AORTIC VALVE REPAIR IN CONGENITAL HEART DISEASE

Thomas L. Spray, MD

Chief, Cardiac Surgery

Alice Langdon Warner Endowed Chair
The Children’s Hospital of Philadelphia
Professor of surgery
The University of Pennsylvania
Disclosures

• I have no disclosures
AORTIC VALVE DISEASE IN CHILDREN

Aortic Stenosis:
- Critical AS of Newborn (Unicommissural)
- Congenital Valvar AS (Bicuspid or Tricuspid)
- Degenerative AS

Aortic Regurgitation:
- Truncus Arteriosus
- Result of In Utero or Perinatal Intervention
- Aortico-LV Tunnel
- Degenerative AR (Bicuspid AV, Marfan’s, etc.)
HISTORY OF AVR IN CHD

• 70’s - … Open valvotomy for critical AS, surgical valvuloplasty for older children
• 80’s – … BAV (1984), fewer open valvotomies
• 90’s – … BAV dominant, Ross/Complex LVOT operations increase
• 2000’s – … AV repair, valve-sparing root replacement, neonatal Ross-Konno
• 2010 – 14 … BAV?, AV Repair resurgence?, stent-mounted AVR, sutureless AVR, “supported Ross”
AV Repair

AV Repair – Freedom From Reintervention

AV Repair – Freedom From Reintervention By Procedure

### Table 2: List of Parameters

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Freedom from reintervention</th>
<th>Freedom from valve replacement</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>P value</td>
<td>HR</td>
</tr>
<tr>
<td>Sex</td>
<td>.948</td>
<td>1.04</td>
</tr>
<tr>
<td>Indication</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td>.971</td>
<td>0.98</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>.806</td>
<td>1.13</td>
</tr>
<tr>
<td>Mixed aortic stenosis/aortic regurgitation</td>
<td>.633</td>
<td>0.66</td>
</tr>
<tr>
<td>Concomitant cardiac procedure</td>
<td>.256</td>
<td>0.48</td>
</tr>
<tr>
<td>Previous cardiac surgery</td>
<td>.963</td>
<td>1.03</td>
</tr>
<tr>
<td>Age at surgery (d)</td>
<td>.229</td>
<td>0.99</td>
</tr>
<tr>
<td>Age &lt;1y at time of surgery</td>
<td>.048</td>
<td>2.89</td>
</tr>
<tr>
<td>Cusp extension technique</td>
<td>.020</td>
<td>3.34</td>
</tr>
<tr>
<td>Use of patch (excluding cusp extension)</td>
<td>.231</td>
<td>2.02</td>
</tr>
<tr>
<td>Initial valve morphology</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unicuspid valve</td>
<td>.908</td>
<td>1.14</td>
</tr>
<tr>
<td>Bicuspid valve</td>
<td>.581</td>
<td>1.34</td>
</tr>
<tr>
<td>Tricuspid valve</td>
<td>.535</td>
<td>0.71</td>
</tr>
<tr>
<td>Body surface area</td>
<td>.229</td>
<td>0.56</td>
</tr>
<tr>
<td>Crossclamp time</td>
<td>.344</td>
<td>1.01</td>
</tr>
<tr>
<td>Bypass time</td>
<td>.087</td>
<td>1.01</td>
</tr>
<tr>
<td>Significant regurgitation before surgery</td>
<td>.260</td>
<td>1.78</td>
</tr>
<tr>
<td>Significant stenosis before surgery</td>
<td>.887</td>
<td>0.93</td>
</tr>
<tr>
<td>Mean peak gradient before surgery</td>
<td>.847</td>
<td>1.03</td>
</tr>
</tbody>
</table>

Boldface types indicate values reaching a P value < .05 by univariate (freedom from valve replacement) and multivariate (freedom from reintervention) analysis. *HR*, Hazard ratio; *CI*, confidence interval.

---

AV Repair – Freedom From Replacement

AV Repair – Freedom From Reintervention

Figure 3. Kaplan-Meier graph demonstrating freedoms from any aortic valve reintervention and from aortic valve replacement (AVR) after surgical aortic valvuloplasty in entire cohort.

Figure E1. Kaplan–Meier graphs demonstrating freedom from aortic valve replacement (AVR) among patients with moderate or greater aortic stenosis (AS) before surgical aortic valvuloplasty and those with less than moderate aortic stenosis. Numbers of patients at risk at 0, 1, 3, 5, 7, 9, and 11 years are listed.

Figure 9. Survival and reintervention free survival in all patients.

Reintervention By Valve Lesion

Figure 11. Reintervention free survival in aortic stenosis, aortic insufficiency and mixed groups.

Fig 2. Kaplan-Meier estimates after aortic cusp extension valvuloplasty: freedom from aortic valve replacement (AVR; blue curve), and freedom from moderate or greater recurrent aortic insufficiency (AI; red curve) or aortic stenosis (AS; green curve; p = 0.2 by log-rank test).

AV Repair – AR Over Time

Fig 1. Durability of the repair: the trajectory of the regurgitant jet width/aortic valve diameter ratio over time is nonlinear and increases over time. The fine solid lines represent individual patient trajectories and the heavy solid line is a smoothing spline that represents the best-fit average trend over time. Time zero was taken to be the date of aortic valve surgery.

AV Repair – AS Over Time

Fig 3. Durability of repair: the univariable exploratory plot shows the trajectory of the peak aortic gradient over time. Aortic gradient remains stable until 1 year post-repair, at which time there is a sustained and nearly linear increase. The lighter solid lines represent individual patient trajectories and the heavy solid line is a smoothing spline that represents the best-fit average trend over time. Time zero was taken to be the date of aortic valve surgery.

Balloon Aortic Valvuloplasty versus Surgical Aortic Valvuloplasty
Aortic Valvuloplasty

Left Heart Growth, Function, and Reintervention After Balloon Aortic Valvuloplasty for Neonatal Aortic Stenosis
Doff B. McElhinney, James E. Lock, John F. Keane, Adrian M. Moran and Steven D. Colan
Circulation. 2005;111:451-458
Neonatal Open Valvotomy – Freedom from reoperation

The Long-Term Outcome of Open Valvotomy for Critical Aortic Stenosis in Neonates

Viktor Hraška, MD, Nicodeme Sinzobahamvya, MD, Christopher Haun, MD, Joachim Photiadiis, MD, Claudia Arenz, MD, Martin Schneider, MD, and Boulos Asfour, MD  (Ann Thorac Surg 2012;94:1519–26)
Open Valvuloplasty –

Freedom from reop based on anatomy

Log-rank test: $p=0.0052$
Open Valvuloplasty – Freedom from AVR

<table>
<thead>
<tr>
<th>Interval (Years)</th>
<th>Freedom (% ± SEM)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>92.4 ± 5.2</td>
</tr>
<tr>
<td>10</td>
<td>82.6 ± 8.0</td>
</tr>
<tr>
<td>15</td>
<td>67.8 ± 12.0</td>
</tr>
<tr>
<td>20</td>
<td>56.5 ± 14.4</td>
</tr>
</tbody>
</table>

Months: 0 24 48 72 96 120 144 168 192 216 240

Freedom in %: 0 20 40 60 80 100

5 Patients at risk
Freedom from AVR by anatomy

Log-rank test: $p=0.0011$
Figure 3. Time-related survival stratified by type of initial aortic valvotomy, adjusted for differences in group characteristics as reflected by a propensity score derived from multiple logistic regression. Solid lines represent parametric determination of continuous point estimates, and dashed lines enclose 70% CI. Abbreviations as in Figure 2.

Are Outcomes of Surgical Versus Transcatheter Balloon Valvotomy Equivalent in Neonatal Critical Aortic Stenosis?
Brian W. McCrindle, Eugene H. Blackstone, William G. Williams, Rekwan Sitiwungkul, Thomas L. Spray, Anthony Azakie, Richard A. Jonas and the members of the Congenital Heart Surgeons Society
Circulation. 2001;104:I-152-I-158
Figure 4. Non-risk-adjusted time-related freedom from subsequent aortic valve-related reintervention after initial aortic valvotomy (n=110), with patients who died before reintervention censored at time of death. Circles represent Kaplan-Meier estimates for freedom from reintervention at each event, with solid lines representing parametric determination of continuous point estimates, and dashed lines enclose 70% CI.
Figure 8. Time-related freedom from aortic valve-related reinter-
vention stratified by type of initial aortic valvotomy, adjusted for

differences in group characteristics as reflected by a propensity
score derived from multiple logistic regression. Solid lines repre-
sent parametric determination of continuous point estimates,
and dashed lines enclose 70% CI. Abbreviations as in previous
figures.
Surgical Valvuloplasty Versus Balloon Aortic Dilation for Congenital Aortic Stenosis: Are Evidence-Based Outcomes Relevant?  
John W. Brown, MD, Mark D. Rodefeld, MD, Mark Ruzmetov, MD, PhD, Osama Eltayeb, MD, Okan Yurdakok, MD, and Mark W. Turrentine, MD
BAV vs SAV – Freedom from >Mod AR

Fig 2. Freedom from more than moderate aortic regurgitation (AR) by the Kaplan-Meier method.
BAV vs SAV – Freedom from reintervention

![Graph showing freedom from AV reintervention over years with surgical and balloon methods compared.](image-url)

- **72%** for surgical method
- **53%** for balloon method
- **p=0.02**

**Patients at risk**:
- 85 at year 1
- 75 at year 2
- 62 at year 3
- 62 at year 4
- 40 at year 5
- 62 at year 6
- 40 at year 7
- 1 at year 8
- 38 at year 9
- 5 at year 10

Note: The graph illustrates the percentage of patients remaining free from AV reintervention over a 20-year period, with a notable difference between the surgical and balloon methods, indicating a statistically significant result.
BAV vs SAV – Freedom from AVR

- Surgical: 80% at 1 year, 75% at 2 years
- Balloon: 75% at 2 years

p = 0.32
## SAV Results

### Table 4. Surgical Aortic Valvotomy (Literature Review)

<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Patients</th>
<th>Age (mean) (y)</th>
<th>Time Interval (y)</th>
<th>Mortality (overall)</th>
<th>Redo Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Justo et al [12]</td>
<td>1996</td>
<td>90</td>
<td>n/a</td>
<td>3.6</td>
<td>15 (17%)</td>
<td>44% (31/70)</td>
</tr>
<tr>
<td>Chartrand et al [13]</td>
<td>1999</td>
<td>67</td>
<td>8.8</td>
<td>10.6</td>
<td>3 (5%)</td>
<td>24% (16/67)</td>
</tr>
<tr>
<td>Lambert et al [14]</td>
<td>2000</td>
<td>121</td>
<td>2.4</td>
<td>9.4</td>
<td>15 (12%)</td>
<td>50% (56/112)</td>
</tr>
<tr>
<td>Detter et al [3]</td>
<td>2001</td>
<td>116</td>
<td>13.7</td>
<td>23.8</td>
<td>29 (25%)</td>
<td>32% (37/113)</td>
</tr>
<tr>
<td>Bogers et al [15]</td>
<td>2001</td>
<td>11</td>
<td>2.7</td>
<td>4.8</td>
<td>1 (9%)</td>
<td>36% (4/11)</td>
</tr>
<tr>
<td>Alexiou et al [1]</td>
<td>2001</td>
<td>44</td>
<td>6.8</td>
<td>10.0</td>
<td>0</td>
<td>18% (8/44)</td>
</tr>
<tr>
<td>Brown [current study]</td>
<td>2011</td>
<td>89</td>
<td>7.1</td>
<td>9.8</td>
<td>2 (2%)</td>
<td>43% (38/89)</td>
</tr>
<tr>
<td>TOTAL</td>
<td>566</td>
<td></td>
<td>10.3 y</td>
<td></td>
<td>12% range, 3%-25%</td>
<td>34% range, 13%-50%</td>
</tr>
</tbody>
</table>
## BAV Results

### Table 5. Balloon Aortic Valvotomy (Literature Review)

<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Patients</th>
<th>Age (mean) (y)</th>
<th>Time Interval (y)</th>
<th>Mortality (overall) (%)</th>
<th>Redo Procedure (%) (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Justo et al [12]</td>
<td>1996</td>
<td>107</td>
<td>5.7</td>
<td>3.1</td>
<td>2</td>
<td>27 (28/105)</td>
</tr>
<tr>
<td>Borghi et al [16]</td>
<td>1999</td>
<td>90</td>
<td>13.7</td>
<td>5.1</td>
<td>16</td>
<td>48 (39/81)</td>
</tr>
<tr>
<td>Jindal et al [17]</td>
<td>2000</td>
<td>74</td>
<td>n/a</td>
<td>5.5</td>
<td>0</td>
<td>14 (10/74)</td>
</tr>
<tr>
<td>Balmer et al [4]</td>
<td>2004</td>
<td>70</td>
<td>2.2</td>
<td>1.7</td>
<td>9</td>
<td>35 (24/68)</td>
</tr>
<tr>
<td>Reich et al [10]</td>
<td>2004</td>
<td>269</td>
<td>n/a</td>
<td>5.3</td>
<td>10</td>
<td>29 (78/269)</td>
</tr>
<tr>
<td>Brown et al [18]</td>
<td>2010</td>
<td>509</td>
<td>2.4</td>
<td>9.3</td>
<td>9</td>
<td>44 (225/509)</td>
</tr>
<tr>
<td>Brown [current study]</td>
<td>2011</td>
<td>69</td>
<td>6.7</td>
<td>5.2</td>
<td>3</td>
<td>47 (32/68)</td>
</tr>
<tr>
<td>Total</td>
<td>2011</td>
<td>1188</td>
<td>5</td>
<td>7</td>
<td>range, 0%-16%</td>
<td>range, 14%-48%</td>
</tr>
</tbody>
</table>
Aortic Valve Reinterventions After Balloon
Aortic Valvuloplasty for Congenital Aortic Stenosis

Brown, D., Dipilato, A.E., Chong, E.C., Lock, J.E., McElhinney, D.B.
Freedom from AVR

Figure 5: Freedom From AVR

Kaplan-Meier curves depict freedom from aortic valve replacement (AVR) after balloon dilation for congenital aortic stenosis (AS) according to (A) age at initial intervention (N = neonate, M = months, Y = years), (B) history of prior aortic valve intervention elsewhere, (C) residual AS gradient after balloon dilation (≤ 29, 30 to 39, or ≥ 40 mm Hg), and (D) severity of aortic regurgitation (AR) after balloon dilation (N = none, T = trace, M = mild, S = moderate-severe).
Freedom from AVR by residual disease

**Figure 6** Freedom From AVR by Residual Valve Disease

Kaplan-Meier curves depict freedom from aortic valve replacement (AVR) after balloon dilation for congenital aortic stenosis (AS) according to 6 empirical groups of combined residual AS and aortic regurgitation (AR), defined as acute residual AS gradient at catheterization ≤35 mm Hg (blue) or >35 mm Hg (red) and AR as 0 to 1 (none or trivial), 2 (mild), or 3 to 4 (moderate-severe).
Surgical Valvotomy and Repair for Neonatal and Infant Congenital Aortic Stenosis Achieves Better Results Than Interventional Catheterization

Figure 1  Distribution of Procedures Over the Study Period

The diagram shows the distribution of cases of aortic valve procedure performed per year in children younger than 1 year in the Royal Children’s Hospital (surgical procedures versus balloon dilation). Balloon dilation was initiated in the early 90s and virtually abandoned in 2006.
BAV vs SAV – Freedom from reintervention in neonates

Figure 6
Freedom From Re-Intervention After Balloon Valvuloplasty Versus Surgery as a Primary Intervention in Neonates (<30 Days) and Infants (31 Days to 1 Year)

The comparison between balloon dilation and surgical valvuloplasty in neonates (A) and infants (B) shows a higher risk of re-intervention in neonates. In neonates and children, the risk of re-intervention was higher after balloon dilation than after surgical valvuloplasty (HR: 4.00; p = 0.001 and HR: 3.08; p = 0.047, respectively).
BAV vs SAV – Freedom from reintervention or AS

**Figure 5**

**Freedom From Re-Intervention or Significant Restenosis After Balloon Valvuloplasty Versus Surgery as a Primary Intervention**

At latest follow-up 19 of the 69 patients who did not undergo a re-intervention had at least moderate stenosis. The risk of re-intervention or stenosis was higher after a primary balloon dilation than after a surgical valvuloplasty. By multivariate analysis, balloon valvuloplasty was the only independent predictive factor of either stenosis or re-intervention (p = 0.009).
Figure 2  Surgical Techniques of Aortic Valve Repair

Techniques of surgical repair have evolved and always include an extensive debridement of the valve by resection of all nodular dysplasia, thinning of the leaflets, and recreation of an interleaflet triangle by resecting the fibrosis immobilizing the commissure. Incised portions of atretic valves are now resuspended with pericardial patches to avoid regurgitation.
Aortic Valve Surgery - Survival

Figure 3  Kaplan-Meier Survival Curve of the Entire Population

Survival of the entire population undergoing an aortic valve procedure in the Royal Children’s Hospital between 1977 and 2009. Ten-year and 20-year survival were, respectively, 88% (95% CI: 81% to 93%) and 83% (95% CI: 71% to 90%).
SUMMARY

• Not clear that valvuloplasty better than heterograft valve replacement in terms of durability in older children....suitable tissue for repair lacking
• Thin, bicuspid valves with pure AR suitable for repair with good medium-term results
• Complex valve repairs offer limited durability …damage from previous intervention and need to augment leaflet tissue…need for growth limits annuloplasty/support options
• Mechanical valves offer durability but significant lifestyle limitations in young patients
• Externally-supported autograft may be best choice in older children/adolescents – awaits longer followup
AV Repair vs. Replacement

AV Repair: Continuing Hazard For Reop/Replacement From First Year
Ross Procedure: Low Hazard For Reop/Replacement Until 6-10 Years, Then Increasing Hazard
Mechanical AVR: Low Hazard For Reop/Replacement Until 20 Years Continuing Hazard For Anticoagulant-Related Complications