The Role(s) of Cath in PVD

- Diagnosis
  - Rule in PVD
  - Rule out other causes
  - Assess for associated lesions
- Assess Severity / Reactivity
- Monitor / Modify Treatment
- Repair Associated Lesions
- Palliation / decompression
  - ASD creation
  - Potts Shunt creation

Catheterization Mechanics in PVD

- Timing
- Pre Cath Management
- Anesthesia / Sedation Medications
- Accurate Data: Its all about the flow!
  - Thermodilution / Fick and VO2 assessment
- Vasodilator Testing: When/What/How
  - 100% O2 / 40 ppm iNO / aerosolized iloprost (0.3–0.5 mg/kg)
- Post Cath Management

Pre Cath Management

- Multi-disciplinary approach
  - PHT team / Cardiology / Cardiac Anesthesia / CICU
  - Risk stratify pre-cath – normal vs high
- Admit
  - CICU vs pre-op
  - Modification of PHT medications / Diuretics
  - In hospital management of PHT infusions

Anesthesia Strategy

  - Spontaneous breathing when possible / safe
  - Agents with minimal cardiopulmonary effects
Risk Stratify

<table>
<thead>
<tr>
<th>Normal</th>
<th>High</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre – op admit</td>
<td>CICU admit</td>
</tr>
<tr>
<td>iNO available cath and post</td>
<td>iNO available pre/ transfer/ cath/ post</td>
</tr>
<tr>
<td>+/- arterial line</td>
<td>Arterial line</td>
</tr>
<tr>
<td>+/- intubation</td>
<td>+/- intubation</td>
</tr>
<tr>
<td>Wean iNO off post testing</td>
<td>iNO weaned slowly in CICU</td>
</tr>
<tr>
<td>CICU post +/- home same day</td>
<td>CICU overnight +</td>
</tr>
</tbody>
</table>

High Risk: history of syncope
suspected supra-systemic Pap
significant associated lung disease
and/or need for chronic oxygen therapy
on prostacyclin (Flolan/Epoprostenol IV,
Remodulin IV/SQ, Tyvaso INH)
other co-morbidities identified by the PH team

Wean iNO off post testing
iNO weaned slowly in CICU

Anesthesia Management

- Moderate Sedation vs General Anesthesia
- Spontaneous Breathing vs Ventilated
- Meds:
  - Oral agents – Versed / Valium
  - IV agents – Dexmedetomidine, Ketamine, Versed
  - Inhaled agents - sevoflurane
- Post cath airway management if intubated
  - Extubated in cath lab vs CICU

UCSF Cath Procedure

- Right and Left heart diagnostic catheterization
  - Vascular US guided access – usually femoral
- Thermodilution catheters when possible
  - if not
  - Fick with measured VO2 (if possible)
- Angiography: limited
  - segmental wedge angiograms
  - Associated lesions

Segmental Wedge Angiogram
Hemodynamic Evaluation

8 mo Adams-Oliver syndrome developed
PV stenosis s/p sutureless repair

100% O2 + 40 ppm iNO

Qs 3.5, Qp 2.1, Qp:Qs 0.6,
Transpulmonary grad: 78 mmHg

R̄p 34.6 Wui
OUCH!!!

Pulmonary Vascular Resistance
(PVR = Rp)

\[
PVR = PA \text{ mean} - LA \text{ mean pressures}
\]

Pulmonary Flow

LA mean = Pulm Vein mean = PCWP
Pulmonary Flow = Qp

PVR reported indexed Woods units = mmHg/L/min/M²

Normal PVR = 18 mmHg − 10 mmHg = 2.3 Wui

3.5 L/min/M²

Normal Range: 1 to 3.5 Wui

Pulmonary Flow Measurement

- Accuracy Matters
- Measurement Options
  - Thermodilution – gold standard - except
    - Intra-cardiac shunt, TR, PI
  - Fick – using O2 content
    - \[ Qp = \frac{VO2}{(PV – PA O2 content) \times Hgb \times 13.6} \]

Common Errors
- Dissolved O2
- Estimating VO2
O2 Content: Dissolved O2

2 mo old - s/p CDH repair - PDA

\[ \text{With Dissolved} \]
\[ R_P = \frac{165}{(9.74 - 5.57) \times (13.6)} = 2.91 \text{ Wi} \]

\[ \text{No Dissolved} \]
\[ R_P = \frac{165}{(8.53 - 5.48) \times (13.6)} = 3.98 \text{ Wi} \]

\[ \text{O2 Content Bound} = \text{Sat} \times \text{Hgb} \]
\[ \text{Dissolved} = \text{PaO2} \times 0.003 \]
\[ \text{Ao} = 0.98 \times 8.7 + 403 \times 0.003 = 8.926 + 1.21 (14\%) \]

Measuring VO2 in Cath Lab

UCSF VO2
2016-17 data
N=34
Age 3-53 yr
Wt 15-95 kg

Thermo VO2 + 4.2%
Ult. Series VO2 + 9.6%
La Farge VO2 + 14.9%
Seckeler Chart VO2 -4.0%
Att est VO2 + 4.2%

Asymmetric Qp or PA pressures: Affect on PVR

20% Left; 80% Right

13 mo ex29 week premie, severe chronic lung disease (tracheostomy), left PV stenosis, & pulmonary hypertension
PVR Calculation

Normal flow and Pressures
(RPA = LPA pressures
Flows aprox 50:50)
Right + Left = Lung bed
so
Rp = MPA - PCWP
Qp

Asymmetric Flows or Pressures
(RPA ≠ LPA pressures
Or Abnl Flows)
Right & Left calculated seperately
so
Rt Rp = RPA – PCWP
Lt Rp = LPA – PCWP
Qrp Qlp

1 = 1 + 1
Total Rp Rt Rp Lft RP

13 mo ex 29 week premie
Severe chronic lung disease
Tracheostomy
Left PV stenosis
Pulmonary hypertension

Vasodilatory Testing

• How
  – Establish baseline – prefer RA breathing spontaneously
  – Adjust to patient’s severity
  – Vasodilator stable x 5+ minutes
• What
  – 100% O2
  – iNO – 40 ppm
  – Inhaled iloprost
• Response
  – ↓ mPAP of ≥ 10 mmHg to <40 mm Hg with a normal CI
  – ↓ mPAP ≥20%, no change in cardiac index, and a decrease or no change in PVR/SVR ratio
• Post testing management
  – DC – wean – continue?

4 yo new Dx PHT with beta thal + G6PD deficiency – response to iNO

2 L NC

100% O2 + 40 ppm iNO
Response to inhaled iloprost

Baseline:
- 100% O2
- 40 ppm iNO

2mo s/p CDH repair: No response?

PVR = Cross sectional bed + vascular tone

Post Cath Management

<table>
<thead>
<tr>
<th>Normal</th>
<th>High</th>
</tr>
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<tbody>
<tr>
<td>+/- extubation in lab</td>
<td>Transfer to CICU intubated</td>
</tr>
<tr>
<td>Wean iNO in lab slowly with monitoring</td>
<td>Transfer on iNO - wean slowly in CICU per PHT team</td>
</tr>
<tr>
<td>CICU post +/- home same day</td>
<td>CICU overnight +/- as needed</td>
</tr>
<tr>
<td>Modify / escalate PHT Rx as needed</td>
<td>Modify / escalate PHT Rx as needed</td>
</tr>
</tbody>
</table>

Risks of Cath in PHT

- Age: 1-17, mean 7.3 yrs
- PVR: 3-97, mean 16.6

Univariate risk factors
- FC ≥ 3
- Gen Anesthesia

### Interpretation of Cath Data

#### Table 3: Hemodynamic definitions of PH (modified from Galiè

<table>
<thead>
<tr>
<th>Definition</th>
<th>Invasive measures</th>
<th>PH group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary PH</td>
<td>mPAP ≥25mmHg</td>
<td>1, 1.5</td>
</tr>
<tr>
<td>RVSP ≥35mmHg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary PH</td>
<td>mPAP ≥25mmHg</td>
<td>2</td>
</tr>
<tr>
<td>RVSP ≥35mmHg</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

#### AHA/ATS Consensus Pediatric PAH: Disease Severity

<table>
<thead>
<tr>
<th>LOWER RISK</th>
<th>DETERMINANTS OF RISK</th>
<th>HIGHER RISK</th>
</tr>
</thead>
<tbody>
<tr>
<td>PVR 0-10 WU/㎡</td>
<td>Hemodynamics</td>
<td>PVR 10-30 WU/㎡</td>
</tr>
<tr>
<td>PA/SVR 0-8.5</td>
<td></td>
<td>PA/SVR 8.5</td>
</tr>
</tbody>
</table>

- **AHA/ATS – Circ 2015**

### Surgical Repair Criteria: Shunt Lesions

- **AVT Complementary**
  - Operation / Standard PH Management
  - + AVT

- **AVT Necessary**
  - AVT
  - Operation / Standard PH Management Post-op / Consider Fenestration
  - - AVT

- **Reoperate Catheterization**
  - High Risk
  - Consider Fenestration
  - Probably Inoperable

- **Operation / Standard PH Management Post-op / Consider Fenestration**
  - + AVT

- **Reoperate Catheterization**
  - - AVT

### Treatment Options in the Cath Lab

- **Treat Associated Lesions**
  - PDA closure
  - ASD
  - Close
  - Modify
  - PV stent repair

- **Decompressive Palliation**
  - ASD creation
  - Potts shunt creation

### 2 mo s/p CDH repair with PDA

- **PDA right to left shunt, supra-systemic PAP post repair**
- **Rx ventilation + iNO + Sildenafil**
- **Shunt reversed (left to right) at 6 weeks – weaned off vent & iNO**
ASD Closure
- 47 yo with Sjogren’s lung disease, ASD, pulmonary HTN
- Baseline RA: Qs 5, Qp 4.7 (0.9), PA 80/27 m 46 Rp 8.2 Wui
- 100% O2 + 40 ppm iNO: Qp:Qs 1.2, PAm 38, Rp 6.5 Wui
- 25 mm sec ASD

ASD Creation
- Palliation for severe symptomatic patients on maximal med Rx
- Trade sustained output for cyanosis – too large – high risk

PA to DAO shunt – reverse Potts
- Palliation for severe symptomatic patients on maximal med Rx
- Trade sustained output for cyanosis (lower body) – too large - very high risk
- Advantage over atrial shunt – immediate decompression of RV, desaturated blood only to lower body

Conclusions
- Cath is a critical tool in the management of children with PHT
  - Initial diagnosis
  - Ongoing medical management
  - Treatment of associated lesions
  - Palliation for end stage and symptomatic pre transplant patients
- Multi-disciplinary structured team approach crucial to optimize outcomes and minimize complications in this very sick population