Surgical Approaches to Advanced Pulmonary Vascular Disease

Children with Congenital Heart Defects with large L>R shunts (VSD, PDA, AVC, TGA/VSD, etc.,) will develop over time pulmonary vascular obstructive disease.

Historically children with bi-directional shunting and/or a pulmonary vascular resistance of 10 Wood units or > were considered inoperable.

Surgical intervention at this stage has been historically fraught with high surgical mortality and in survivors the development of pulmonary hypertension.

Historical Perspectives

Several institutions reviewed the results of VSD closure, even re-cath some children following surgery years later.


All concluded that operating on children late in age, with elevated pulmonary vascular resistance increased early mortality and in a significant number resulted in increasing pulmonary artery pressure over time with late deaths as progressive pulmonary vascular obstructive disease became apparent.

Historical Perspectives

The development of pulmonary vascular obstructive disease was analyzed. Several institutions performed analysis of the pulmonary vascular bed in children with L>R shunts and pulmonary hypertension; 1- Heath and Edwards (Thorax-1958) 2- Rabinovitch (Circulation-1984), 3- Fried (Pediatr Cardiol-1986). Thus establishing that pulmonary vascular changes occurred in this set of patients, both before and after operation.
Historical Perspectives

Based upon previous surgical results and pathological studies a number of institutions initiated earlier surgical intervention in children with L>R shunts, to prevent the progression of pulmonary vascular obstructive changes. Castaneda (ATS-1976), Malm (JTCVS-1978), Stark & de Leval (BHJ-1979), Kirklin (JTCVS-1980).

Current Era

The strategy to operate on children at a younger age reduced the occurrence of the development of pulmonary hypertension late post-operatively, but lead to another issue, pulmonary hypertensive crisis (PHC) secondary to the reactivity of the pulmonary vascular bed in these younger children Bando (1996-JTCVS) with high flow pulmonary hypertension. Mortality from these events was up to 20% depending upon the defect.

Current Era


Post- Operative Pulmonary Hypertension

The occurrence of pulmonary hypertension following surgery for congenital heart disease appears to be decreasing.

The mortality and morbidity from pulmonary hypertension is still significant.

Improved care and earlier age at operation may be contributory.
VSD Closure Current Status: North America

- VSD operations performed at 74 sites in North America between 2005 and 2009.
- 5,150 operations
- Aggregate mortality, 0.64%
- Center range mortality, 0% - 5.1%
- Median age, 10 months, ATS-2011, 2010

VSD Mortality in non-First World

Why is the mortality in non-first world countries for VSD still higher?

* Advanced age at presentation, patients present with PHT and elevated PVR
* Few ECMO programs
* Poor or NO access to sophisticated medicines to treat PHT

VSD Closure; Past and Current Status, Non-First World Countries

- Mortality in Russia between 1964-85 was 19.2% for completion stage of two-stage repair and 6% for primary banding stage.
- Mortality in Turkey for isolated VSD closure between 1983-1999 was 6.4%.
- Mortality in South Africa between 1980-90 was 11% for black children.
- Mortality in Russia for “radical correction” of VSD in 1991 was 11%.

Flap Valved Double Patch (DFV) VSD Closure: History

- 1994- First asked to develop an operation which would decrease the mortality of children with PHT undergoing VSD closure.
- 1995- Zhou published initial results using same principal, 9.2% mortality with closure of simple VSD’s. (ATS, 1995)
- 1996- First VSD closed utilizing technique we had adopted from the atrial flap technique previously published by several groups.
- 1997- First TGA/VSD correction with DFV
- 1998- Published our experience with DFV 0% mortality.
Flap Valved Double Patch (DFV) VSD Closure; Artist’s Concept

Preliminary Experience with DFV

Diagnosis, Mortality and Follow-up

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>VSD</th>
<th>VSD/PDA</th>
<th>VSDmultiple</th>
<th>CAVC</th>
<th>ASO/VSD</th>
<th>Early Mortality</th>
<th>Late Mortality</th>
<th>Pre-op Sats</th>
<th>Pre-op PVR</th>
</tr>
</thead>
<tbody>
<tr>
<td>N=18</td>
<td>13</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>0/18</td>
<td>1</td>
<td>89%</td>
<td>11.4±4.1 W units</td>
</tr>
<tr>
<td>Maximal follow-up</td>
<td>18 months</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Intermediate Experience with DFV

Diagnosis, mortality and follow-up

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>N=56</th>
<th>Complex defects</th>
<th>TGA, DORV, AVC, IAA, Truncus, APW, etc.</th>
<th>N=35</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD ± ASD, PDA</td>
<td>13</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Mortality 3.6% (s), 14.3% (c)*</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Maximal follow-up 8 years.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Survival at Intermediate Follow-up

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV

Intermediate Experience with DFV
Preoperative Diagnosis and Early Mortality- Simple Defects

- VSD 89/5 (5.6%)
- VSDm 4/0 (0%)
- VSD/ASD 13/1 (7.8%)
- VSD/ASD/PDA 8/0 (0%)
- VSD s/p PAB 4/0 (0%)
- VSD/Coarct 1/0 (0%)
- VSD/SAM 1/0 (0%)
- VSD/AI 1/0 (0%)
- VSD/LPAatr. 1/0 (0%)
- VSD/TVRpr 1/0 (0%)

**TOTAL** 123/6 (4.8%)

Preoperative Diagnosis and Early Mortality- Complex Defects

- DORV (ALL) 24/2 (8.3%)
- DORV 18/2 (11.2%)
- DORVm 3/0 (0%)
- DORV/PDA 2/0 (0%)
- DORV/PAB 1/0 (0%)
- AVC (ALL) 20/4 (20%)
- AVC 11/2
- AVC/PDA 7/2
- AVC/PAB 2/2 (100%)
- Truncus Arteriosus (ALL) 10/4 (40%)
- Truncus Arteriosus 9/3 (33%)
- Truncus/LPA atr. 1/1 (100%)
- IAA/VSD (ALL) 8/1 (12.5%)
- IAA/VSD 7/0 (0%)
- IAA/DORV 1/1 (100%)
- Mitral Valve (ALL)/VSD 4/1 (25%)
- SVMR/VSD 3/0 (0%)
- MR/VSD/ASD 1/1 (100%)
- Miscellaneous Defects (ALL) 12/2 (16.7%)
- TGA/VSD 2/2 (100%)
- AP Window 2/0 (0%)
- TAPVC/VSD 2/0 (0%)
- LPA atr/VSD 2/0 (0%)
- Hypo Arch/PDA/VSD 2/0 (0%)
- PDA 2/0 (0%)

**TOTAL** 78/14 (17.9%)

Sildenafil in the Post-op Patient

- 35 consecutive patients with DFV who demonstrated elevated (50% or >) PA pressures received oral Sildenafil 1.0- 2.0 mgs/kg/dose (NG/oral).
- Mean age 7.2 years (1-28 yrs), PVR 7.8 (with oxy prov)
- 34/35 survivors (97.1% survival), the death was secondary to pneumonia 21 days post-op.

Current Protocol for VSD with Elevated PVR

- All patients undergo catheterization with oxygen provocation, NO if available.
- Children with simple defects who show response of 20% undergo Sildenafil dosing for 1 week then operative closure with DFV.
- Children with complex defects are treated depending upon defect and age. Sildenafil following cath at a minimum of 1 week before operation to 3 months.
- Intra-operatively, 3mgs/kg via NG post-induction, milrinone 50-75 mcgs/kg load with cross-clamp removal or for complex defects with re-warming.
- Post-operatively, all children allowed to awaken from anesthesia and are extubated when respiratory efforts satisfactory.
- Milrinone (0.5 – 1.0 mcgs/kg/min) and Sildenafil (2mgs/kg/dose, 4/daily) are maintained for 36 hours, milrinone weaned, ACE inhibitor started in concert with Sildenafil. Routine inotropes weaned as clinically indicated.
Where do we go?

- Currently obtaining follow-up on all children
- Developing research protocols to enable determination of pulmonary vascular endothelial response to; DFV, pre-treatment with Sildenafil, Endothelin blockers, other agents.
- Refining pre-treatment protocols for complex defect patients.

Summary

The treatment of children with bi-directional shunting across L>R shunts has been shown to result in low mortality and morbidity when the DFV technique is used.

Elevated PVR is not an indication of inoperability.

Earlier operation is the treatment of choice, but the situation outside the First World prohibits this approach.