Surgical Repairs for LVOT Obstructive Lesions

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Presented by
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Faculty Disclosure Statement:
I do not have any relevant financial relationship to disclose.
LVOT obstruction with normally related great vessels

- May be isolated, i.e. localized to one level
- May be multilevel and/or syndromic,
  - Hypoplastic Left Heart Syndrome
  - Shone Complex
  - Williams Syndrome
- May be associated with aortopathy
- May be genetically based and familial
- Is generally treatable but not often curable
- May be associated with a 1 or 2 V. repair
Hypoplastic Left Heart Syndrome

- Etiology: unclear
- Abnormal fetal flow patterns lead to stenosis or atresia of the left heart valves with hypoplasia of the LV
- Obstruction at the atrial septal level may play a role in underdevelopment of left heart structures
- Fetal echocardiography may permit early intervention
Hypoplastic left heart syndrome

Most common congenital cardiac lesion causing death within the first year after birth

Gillum et al, Epidemiology of congenital heart disease in the US. Am Heart J 1994
Evolving Survival Curves

Mahle et al, Circulation 2000
Arch reconstruction with continuous brain perfusion
Completed operation
## STS Outcomes 2008-2011
89,140 operations/ 103 centers

<table>
<thead>
<tr>
<th>PROCEDURE</th>
<th>INCIDENCE%</th>
<th>MORTALITY %</th>
<th>COMPL. SCORE</th>
<th>COMPL. LEVEL</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD patch</td>
<td>3.5</td>
<td>0.2 (0)</td>
<td>3.0</td>
<td>1</td>
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<tr>
<td>VSD patch</td>
<td>6.8</td>
<td>0.6 (0)</td>
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<tr>
<td>AVC repair</td>
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<td>2.4 (0)</td>
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<td>3</td>
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<tr>
<td>Art. Switch-IVS</td>
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<td>2.1 (0)</td>
<td>10.0</td>
<td>4</td>
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<tr>
<td>BT shunt</td>
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<td>6.4 (0)</td>
<td>6.3</td>
<td>2</td>
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<td>Norwood proc.</td>
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<td>16.5 (8.5)</td>
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<td>Cardiac Tx</td>
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<td>4.9</td>
<td>9.3</td>
<td>3</td>
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<tr>
<td>ALL ops.</td>
<td>100.0</td>
<td>3.4 (2.2)</td>
<td>7.3</td>
<td>2.4</td>
</tr>
</tbody>
</table>
Figure 1. Flow diagram depicting procedural, pregnancy, and postnatal outcomes among 70 fetuses that underwent attempted prenatal aortic valvuloplasty for AS with evolving HLHS.

240 fetuses evaluated
- Termination: 28
- No Rx, decision prenatal: 10
- Intrauterine death: 3
- No Rx, decision post-natal: 11
- Referred for Tx: 3
- 185 Norwood
• 185 Norwood (77.1%)
• Overall operative survival = 83.8%
• Standard risk group = 92.8%
• High risk group = 56.5%
• Why not do something different for the “high risk” hypoplastic?
The hybrid stage 1 palliation

How can the Hybrid Technique help in the management of HLHS?

- Stabilize poor surgical candidates, e.g. a good candidate with an acute problem
- Stabilize high risk candidates while their fate is being determined: Norwood vs OHT
- Avoid the “lock step” 3 stage approach for high risk sub group, e.g. obstructed atrial septum, LBW, extreme prematurity, high PVR
How can the Hybrid Technique help in the management of HLHS?

- The Hybrid can be Stage Ia in a multistage, i.e. > 3 stages, approach
- The Hybrid approach can help “sort out” the questionable LV
- Since the relationship between brain development, CBF, hypoxia and CPB is very complex. An eclectic approach using a flexible protocol may be the ideal method for treating complex, high risk HLHS patients
Lily: ex-1200 gm preemie/HLHS with AS/MS; S/P PA bands, PGE, no stent, Norwood/Sano @ 2200 gms.
The Shone Complex

- Shone, J D, Sellers, RD, Anderson, RC, Adams, P J r., Lillelei, CW, and Edwards, J E:
  - The developmental complex of “parachute mitral valve”, supravalvular ring of left atrium, subaortic stenosis and coarctation of the aorta
Subaortic Stenosis

- Isolated or Complex
- Etiology: steeper aorto-septal angle?
- Fibrous: discrete or tunnel
- Dynamic: IHSS
- Associated lesions:
  - VSD/ ASD
  - Arch anomaly/ PDA
  - Aortic Valve hypoplasia/ pathology
Modified Konno-Rastan procedure performed through a transaortic transventricular approach

Modified Konno-Rastan procedure performed through a transaortic transatrial approach

Aortic Valve Stenosis

- Isolated or Complex
- Neonatal critical AS is a special case
- Outcome is a function of age and size
- Associated lesions:
  - LVOT Hypoplasia
  - Sub AS
  - VSD/ ASD
  - Arch anomaly/ Coarctation/ PDA
Surgical Valvuloplasty is NOT a valvotomy

Surgical Valvotomy

Pre and Post-op gradients after surgical valvuloplasty

Fratz et al: Aortic Valvuloplasty in Pediatric Patients Substantially Postpones the Need for Aortic Valve Surgery

Circulation 2008: Munich HZ

- 188 patients
- Up to 17.5 years of follow-up
- < 1/12 years old: 60% free of valve surgery @ 10 years
- > 1/12 years old: 70% free of valve surgery @ 10 years
Figure 3. Survival after 10 years free from aortic valve surgery or second AoVP was 47% (95% CI, 32 to 62) in group <1 month and 63% (95% CI, 52 to 85) in group ≥1 month.

Figure 4. Survival after 10 years free from aortic valve surgery was 59% (95% CI, 45 to 73) in group <1 month and 70% (95% CI 59 to 81%) in group ≥1 month.


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Figure 5. Survival after 10 years was 71% (95% CI, 57 to 85) in group <1 month and 98% (95% CI, 96 to 100%) in group ≥1 month.
COMPLEX AORTIC VALVE REPAIR

COMPLEX AORTIC VALVE REPAIR IS NOT CURATIVE!

The “Scientific Basis” of the Ross Operation (Mr. Ross)

- At birth aortic and pulmonary valves are similar, if not identical
- In fetal life, the aortic and pulmonary valves function under similar hemodynamic conditions
- Therefore: When the post-natal pulmonary valve is asked to become an aortic valve it should easily adapt to its new role as the perfect AVR
The Ross Procedure: Systematic review and meta-analysis

• 39 reports from 2000-2008
• Consecutive/ pediatric/ adult
• “The Ross procedure provides satisfactory results for children and young adults. Durability limitations become apparent by the end of the first post-op decade, particularly in young patients”
Incision through the ascending aorta, aortic annulus, and right ventricular outflow tract (RVOT), exposing the interventricular septum.
Insertion of a prosthetic aortic valve with patch enlargement of the left ventricular outflow tract and the aortic annulus, and a second patch to close the right ventricular outflow tract.
Surgical techniques of posterior aortic root enlargement reported so far (Nick's-white arrow, Nunez's-black arrow, Manouquian's-black plus black dotted arrows)

Outcomes after mechanical AVR in children and young adults

- 30 pts. Median Age 14.3, range 7.6-24.7 yrs.
- Follow-up: mean age 21 yrs. Range 13-31 yrs.
- Median follow-up: 5.9 yrs.
- No mortality, early or late
- One reop for thrombosis, successful redo AVR
The size of prostheses and the type of insertion technique plotted by body weight of the patients at the time of initial implantation of the aortic prosthesis.

Freedom from events.

Postoperative years

<table>
<thead>
<tr>
<th>Patients/valves at risk</th>
<th>Survival rate</th>
<th>Re-AVR free</th>
<th>Event free</th>
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<tbody>
<tr>
<td></td>
<td>45</td>
<td>38</td>
<td>21</td>
</tr>
<tr>
<td></td>
<td>46</td>
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</tr>
<tr>
<td></td>
<td>46</td>
<td>36</td>
<td>20</td>
</tr>
</tbody>
</table>


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Supravalvular Aortic Stenosis

- Williams Syndrome
- Non-Williams SVAS
- May involve the coronary ostia!
- Often associated with pulmonary artery stenoses (we have used 7 patches!)
- May involve the entire aorta and its branches
- The aortic valve is usually trileaflet and reasonably functional (rarely replaced)
A preoperative catheterization of a supravalvular aortic stenosis patient that highlights the supravalvular stenosis

Aortogram showing a diffuse SVAS, involving the aortic arch vessels.

ArnÁiz E et al. MMCTS 2008;2008:mmcts.2006.002329
Lateral view of aortogram showing obstructive lesions located at the origin of the arch vessels.
Transection of the aorta at the level of the supravalvular stenosis (the sinotubular junction).
Explanatory diagram of the three perpendicular incisions in the aortic sinuses.

ArnÁiz E et al. MMCTS 2008;2008:mmcts.2006.002329

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Accommodation of the three pericardium patches in each Valsalva sinus.

 ArnÁiz E et al. MMCTS 2008;2008:mmcts.2006.002329
End-to-end anastomosis that connects proximal and distal aortic sides.

ArnÁiz E et al. MMCTS 2008;2008:mmcts.2006.002329
Peak pressure gradients according to type of repair, preoperatively, immediately after surgery and at last follow-up

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<tbody>
<tr>
<td>A valvuloplasty</td>
<td>1.1</td>
<td>1.8 (4.5)</td>
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<td>3</td>
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<tr>
<td>SubAS discrete</td>
<td>1.7</td>
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<td>6.3</td>
<td>2.5</td>
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<tr>
<td>SubAS Konno</td>
<td>0.1</td>
<td>0.0 (0)</td>
<td></td>
<td>3-4?</td>
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<td>AVR mech.</td>
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<td>1.9 (0)</td>
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<td>Ross AVR</td>
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<td>1.2 (0)</td>
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<tr>
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<td>Supra AS</td>
<td>0.2</td>
<td>1.3 (0)</td>
<td>5.5</td>
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MH 23yr.old S/P Ross 2001w 22mm. ascending aortic graft
CJ  22 yr. old S/P Ross 1989 (pre and post valve sparing root 2008)
NE 17 yr.old athlete Ross 1997; PVR/Aortic wrap 2008
(A) The Damus-Kaye-Stansel operation constructed with the hood technique keeps the pulmonary trunk free of distortion and tension on the valve

AGA 6 yr. old S/P DSK 2002 pre and post root revision 07/07
Prior to, and s/p DKS conversion
Summary I

- HLHS outcomes are improving
- Sub-aortic stenosis is easily treatable but sometimes recurs
- Complex aortic root surgery is feasible in small infants
- Modern imaging modalities provide excellent “roadmaps” for complex repairs
- Avoid AVR in infancy, if possible*

* Surgery rather than balloon valvuloplasty?
Summary II

- The Ross procedure as a root replacement is a very useful tool but it is not curative.
- The Ross procedure and the DKS procedure can provide an annuity for surgeons.
- The surgical treatment of failed Ross operations has created a new category of complex second and third procedures, e.g. “Reverse Ross”.
- Long term follow-up of our DKS patients reveals new and unusual pathology of the valves, aorta and the airways.
• Mechanical valves are a viable option
• Advances in mechanical valve design and anticoagulant management may further alter our thinking in the future
• Decellularized homografts/heterografts may become an option
• Ultimately, prosthesis choice is dictated by anatomy, pathophysiology, age, gender and lifestyle
• The feasibility of late TAVI/TPVI (valve in valve) may also alter prosthesis choice