Hemodynamic assessment prior to closure of heart defects in presence of pulmonary hypertension

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Hemodynamic assessment in heart defects with pulmonary hypertension?

1. Why worry about pulmonary hypertension?
2. How do we decide on surgical closure of heart defects in presence of pulmonary hypertension?
3. Should we do anything different for catheter closure of heart defects?
4. Should we do anything different after the availability of newer therapy for pulmonary hypertension?
Congenital Heart Disease (L-R shunts) and Pulmonary Hypertension

- **Left-to-right shunt**
  - Increased pulmonary blood flow (shear stress/circumferential stretch)
  - Endothelial dysfunction and vascular remodeling
    - Smooth muscle cell proliferation, increase in extracellular matrix, intravascular thrombosis
  - Increase in PVR
  - Inverted shunt: right-to-left
  - Cyanosis (Eisenmenger syndrome)

Congenital Heart Disease

<table>
<thead>
<tr>
<th>Year</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1940</td>
<td>Surgery for PDA</td>
</tr>
<tr>
<td>1950</td>
<td>Paul Wood: Eisenmenger syndrome, Open heart surgery for common shunts</td>
</tr>
<tr>
<td>1960</td>
<td>Early Open Heart surgery in Infants</td>
</tr>
<tr>
<td>1970</td>
<td>Infant open heart surgery widely established in developed countries</td>
</tr>
<tr>
<td>1980</td>
<td>Infant open heart surgery in selected parts of the developing world</td>
</tr>
<tr>
<td>1990</td>
<td></td>
</tr>
<tr>
<td>2000</td>
<td></td>
</tr>
</tbody>
</table>

100% Percentage of infants with large VSD receiving timely surgery

Developed world

Developing world
Operations for CHD (Global Situation): Data from Year 2000

- Total Open Heart Procedures: 1,211,624
- Total Congenital operations: 159,482
- Adult Congenital: 25,556
- Pediatric Congenital: 81,506
- Infant/Newborn: 52,419
Estimated proportion of infants and newborns in India with critical CHD undergoing surgery within the first year of life.

- **1998**: Estimated number of infants with critical CHD
- **2004**: Number undergoing surgery in the first year of life
- **2008**: Number undergoing surgery in the first year of life

** RAW TEXT END**
Parts of The World Where the Average Child in the Region has Access to Congenital Heart Surgery
Late Diagnosis of CHD: Consequences

- Survivors selected by natural history
- Shunt lesions: Most children with large shunts with \( \uparrow \text{Qp} \) would not survive beyond 2 years of age
- Relatively large proportion of \( \uparrow \text{PVR} \) among survivors
Why worry about pulmonary hypertension in CHD?

- Most of the world has limited access to congenital heart surgery
- ASD can present late anywhere
- An occasional VSD or PDA escape detection may present later in childhood
- Insights into mechanisms of development and progression of pulmonary hypertension
What determines the development of pulmonary vascular obstructive disease (PVOD)?

Pre vs. post tricuspid
Size
Associated lesions: pulmonary venous hypertension

Anatomy of defect

Associated conditions

Lungs and airways
Altitude
Syndromes: Tri-21

Time

Unknown influences

Genetic???
Determinants of development of PVOD in L-R shunts

• Pre-tricuspid shunts:
  – gradual ↑ in Qp as RV accommodates and enlarges
  – ASD, PAPVC

• Post tricuspid shunts: Direct transmission of pressure head:
  – VSD (systolic),
  – PDA, AP-Window (systolic and diastolic)
Large Fossa ovalis ASD
SV ASD
Unrestrictive VSD or PDA
Single ventricle variants
Truncus
TGA
VSD/PDA

Likelihood of operability

100%

Age

Infancy
Early childhood
Adolescence
Adulthood

Concepts: Defect vs. PVOD Risk

Likelihood of operability
Risk of development of PVOD: Other (unknown) influences

Remarkable individual variability

- ASD with PAH in an infant
- VSD with shunt reversal in an infant
- Operable AP window in a teenager
- Operable large VSD in an adult

Prediction for an individual patient is sometimes quite challenging
Case example

• 5 month old, first seen in 2002, large fossa ovalis ASD, pulmonary hypertension, RV systolic pressure 54 + RA pressure, L-R shunt (vigorous)
• Symptomatic, tachypnea, failure to thrive, heart failure
• Catheterized to “understand the hemodynamics better”
Case example....

Operated in the same admission with uneventful post-operative course

<table>
<thead>
<tr>
<th>HEMODYNAMIC DATA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Condition 1:</td>
</tr>
</tbody>
</table>

A) Oxymetry and pressure data were recorded for SVC, RA, PA, LV, AO, PV, in
   Basal state and After administration of O2 at 10L/min for 10 minutes which revealed
   - Large L->R Shunt
   - QP/QS::3.1>1
   - PVRi:2.3
   Large L->R Shunt
   QP/QS::>4
   PVRi:0.29

B) Elevated right heart filling pressure and Moderate PAH.

C) Angios:
   - LUPV Injection PA view normal drainage to LA
   - PA angiogram PA view showed confluent PA anatomy, RPA:6.8MM, LPA:5.7MM, normal arborization, no evidence of pulmonary stenosis.
   - LV angio LAO, Cran. View showed normal LV, normal coronaries, and arch vessels.

PLAN

Atrial Septal Defect Closure
Seven years later...
Case Example #2

• 17 year old
• Detected to have CHD in the early infancy. Cardiac cath planned but could not be accomplished.
• Relatively “asymptomatic”
• Occasional episodes of respiratory infection+
• AV malformation of calf, heart disease needed to be fixed before vascular surgery.
Case #2: Physical Exam

- HR: 110/min, RR: 32/min.
- SpO2 on room air 97%.
- CVS: S1 normal, S2 normally split with loud P2, grade 5/6 Pansystolic Murmur at present at LLSB with a clear mid diastolic murmur at left 4th ICS.
Case #2: Echo

- Large inlet VSD (2 cm), gradient of 35 mm Hg
- Predominant L-R flows
- Grade I straddling of tricuspid valve
- LA and LV enlargement
- Flow acceleration in the pulmonary valve (gradient of 30 mm Hg)
Case #2: PFT

<table>
<thead>
<tr>
<th>Parameter</th>
<th>UM</th>
<th>Description</th>
<th>Pred.</th>
<th>SD</th>
<th>TEST#1</th>
<th>%Pred.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Best FVC</td>
<td>l(btps)</td>
<td>Best Forced Vital Capacity</td>
<td>4.59</td>
<td>0.56</td>
<td>2.18</td>
<td>47.4</td>
</tr>
<tr>
<td>Best FEV1</td>
<td>l(btps)</td>
<td>Best Forced Exp Volume in 1 sec</td>
<td>3.95</td>
<td>0.47</td>
<td>1.85</td>
<td>46.7</td>
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<td>0.47</td>
<td>1.85</td>
<td>46.7</td>
</tr>
<tr>
<td>PEF</td>
<td>l/sec</td>
<td>Peak Expiratory Flow</td>
<td>9.03</td>
<td>1.11</td>
<td>7.02</td>
<td>77.7</td>
</tr>
<tr>
<td>PIF</td>
<td>l/sec</td>
<td>Peak Inspiratory Flow</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>FEV1/FVC%</td>
<td>%</td>
<td>FEV1 as % of FVC</td>
<td>84.0</td>
<td>7.2</td>
<td>84.9</td>
<td>101.1</td>
</tr>
<tr>
<td>FEF25-75%</td>
<td>l/sec</td>
<td>Forced mid-expiratory flow</td>
<td>4.81</td>
<td>0.96</td>
<td>2.64</td>
<td>55.0</td>
</tr>
<tr>
<td>MEF75%</td>
<td>l/sec</td>
<td>Max Exp Flow @ 25% FVC</td>
<td>7.63</td>
<td>1.57</td>
<td>6.61</td>
<td>86.7</td>
</tr>
<tr>
<td>MEF50%</td>
<td>l/sec</td>
<td>Max Exp Flow @ 50% FVC</td>
<td>5.09</td>
<td>1.21</td>
<td>3.14</td>
<td>61.7</td>
</tr>
<tr>
<td>MEF25%</td>
<td>l/sec</td>
<td>Max Exp Flow @ 75% FVC</td>
<td>2.42</td>
<td>0.72</td>
<td>1.27</td>
<td>52.7</td>
</tr>
<tr>
<td>FET100%</td>
<td>sec</td>
<td>Forced Expiratory Time</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IC</td>
<td>l(btps)</td>
<td>Inspiratory Capacity</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Restrictive physiology
Case #2: Hemodynamic data

<table>
<thead>
<tr>
<th>Chamber</th>
<th>Sys(v)</th>
<th>Dias(a)</th>
<th>Mean</th>
<th>O2 %</th>
</tr>
</thead>
<tbody>
<tr>
<td>SVC</td>
<td></td>
<td></td>
<td></td>
<td>66.3%</td>
</tr>
<tr>
<td>RA</td>
<td>11</td>
<td>12</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>PA</td>
<td>85</td>
<td>29</td>
<td>51</td>
<td>87.8%</td>
</tr>
<tr>
<td>PA wedge</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PV</td>
<td></td>
<td></td>
<td>98 (a)</td>
<td></td>
</tr>
<tr>
<td>RV</td>
<td>75</td>
<td>ED= 10</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ao</td>
<td>110</td>
<td>60</td>
<td>88</td>
<td>97.5%</td>
</tr>
</tbody>
</table>
## Case #2: Hemodynamic data

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Condition 1 (room air)</th>
<th>Condition 2 (FiO2 100)</th>
<th>Condition 3 (NO 40PPM + O2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>O2 Consumption</td>
<td>135</td>
<td>143</td>
<td>143</td>
</tr>
<tr>
<td>Qp</td>
<td>7.54</td>
<td>20.22</td>
<td>19.13</td>
</tr>
<tr>
<td>Qs</td>
<td>2.47</td>
<td>3.54</td>
<td>5.39</td>
</tr>
<tr>
<td>Qp/Qs</td>
<td>3.06</td>
<td>5.72</td>
<td>3.55</td>
</tr>
<tr>
<td>PVRI</td>
<td>5.43</td>
<td>1.98</td>
<td>1.46</td>
</tr>
<tr>
<td>SVRI</td>
<td>29.2</td>
<td>16.7</td>
<td>17.26</td>
</tr>
</tbody>
</table>

VSD closure done; Normalization of PA pressures in the post op. period
Other (?Genetic) Influences

Risk of PVOD

Least

Most

IPAH??
Operability in L-R shunts

• How do we decide on operability of L-R shunts today?
  – What tools do we have?
  – What are their limitations?
Deciding operability of L-R shunts

• Clinical evaluation
• Chest X-ray and ECG
• Measurement of oxygen saturation
• Echocardiography
• Resting and post exercise ABG (PO2)
• Cardiac catheterization
• MRI

Viswanathan S, Kumar RK, Assessment of operability in congenital cardiac shunts with increased pulmonary vascular resistance, Cathet Cardiovasc Interv. 2008; 71:665-70
What principles govern decision on operability?

• Post tricuspid shunts:
  – Operable if significant shunt in the basal state irrespective of PA pressure

• Pre-tricuspid shunts:
  – Pulmonary hypertension (anything more than mild) warrants concern especially if basal shunt is not obvious
Deciding operability: Principles

• Age: benefit of doubt to younger patients.
• Lung, airway and ventilation issues can elevate PVR and confound assessment
• Pulmonary venous hypertension can result in reversible elevations in PVR
Clearly Operable: Cath not required
26 year old
Blue
Single loud
S2
Clearly Inoperable: Catheterization unnecessary
Clinical spectrum of post-tricuspid shunts with PAH

**Operable**

| Clear clinical/noninvasive evidence of a large left – right shunt | • Failure to thrive, ↑ precordial activity, mid diastolic murmur at apex, |
| • Cardiac enlargement, ↑ pulmonary blood flow |
| • Q in lateral leads on ECG, good LV forces |
| • LA/LV enlargement, exclusively L-R flows across the defect |

**Inoperable**

| Clear evidence of shunt reversal resulting from high PVR. | • Cyanosis, quiet precordium, no MDM |
| • Normal heart size, peripheral pruning |
| • No Q in lateral leads, predominant RV forces |
| • No LA LV enlargement, significant R-L flows across the defect |

**Borderline clinical non-invasive data: uncertain operability**
Ideally:

- **Operable**
  - Cath
  - PVR < 5-7 Wood units
  - Decline in PVR after vasodilators

- **Inoperable**
  - PVR > 5-7 Wood units
  - No decline in PVR after vasodilators
PVR Estimation by Cardiac Catheterization

PVR = \[ \frac{\text{Pulmonary artery mean pressure} - \text{Pulmonary venous mean pressure}}{\text{Pulmonary blood flow}} \]

\[ \frac{\text{Oxygen consumption}}{\text{PVO2 content} - \text{PA O2 Content}} \]
Sources of Error / Limitations in Catheterization Data

- Assumed oxygen saturations
- Assumed pulmonary vein saturation
- “Non-physiologic” state
- Calculated PVRI (basal and post-pulmonary vasodilator) has not been adequately standardized against the gold standard “surgical outcome”
Limitations of Echo for CHD

In the Real World……

Operable

Borderline

Operable

Inoperable

Operable

Inoperable

Borderline
What else can be done in the cath lab?

• Test occlusion of the defects:
  – ASD
  – PDA

• Little validation with long term data

• Immediate reduction of PA pressure may not translate into long term benefits
Illustrative Example

• 16 year old boy, 9.4 mm duct
• Nearly systemic PA pressures (Ao 120/60, mean: 90; PA 110/60: mean: 80)
• LL O2 Saturation: 96%
• Qp/Qs: 1.15:1 (Qp 3.8; Qs: 3.3)
• Basal PVRI: 18.75 Wood Units; PVRI/SVRI ratio: 0.66
Illustrative Example

Balloon Occlusion
Illustrative Example

5 minutes after balloon occlusion

Ao: 125/77 (96)
PA: 66/18 (41)
Pulmonary Hypertension in ASD

- Clinical clues: limited utility
- Clear evidence of flow reversal (sats < 90%) suggests ↑↑ PVR
- Echo evidence of elevated PA pressure (RVSP > 50-60 mm Hg): Cause for concern, need for careful assessment of hemodynamics
- A decline in PO2 on exercise (>mm Hg 10 mm Hg) suggests high PVR (>7 wood units)
Should we have different thresholds for closing defects in the cath lab (vs. surgery)

- Answer outside the cath lab: No
- Practice inside the cath lab: ??
- Since it is easier: Lower thresholds?
- Think long-term
- Fenestrated ASD device
Should we change our practice because we now have some drugs for PAH?

- Lifelong requirement?
- Outcomes when compared to natural history?
- Expense and availability
The Way Forward......

We need data

- Need for good prospective studies
- Cut-offs
- Basal PVR, O2, NO, O2+NO
- Test-occlusion in the cath lab
- Meticulous follow-up after closure
- SCAMPS??
Standardized clinical assessment and management plans (SCAMP)

1. All patients assessed and managed the same way unless the data center is notified in writing
2. Precise entry criteria
3. Prescribed and enforced follow-up
4. Deviations from protocol are allowed but must be justified
5. *Not randomized, not research, no IRB*
6. Lends itself to multicenter study
Conclusions

• We need better data on our patients.
• Until then: Comprehensive approach and look at all data available (clinical, non-invasive, hemodynamic)
• Guidelines based on combined experience and wisdom
• Holistic and not “hole-istic”