Balloon Valvuloplasty for Congenital Aortic Stenosis: Is it TTT of Choice?

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Disclosures

None related to this presentation
NB with AS - Case 1

- Newborn male
- 2.8 kg
- Severe AS
- Vmax = 5.3 m/sec; PSG=110; M=61
- Mod. LVH with good systolic function

Severity of AS

<table>
<thead>
<tr>
<th></th>
<th>ACC/AHA</th>
<th>ESC</th>
<th>ACC/AHA</th>
<th>ESC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart catheter peak-to-peak gradient</td>
<td>cw Doppler Vmax</td>
<td>Bernoulli peak instantaneous gradient</td>
<td>Bernoulli mean instantaneous gradient</td>
<td>Echo aortic valve area</td>
</tr>
<tr>
<td>Trivial</td>
<td>&lt; 30 mmHg</td>
<td>&lt; 3 m/s</td>
<td>&lt; 36 mmHg</td>
<td>&lt; 25 mmHg</td>
</tr>
<tr>
<td>Slight</td>
<td>30-50 mmHg</td>
<td>3-4 m/s</td>
<td>36-64 mmHg</td>
<td>25-50 mmHg</td>
</tr>
<tr>
<td>Moderate</td>
<td>&gt; 50 mmHg</td>
<td>&gt; 4 m/s</td>
<td>&gt; 64 mmHg</td>
<td>&gt; 50 mmHg</td>
</tr>
<tr>
<td>Severe</td>
<td></td>
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</tbody>
</table>
What is a critical cAS?

- **Reduced effective orifice area**, usually resulting from thickening and reduced motion of the valve leaflets; Incomplete opening of the semilunar valve or valve ring being small, or a combination;
- Critical AS-----lead to **decompensation of LV** if not treated.
- **Ductal dependent** ------the stenosis is so severe that additional perfusion of the circulation through an open ductus is necessary to sustain life.
**Therapy for cAS- Pre-balloon**

- If **HF** is present it is advisable to treat it before the valve procedure.
- With **prostaglandin** the systemic perfusion may be supported.
- If there is isolated impairment of LV pumping function with dilated ventricle and decrease CO, **inotropic support** and **loop** diuretics are indicated.

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**In-Utero AS**

- **In-Utero AS**
  - **Small LV**
  - **LV dysfunction**
  - **HLHS**
  - **ECF**
Predictors of Failure of Biventricular Repair by Multivariate Analysis

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>P</th>
<th>OR</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>All patients (n=72)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate/large VSD</td>
<td>0.001</td>
<td>22</td>
<td>(3.6, 136)</td>
</tr>
<tr>
<td>Unicommision AoV</td>
<td>0.006</td>
<td>16</td>
<td>(2.2, 112)</td>
</tr>
<tr>
<td>Smaller MV annulus</td>
<td>0.02</td>
<td>2.2*</td>
<td>(1.1, 4.5)</td>
</tr>
<tr>
<td>Smaller LVEDVM</td>
<td>0.03</td>
<td>1.9*</td>
<td>(1.1, 3.3)</td>
</tr>
</tbody>
</table>


Valve Morphology?

- Bicuspid
- Bicuspid with raphe
- Bicuspid with raphe + calcification, retraction

**TRICUSPID**
25%

**BICUSPID**
60%

**MONOCUSPID**
15%
Valve Morphology

**Excellent Surgical results and Tricuspid valve**

<table>
<thead>
<tr>
<th>Absence of reintervention in 10 years</th>
<th>Bicuspid</th>
<th>Tricuspid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bhabra M. Ann Thorac Surg 2003 (Birmingham)</td>
<td>33%</td>
<td>92% (p &lt; 0.01)</td>
</tr>
<tr>
<td>Hraska V. Ann Thorac Surg 2012 (St Augustine)</td>
<td>50%</td>
<td>90% (p = 0.03)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Absence of Replacement in 10 years</th>
<th>Bicuspid</th>
<th>Tricuspid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bhabra M. Ann Thorac Surg 2003 (Birmingham)</td>
<td>57%</td>
<td>100% (p &lt; 0.01)</td>
</tr>
<tr>
<td>Hraska V. Ann Thorac Surg 2012 (St Augustine)</td>
<td>60%</td>
<td>100% (p &lt; 0.01)</td>
</tr>
</tbody>
</table>

And Bicuspid better than Monocuspid

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Aortic Insufficiency

- Increased risk with larger balloon: annulus ratio
  (but Hamidi Manesh et al reports differently#)
- Increased risk with young age *
- Increased risk with bicuspid, asymmetrically thick valves
- Increased risk with time


Conclusion: This study did not show any significant relationship between the balloon to annulus ratio during interventional dilatation and the degree of aortic regurgitation following the procedure. (J Interven Cardiol 2013;26:200–207)
BAV in neonates and young infants- AUBMC Experience

- Retrospective review of medical records, cath lab & surgical data base and echocardiograms
- All patients treated by BAV are included
- 2005-2016
- Excluded: Sub or supra AS, ECMO, prenatal aortic valve modification, and patients intended for SV palliation at the time of the BAVP.
30 patients

- Mean Age: **74 days** (1 – 300 days);
- Isolated 63% (19 patients)
- M: F ratio is 4:1
- **LV dysfunction** was a common finding in (56%).
- Prostaglandin was used in 20% of patients prior to the procedure

### Baseline Characteristics

<table>
<thead>
<tr>
<th></th>
<th>Newborns (n=15)</th>
<th>Infants (n=15)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mean Body Weight</strong></td>
<td>3.6 kg (1.9-7)</td>
<td>5.3 kg (2.9-9.6)</td>
</tr>
<tr>
<td><strong>Mean Aortic Annulus Diameter</strong></td>
<td>6.4 mm (4.7-8)</td>
<td>8.2 mm (6-11)</td>
</tr>
<tr>
<td><strong>Aortic Valve Anatomy</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unicuspid</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Bicuspid</td>
<td>6 (40%)</td>
<td>6 (40%)</td>
</tr>
<tr>
<td>Tricuspid</td>
<td>9 (60%)</td>
<td>9 (60%)</td>
</tr>
</tbody>
</table>
Coarctation of COA: 7 (21%) (3 Surgery and 3 Balloon); 1 trivial COA
VSD: 2 (6%); muscular small; no intervention
MV regurgitation: 1 (3%); surgery at 4 years repair of cleft
Ebstein: 1 (3%); mild; also COA
Shone; MS, AS and CoA: 1 (3%); also COA;

IMMEDIATE RESULTS:
- Mortality 0% in Hospital
- Residual gradient > 35mmHg: 3.3% (1 pt.; 40 mmHg)

LATE RESULTS (Mean follow-up 7 yrs; Range: 0.5 – 11 yrs)
- Mortality 1 late death (at home age of 3 months)
- Aortic stenosis: Trace to mild (70%); Mild to Moderate (27%); At least Moderate (3%)
- Re-intervention: 13%
Gradient Reduction (mmHg)

![Bar chart showing gradient reduction before and after BAV]

- Pre-BAV: 78 mmHg
- Post-BAV: 21 mmHg

p < .001

AR and Valvuloplasty

![Bar chart showing number of patients with varying levels of AR]

- Pre-BAV: 30 patients
- Immediate Post-BAV: 13, 9, 7, 1 patients
- Late Post-BAV: 4, 5, 9, 11 patients

Legend:
- 0-tr
- 1+
- 2+
- 3+
Immediate vs Late Results

Optimal outcome: residual PSG less than or equal to 35 mmHg and trivial or no AR.
Adequate outcome: residual PSG less or equal to 35 mmHg, and mild AR.
Inadequate outcome: Residual PSG above 35 mmHg, or more than moderate AR

30 Patients

- 56% presented with LV dysfunction
- 20% required PGE1 infusion

- 1 (3%) died at age of 3 months at home; small LV
- 4 (13%) required intervention
- 30 Balloon valvuloplasty
- 11/29 (38%) inadequate result (AR moderate or more)
- Ross (2) Mechanical (1) after 10 years
- MVR and Konno (1)

11/29 (38%) inadequate result (AR moderate or more)
Balloon vs. Surgery Pendulum Swings?

- (AS) accounts for 2/3 of the LVOTO lesions. (SAV) was the standard therapeutic intervention.

- In 1984 Lababidi et al. described (BAV),

- At most centers, BAV has become the standard of care for infants and children with AS requiring intervention.

- Indeed, a recent survey of 13 unselected North American pediatric cardiology interventional units when asked how infants would be typically managed, 77% reported almost always a BAV; and 23% a variable approach with the type of intervention decided upon on a case-by-case basis.

Aortic Valve and Surgery

Newborns: 30-days Mortality

Procedure and Hospital Mortality after 2010
Surgery vs. Balloon: 4-9%
Newborns: 10-year survival WITHOUT re-intervention

Overall 2368 patients from 20 studies were included in the analysis, including 1835 (77%) in the BAV group and 533 (23%) in the SAV group.
no difference in long-term survival or freedom from aortic valve replacement but significantly more re-intervention in the BAV group

Garick D. Hill et al. J Am Heart Assoc 2016;5:e003931
Figure 1: Overall survival stratifies by strategy.

Figure 2: Actuarial (Kaplan-Meier) freedom from any reintervention.

Figure 4: Actuarial (Kaplan-Meier) freedom from Ross procedure.
Of the 1,026 isolated BAV procedures captured in IMPACT, 2.4% of patients did not survive to hospital discharge.

Of the 1,026 isolated BAV procedures captured in IMPACT, 718 (70%) were “successful.”

BAV is an effective treatment for congenital AS with low rates of mortality and adverse events.
Conclusions

- BAV is a safe and effective modality to treat cAS in newborns and young infants, even in the presence of additional anomalies (i.e., CoA).
- Early and late mortality rates are low.
- AR remains a major concern and may increase with time and may require surgical intervention.
- Repeat BAV successful for recurrent AS.

Conclusions

- If the structures of the left side of the heart are severely hypoplastic/valve ring is too small or in cases of ECF, IV repair may be necessary.
- Balloon versus surgery is center specific and randomized data are needed to assess the superiority of a procedure over another.
- No clear data documenting the superiority of BAV vs. SAV; at our Center BAV is TTT of Choice.