The Surgical Management of the Bicuspid Aortic Valve in Children

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The Surgical Management of the Bicuspid Aortic Valve in Children

- Critical aortic valve stenosis in neonates
  - Univentricular vs. Biventricular repair
  - Balloon valvuloplasty vs. surgical valvotomy
- Aortic valve repair
- Aortic valve replacement
Critical Aortic Valve Stenosis in Neonates

- **BAS** associated with a spectrum of disease:

  - Aortic Valve Stenosis
  - Borderline LV
  - Hypoplastic LV
  - Biventricular
  - Univentricular

Risk Stratification
- Rhodes Score
- Colan Score
- (UVR-SA Value)

- AV Dimension
- Aortic Dimension
- LVOT Dimension
- MV Dimension
- EDFE
- Tricuspid Regurgitation
- Left Ventricular Length
Critical Aortic Valve Stenosis in Neonates

- Improving results of Norwood/Sano Procedures
- Increased mortality associated with “discordant decisions” (biventricular vs. univentricular repair)
- Influence of Ross Konno
- Considers the early hazard phase only
- Cross-over policy (initial balloon dilatation)
Critical LVOT obstruction: The disproportionate impact of biventricular repair in borderline cases

Balloon valvuloplasty versus surgery

- Risks of mortality and re-operation on the aortic valve were similar up to 5 years


- Some patients with borderline LV would now undergo single ventricle repair
- Surgical techniques were limited
Current Surgical Techniques

- Commissurotomy
- Resection of dysplastic myxoid nodules
- Thinning of leaflets
- Re-suspension of leaflets

Y d’Udekel et al, 2011
Surgical Valvotomy

V Hraska et al, 2006
Surgical Valvotomy

V Hraska et al, 2006
Surgical Valvotomy

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Surgical Valvotomy

V Hraska et al, 2006
### V Hraska et al, 2006

<table>
<thead>
<tr>
<th>Table 1. Balloon valvotomy</th>
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<tr>
<td><strong>Place of study</strong></td>
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<tr>
<td>Children’s Hospital Boston, USA [13]</td>
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<td>Children’s University Hospital, Zurich, Switzerland [11]</td>
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<th>Table 2. Open valvotomy</th>
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<td><strong>Place of study</strong></td>
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<tr>
<td>Department of Cardiac Surgery Southampton, UK [17]</td>
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<tr>
<td>German Pediatric Heart Center, Sankt Augustin, Germany [18]</td>
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Tricuspidisation with cusp extension

Tricuspidisation with cusp extension


Valve Replacement in Children

- Mechanical
- Bioprosthesis (Xenograft)
- Homograft
- Autograft (Ross Procedure)
Mechanical Valves

- High peri-operative mortality in children under 5 years of age.
- Constant phase of mortality associated with thrombo-embolic and anticoagulation related events (0.3 per 100 patient years).
- Better long term survival after mechanical AVR in adolescents than in adults.
Bioprosthesis

- Inverse relationship between age and rate of degeneration.

- 50% of patients will require re-operation within 8 years.
Homografts

- Rate of degeneration similar to bioprosthesis.
- In most cases must be inserted as a root replacement.
- Re-operation can be extremely difficult due to calcification.
Ross procedure

- In theory a permanent substitute for the aortic valve with potential for growth:
  - Degeneration of and need for replacement of the pulmonary homograft.
  - Inappropriate increase in size of the aortic root associated with neo-aortic regurgitation.
  - Estimated that 25% of the cases will require re-intervention on the aortic root or valve at 20 years.
Dilatation of the aortic root after the Ross operation modifications

- Root replacement vs. the inclusion technique.
- Technique of root replacement.
- Age at operation.
Ross Procedure

Y d’Udelem et al, 2011
The Ross Procedure

Technical Considerations:

1. Trim excess muscle from the autograft.
2. Correct annular mismatch and place pulmonary annulus inside the aortic annulus.
3. Aortic anastomosis just above the sinotubular junction.
4. Tailor or replace ascending aorta.
5. Control post-operative blood pressure.
The Ross Procedure

Ismail El-Hamamsy et al

M. Mostafa Mokhles, et al
The Ross Procedure
1999-2010

- 1\text{st} OPERATION = 50
- 2\text{nd} OPERATION = 17
- 3\text{rd} OPERATION = 4
The Ross procedure

Previous Procedures:

- Valvotomy = 11
- Subaortic Resection = 9
- Homograft Root Replacement = 4
- VSD Closure = 2
Cumulative survival free of reoperation (%)

Time (years)

Number at risk

Cumulative survival free of reoperation (%)

94.6

0 2 4 6 8 10

0 2 4 6 8 10
Ross Autograft and Pulmonary Homograft 10 years post-op

Aortic Root 41 mm at sinus level in systole
Influence of age on aortic dilatation after the Ross operation

- Aortic and pulmonary roots and semilunar leaflets have a similar structure during foetal life.

- Haemodynamic changes after birth initiate a re-modelling process which creates different vessel wall characteristics.
Influence of age on aortic dilatation after the Ross operation

- Remodelling capacity progressively decreases during childhood (progressively decreasing number of immature cells).

- In young age groups aortic and pulmonary roots are more alike with greater numbers of immature cells and higher content of fibrin, elastin and collagen fibres.
Freedom from aortic valve/root reoperation per age group

Freedom from RVOT reoperation per age group

M.L. Rito et al, 2013
M.L. Rito et al., 2013
Bicuspid Aortic Valve (Neonate)

- Biventricular Repair
  - Balloon dilatation
    - Success
  - Aortic valvotomy
    - Failure
- Univentricular Repair
  - Ross operation
Bicuspid Aortic Valve (Older Children)

Aortic Stenosis

Aortic Regurgitation

Repair (Valvulotomy /Valvuloplasty)

Success

Failure

Re-repair

Ross