Supravalvar Aortic Stenosis in Infants

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No disclosures
Supravalvar Aortic Stenosis

- Elastin arteriopathy
- 7q11:23 deletion
- Patient types
  - Williams-Beuren
  - Familial
  - Sporadic
# Age and Early Outcome

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>N</th>
<th>Age*</th>
<th>Deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kaushal</td>
<td>2010</td>
<td>20</td>
<td>3.6</td>
<td>10 mo., Diffuse type, Severe PA disease</td>
</tr>
<tr>
<td>Scott</td>
<td>2009</td>
<td>25</td>
<td>4.7</td>
<td>14 mo., Diffuse type, Cath lab arrest</td>
</tr>
<tr>
<td>Metton</td>
<td>2009</td>
<td>34</td>
<td>5.5</td>
<td>3 mo., Diffuse type, Left coronary obstruction</td>
</tr>
<tr>
<td>Hickey</td>
<td>2008</td>
<td>47</td>
<td>4.3</td>
<td>2 deaths, ages not specified</td>
</tr>
<tr>
<td>Brown</td>
<td>2002</td>
<td>101</td>
<td>6.1</td>
<td>1 death, ? age not specified</td>
</tr>
<tr>
<td>McElhinney</td>
<td>2000</td>
<td>36</td>
<td>4.0</td>
<td>24 mo, Diffuse type, Severe PA disease</td>
</tr>
</tbody>
</table>

*mean or median
SAVS in Infants

- Is younger age a risk factor?

- Complicating features are more prevalent in younger patients:
  - Diffuse disease
  - Bi-ventricular obstruction
  - Coronary artery obstruction
“Congenital Supravalvular Aortic Stenosis and Sudden Death Associated with Anesthesia: What’s the Mystery?”

- Review of all reported cases:
  - Bi-ventricular obstruction, or coronary obstruction
  - Most patients were young
    - 7 of 15 < 1 yr.
    - 10 of 15 < 2 yr.

Diffuse SVAS

Stamm, *JTCVS* 1999

Brown, *EJCTS* 2002
Diffuse SVAS Prevalent in Infants

  – 28% (7/25) overall had diffuse disease
  – 75% (6/8) <10 kg. had diffuse disease
    – Only 1 patient >10 kg. had diffuse disease

  – 60% (6/10) infants operated for SVAS had diffuse disease

  – 5 of 6 patients w/ diffuse disease were infants
Bi-ventricular Obstruction

SVPS

Pulmonary arteriogram

SVAS

Levophase
Bi-ventricular Obstruction

- **Stamm, et al. JTCVS 2000**
  - SVAS + right heart obstruction referred at younger age
  - ↑ RV pressure :↓ age

  - All SVAS+SVPS were infants, (n=5)
Coronary Involvement
SVAS with Coronary Obstruction

N=9

Type I
Ostial Narrowing
n=5

Type II
Cusp-Ridge Fusion
n=2

Type III
Fusiform Narrowing
n=2

7 patch angioplasties

*5 of 9 ≤ 2 yrs.

Thistlethwaite, et al. JTCVS 2000
Repair of SVAS

McGoon

1 sinus

Doty

2 sinus
3 Sinus Repair

surgeon
## 3-Sinus Repairs

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Total</th>
<th>Types (Brom, Meyers, Chard, Other)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kaushal</td>
<td>2010</td>
<td>8</td>
<td>8 Brom</td>
</tr>
<tr>
<td>Scott</td>
<td>2010</td>
<td>15</td>
<td>13 Meyers, 1 Brom, 1 Other</td>
</tr>
<tr>
<td>Metton</td>
<td>2009</td>
<td>23</td>
<td>23 Brom</td>
</tr>
<tr>
<td>Cruz-Castaneda</td>
<td>2009</td>
<td>9</td>
<td>9 Brom</td>
</tr>
<tr>
<td>Kocyildrin</td>
<td>2009</td>
<td>11</td>
<td>11 Brom</td>
</tr>
<tr>
<td>Kang</td>
<td>2001</td>
<td>7</td>
<td>4 Chard, 3 Meyers</td>
</tr>
<tr>
<td>McElhinney</td>
<td>2007</td>
<td>7</td>
<td>7 Meyers/Chard</td>
</tr>
<tr>
<td>Stamm</td>
<td>1999</td>
<td>6</td>
<td>4 Meyers, 2 Brom</td>
</tr>
<tr>
<td>HazeKamp</td>
<td>1999</td>
<td>13</td>
<td>13 Brom</td>
</tr>
<tr>
<td>Meyers</td>
<td>1993</td>
<td>6</td>
<td>2 Brom, 4 Meyers</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td></td>
<td><strong>105</strong></td>
<td><strong>69 Brom, 24 Meyers, 4 Chard, 1 Other</strong></td>
</tr>
</tbody>
</table>
Brom Repair for Diffuse SVAS
Brom Repair

ST junction is 7mm
Need ~ 14mm
Circumference = $\pi \times$ diameter
$\pi$ approx = 3
Need to add $(14-7) \times 3 = 21$ mm
3 patches →
Each patch ~ $(14-7)$ = 7 mm
Discrete SVAS

Diastole

Systole