>
</tr>
</thead>
<tbody>
<tr>
<td>PEEP</td>
<td>No PEEP</td>
</tr>
<tr>
<td>High airway pressure</td>
<td>Low airway pressure</td>
</tr>
<tr>
<td>Atelectasis</td>
<td>Normal FRC</td>
</tr>
<tr>
<td>Low FiO$_2$</td>
<td>High FiO$_2$</td>
</tr>
<tr>
<td>Acidosis</td>
<td>Alkalosis</td>
</tr>
<tr>
<td>Hypercapnia</td>
<td>Hypocapnia</td>
</tr>
<tr>
<td>Increased hematocrit</td>
<td>Low hematocrit</td>
</tr>
<tr>
<td>Sympathetic stimulation</td>
<td>Blunted stress response</td>
</tr>
<tr>
<td>Pain and agitation</td>
<td>Nitric oxide</td>
</tr>
<tr>
<td>Epinephrine, dopamine</td>
<td>Vasodilators (Milrinone, prostacyclin, etc.)</td>
</tr>
<tr>
<td>Direct surgical manipulation</td>
<td></td>
</tr>
<tr>
<td>Vasoconstrictors: phenylephrine</td>
<td></td>
</tr>
</tbody>
</table>
RV and PAH

Chronic changes:
- Hypertrophy of RV => dilation => decreased RV CO
- Septal deviation => impaired LV diastolic and systolic function
- Initially only with exercise, later at rest: dyspnea and chest pain

Acute rise in PA pressures and PVR
- Acute increase in RV afterload => RV dilation and decompensation, ischemia
- Triggered by hypoxia, hypercapnia, acidosis, hypothermia, noxious stimulus

=> Pulmonary Hypertensive crisis
Pulmonary Hypertensive Crisis?
Bradycardia – Hypotension – Cardiac Arrest

Pulmonary Hypertension => RV pressure load

RV systolic dysfunction -> RV diastolic dysfunction

Reduced RV output and diastolic RV Hypertension

Tricuspid regurgitation -> Diastolic Ventricular Interdependence

Reduced LV Filling and decreased Cardiac Output

RV ischemia -> Reduced LV assistance to RV EF -> Hypoxia and Acidosis

RV systolic dysfunction

RV diastolic dysfunction
Treatment

Decreasing PVR

• 100% oxygen
• Hyperventilation
• Adequate anesthesia
• Adjust ventilation
• Call for NO
• Treat acidosis with Bicarbonate
• Treat hypothermia

Supporting RV

• Inotropic support of RV
• Judicious fluid administration
• Maintaining coronary perfusion
• Support for LV
• With RV ischemia:
  => Vasoconstrictors
  • Norepinephrine
  • Phenylephrine
  • Vasopressin (?)
Preoperative Evaluation

• Careful risk/benefit evaluation of planned procedure

• History and Physical exam
  • CV symptoms, exercise tolerance, h/o syncope, arrhythmia etc. ?
  • Recent changes ?
  • Current medications?
  • Previous Anesthesia ?

• Review of diagnostic studies
  • ECG: evidence of ventricular hypertrophy, ST changes, arrhythmia
  • Echo: anatomy, RV size and function, estimated RV pressure, PFO or ASD ?, LV function
  • Cath: “gold standard”, pressures and response to vasodilators (oxygen and NO)

• Discussion with Cardiologist
Anesthetic Considerations

• Major anesthetic risk even for minor procedures
• **Perioperative risk not just an anesthetic risk!** => Day Surgery ??
• All techniques have been used successfully
• Maintain optimal PVR and avoid triggers of ↑ PVR
• Possible severe cyanosis if “pop off” present: => paradoxical emboli
• Spontaneous vs controlled ventilation?
  • Positive pressure ventilation = afterload for RV as is Hypoventilation with hypoxia and hypercarbia
• **Never switch off** NO or infusion of prostacyclins: => Rebound pHTN
Anesthesia Plan

• Thorough preoperative risk/benefit discussion with team
• Careful sedation if indicated and necessary
• Adequate ventilation strategy:
  • High inspired oxygen concentration, avoidance of hypercarbia
  • Mild hyperventilation without excessive peak airway pressures
  • Adequate tidal volume to avoid atelectasis, long expiratory phase, no or minimal PEEP
• Adequate depth of anesthesia for periods of intense stimulation
• Maintenance of normal preload for hypertrophied RV
• Early use of inotropic support for RV, Nitric oxide available.
• Maintenance of adequate coronary perfusion pressure
• Adequate postoperative monitoring and pain control
Back to our OR schedule …

Third Case:
• 1 mo with **PA/IVS** s/p repair for lap G-tube

“Can it get any worse?
Why me?
What did I do?”

What’s the story on this kid?

What is PA/IVS and what kind of repair?
Patient’s History

• Prenatal U/S diagnosis: **Pulmonary atresia with intact ventricular septum, hypoplastic RV**
• Born at 36 weeks via C/S for NRAFHR
• Stayed in NICU x 3d for r/o sepsis, on PGE₁
• Cath lab for possible RVOT perforation and PDA stent
  - **RV dependent coronary circulation**, arrhythmias with catheter manipulation
• To OR for Blalock Taussig Shunt and PDA ligation
• Prolonged ICU course post op
  - on multiple inotropes, extubated POD 7
Pulmonary Atresia and Intact Ventricular Septum

• ~ 3% of CHD: 4-8 / 100,000 live births
• Problem develops later than PA/VSD
  • Ventricular septum formed, PA’s often normal size

• Heterogeneous morphology:
  • Pulmonary valve atresia
  • Hypoplastic Right Ventricle
  • Hypoplastic tricuspid valve, tricuspid regurgitation
  • Interatrial communications: PFO or ASD
  • Duct-dependent pulmonary blood flow
  • Sinusoids & RV dependent coronary circulation?
Sinusoids

• Back to Embryology: Coronary arteries
  Plexus of endothelial lined channels penetrate the myocardium
  => Intratrabecular sinusoids
  Regression and merging of channels => Mid myocardial Network
  ⇒ Coronary arteries grow into aorta (“Invasion of aorta”)

Blood in hypoplastic RV with PA/IVS under high pressure
  → If no outflow: Sinusoids persist in developing fetal myocardium
  → Extensive Ventricular-coronary communications
  → Turbulent flow and endothelial injury result in coronary stenosis
  → Mechanism: Competitive flow – less saturated blood?

Moss and Adams’, 6th ed., Vol 1, Chapt 1, page 17
RV-Dependent Coronary Circulation

• Development of proximal coronary stenosis or atresia
  • Myocardium dependent on RV pressure for adequate perfusion
  • Functional single ventricle at risk for ischemia

• Dual coronary blood flow or distal stenosis
  • Risk for “RV steal” with low RV pressures

=> RV decompression can result in significant LV infarction
Management of PA/IVS

PGE₁ for ductal dependent pulmonary blood flow

Echo and Angiography
RV size – TV size and function – ASD/PFO – PDA – Sinusoids/RVDCC

Single Intervention or Surgery
- e.g. PV perforation, BAS

Multiple Procedures
- e.g. BTS-BDG-Fontan
- RV-PA conduits, TV repair

Transplant
- Bridge with PDA stent or BTS

Biventricular Repair

1 ½ Ventricle Repair

Single Ventricle Repair
Sinusoids, RVDCC and Outcome

Retrospective review: 1989-2004
32 patients with PA/IVS and RVDCC
Single Ventricle palliation
Median F/u 5.1y (9mo – 14.8y)
Overall mortality 18.8%
All deaths within 3 months of BTS

Retrospective review: 2000-2012
58 patients with PA/IVS, F/u 8.2y (0-11y)
17 (30%) Single ventricle palliation
10 (59%) RVDCC => survival 40%
7 (41%) non-RVDCC => survival 100%

Natural History of Pulmonary Atresia With Intact Ventricular Septum and Right-Ventricle–Dependent Coronary Circulation Managed by the Single-Ventricle Approach
Kristine J. Guleserian, MD, Laurie B. Armsby, MD, Ravi R. Thiagarajan, MD, Pedro J. del Nido, MD, and John E. Mayer, Jr, MD
Departments of Cardiovascular Surgery and Cardiology, Children’s Hospital Boston, Harvard Medical School, Boston, Massachusetts

Pulmonary Atresia/Intact Ventricular Septum: Influence of Coronary Anatomy on Single-Ventricle Outcome
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More Data from the Literature

- Overall mortality for PA/IVS: 19-42%
- RV-to-coronary artery fistulas in 31-68% of patients with PA/IVS
- RV-dependent coronary circulation present in 3-34%
  - Significant part of LV perfused via ventriculo-coronary fistulae by hypertensive RV
  - True prevalence unknown, different definitions, angiographic practices etc.
    - Multi-institutional studies: 5-9%, single institutional: 25-35% (referral pattern ?)
  - increased early mortality within 3 months, at or around time of BTS
    => Ischemia and significant LV dysfunction
- Aorto-coronary atresia: associated with 100% mortality => Transplant
- No real regression of ventriculo-coronary fistulae/stenosis over time!
  - No evidence of switch from RVDCC to non-RV dependent circulation
At risk for coronary ischemia and LV dysfunction!

Potential triggers:
- Brief period of hypotension
- Volume depletion
- Tachycardia (e.g. with fever)
- Diastolic run off via BTS

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Anesthesia Concerns for Non-Cardiac Surgery

• Preoperative Echo: LV function?
• Adequate Preload - “keep RV full”:
  • short NPO times, prehydration
• Monitoring for LV ischemia: 5 lead ECG, baseline strip
• Maintain adequate coronary perfusion pressures
  • Avoid hypotension and tachycardia
  • Hemodynamic support with norepinephrine, low dose epi or vasopressin
  • Avoid excessive diastolic run off via BTS, adjust ventilation accordingly
• Avoid increased myocardial $O_2$ consumption
• Single Ventricle Physiology with parallel circulation
Laparoscopic Surgery and PA/IVS with RVDCC

High Risk Procedure!

Abdominal insufflation: => Changes in pulmonary mechanics and CV filling
- Volume loaded ventricle unable to handle increased afterload
- Loss of preload and potential hypotension: **Increased risk for ischemia!**
- Limited pulmonary blood flow restricts ability to “blow off” additional CO$_2$
- Increased PVR can “unbalance” circulation

**Anesthesia:**
- Careful risk/benefit discussion
- Pre- and postoperative echocardiography
- Invasive blood pressure monitoring, 5 lead ECG, frequent ABGs.
- Low abdominal insufflation pressures (8-12mmHg)
- Transfusion to hematocrit of 40-45%
- Planned postoperative ICU admission
Summary: Williams, Pulm. HTN & Sinusoids

- High risk cardiac patients
- Careful risk/benefit discussion
- Thorough preoperative evaluation
  - Review of most recent cardiology note and imaging studies
  - Discussion with cardiologist
- Choice of appropriate venue, timing and staffing
- Detailed anesthesia plan and extended monitoring
  - 5 lead ECG, emergency drugs, defibrillator pads, extended PACU stay/ICU admission etc.

At risk for ischemia and ventricular dysfunction
=> Maintenance of adequate coronary perfusion pressure
References for Williams Syndrome


References for Pulmonary Hypertension


References for Sinusoids


5. Svrivastava D, Baldwin S. Molecular Determinants of Cardiac Development. In Moss and Adams’ “Heart Disease in Infants, Children and Adolescents” Vol 1. 6th ed. 2001 Lippincott Williams & Wilkins