Management of Infants with Coarctation and Ventricular Septal Defect

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Introduction

- The presentation in infancy of coarctation with an associated hemodynamically significant ventricular septal defect (VSD) is common.
- The optimal surgical strategy for these children is slightly controversial.

Two-stage Repair

- Initial repair of the coarctation through a left thoracotomy with/without pulmonary artery banding
- Then followed with closure of the VSD

Advantages:
- Avoidance of cardiopulmonary bypass and circulatory arrest in infancy
- VSD is closed when the child is older and bigger, arguably a simpler cardiac procedure than neonatal VSD closure

Disadvantages:
- Requires 2 operations with the infant only palliated between the 2 stages
- If a pulmonary artery band is placed, there are the risks: distortion of the branch pulmonary arteries, deformation of the pulmonary valve itself
- If there is associated proximal aortic arch hypoplasia, it is difficult to address satisfactorily through a thoracotomy incision

Conte and colleagues from Marie-Lannelongue Hospital in Paris (1992)
- 102 patient with CoA + VSD
  - 97 pt: end-to-end coarctation repair through a thoracotomy with 48 pt: placement of a pulmonary artery band
  - All of the patients with multiple VSDs, but only 31% of the neonates with single VSDs, had concomitant pulmonary artery banding
  - 10-year actuarial survival rate: 94; 5%
  - Recoarctation rate: 13%
- Importantly, 30% of these patients had spontaneous closure of the VSD
- Authors recommend: limitation of pulmonary banding to those children with multiple VSDs.

Isomatsu and associated in Tokyo (2001)
- 79 infants < 3 ms of age with CoA+VSD
- All 2 stage:
  1) Subclavian flap repair of the coarctation with a concomitant pulmonary artery band
  2) The VSD was closed after an interval of 10.4 ; 9.6 ms with removal of the pulmonary artery band
- Early mortality rate: 2.5%
- 10-year actuarial survival rate: 92.3%
- Recoarctation rate at 10 years: 9.6%
Bonnet and colleagues from Paris (1999)
- 11 infants < 30 days of age with CoA + medium-sized VSD (4~7 mm, mean: 5 ± 0.7 mm)
- End-to-end repair of the CoA and placed an absorbable pulmonary artery band with polydioxanone:
  - No early or late deaths
  - Only 1 recoarctation
  - 4 patients: complete spontaneous closure of the VSD, 6 had partial closure with no significant residual left-to-right shunt
  - The pulmonary artery band: all completely resorbed by 6 months without any residual PA stenosis or distortion

Brouwer and colleagues from The Netherlands (1996)
- 80 infants < 3 months of age with coarctation and VSD:
  - 64 infants (80%): 2-stage repair, only 10 had pulmonary artery banding
  - 16 infants (20%) had a 1-stage repair using hypothermic circulatory arrest for the coarctation repair
  - Freedom from recoarctation rate at 5 years:
    - In the 2-stage group was 91.3%
    - In the 1-stage group of only 60% (P=0.018)

Alsoufi and colleagues from the Hospital for Sick Children in Toronto:
- 141 neonates with coarctation and a VSD:
  - 2-stage strategy (n = 89) vs 1-stage approach (n = 52)
    - 5-year survival rate (91.3% vs 88.7%): similar
    - 12-year freedom from echocardiographic arch obstruction rates (80.8% vs 73.4%): similar
  - Only 42 (47%) of the 2-stage patients required subsequent VSD closure

Single-Stage Repair (One Incision)
- Advantages:
  - Complete repair of all the cardiac defects at 1 operation without interim palliation
- Disadvantages:
  - The use of circulatory arrest or regional cerebral perfusion

Sandhu and colleagues from the University of Michigan:
- 60 neonates < 28 days of age
- 19 neonates had aortic arch obstruction and VSD:
  - Operative mortality rate: 5.3%
  - 1 pts: early death
  - 10 pts: a recoarctation rate of 11%

Gaynor and colleagues from the Children’s Hospital of Philadelphia (2000):
- 25 infants < 3 months of age with coarctation and VSD
- Repaired the coarctation and arch:
  - Primarily (n = 4) or with a patch (n = 21)
  - Average circulatory arrest time: 38 ± 12 mins.
  - 4% early mortality rate with no late deaths
Ishino and colleagues from Okayama, Japan (2000)
- 11 patients: 1-stage technique of coarctation repair through a sternotomy
- They used regional cerebral perfusion:
  - Direct ascending aortic cannulation
  - Perfusion through a polytetrafluoroethylene graft sewn to the innominate artery
  - Allowed both myocardial and cerebral perfusion during the coarctation repair
- VSD was then closed with standard perfusion techniques and cardiopulmonary arrest

Single-Stage Repair (Two Incisions)
- Repair of the coarctation through a left thoracotomy followed by VSD closure through a sternotomy under the same anesthetic
- Potential advantages:
  - Repair of all the defects in infancy, avoiding palliation
- Disadvantages:
  - The need for circulatory arrest or regional cerebral perfusion for the coarctation repair

- 36 consecutive, nonrandomized infants (age <90 days)
- 11 pt: coarctation repair; pulmonary artery banding followed by VSD closure with 2 separate operations (2 stage)
- 10 pt: coarctation repair and VSD closure on cardiopulmonary bypass (1 stage, 1 incision)
- 15 pt: coarctation repair without cardiopulmonary bypass through a thoracotomy followed by VSD closure during the same operation (1 stage, 2 incisions)

Outcome
- No deaths in any of the groups
- Only 1 episode of significant recoarctation in a patient in the 1-stage, 1-incision group (who was successfully balloon dilated in the cardiac catheterization laboratory 3 months postoperatively.)

Table 1. Patient Characteristics at the Time of Index Operation

<table>
<thead>
<tr>
<th></th>
<th>Two Stage</th>
<th>One Stage</th>
<th>One Stage, Two Incisions No. = 100</th>
<th>No. (Percent)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (increase y)</td>
<td>0.045±0.009</td>
<td>0.04±0.009</td>
<td>0.04±0.009</td>
<td>0.05±0.009</td>
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<tr>
<td>Weight (kg)</td>
<td>5.1±0.9</td>
<td>5.1±0.9</td>
<td>4.9±0.8</td>
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<td>BSA (m²)</td>
<td>0.9±0.08</td>
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<td>Myocardial arrest</td>
<td>0.3±0.5</td>
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<tr>
<td>Perfusion time (min)</td>
<td>4.5±2.3</td>
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<td>Surplus with Z score</td>
<td>1.2±0.8</td>
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Table 2. Operative Results

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<td>Cardiac catheterization, mean time (min)</td>
<td>75±1.9</td>
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<td>Cardiopulmonary bypass time (min)</td>
<td>30±1.5</td>
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<td>Definitive repair, mean time (min)</td>
<td>75±1.9</td>
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<td>Total postoperative ICU Stay (days)</td>
<td>10±2.3</td>
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<td>1-stage, 1-incision group in 1-stage, 1-incision group and 1-stage group</td>
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No statistically significant differences
Gaynor JW from Children’s Hospital of Philadelphia (2005):

- duration of deep hypothermic circulatory arrest was associated with postoperative seizures after neonatal heart surgery (a putative marker for brain injury.)

**Recommendation**

1. 1-stage, 2-incision approach:
   - for infants with coarctation of the aorta and a hemodynamically significant VSD

2. 2-stage approach:
   - for infant with coarctation and multiple VSDs or a Swiss cheese septum

3. 1-stage, 1-incision approach:
   - for the infant with associated significant hypoplasia of the proximal aortic arch (between the innominate artery and the left carotid artery)

Thanks for your attention!