Early repair of Complete Atrio-ventricular Canal Malformations

Presented by
Hesham Shawky
Professor of Cardiothoracic Surgery,
Faculty of Medicine, Cairo University
Complete A-V Canal Defect

CAVC’Ds are the commonest form of AVC which accounts for about 3% of all major congenital cardiac defects.

Introduction

Normal heart anatomical specimen cut in short axis at the base demonstrating where the AV junction has a figure-of-8 configuration.

AVSD heart where the aortic valve (instead of being wedged) between the AV valve annuli is anterior of the AV junction. (Moss and Adams' Heart Disease in Infants, Children, and Adolescents: 7th Edition, 2008, chapter 31)
Introduction

Right anterior oblique view with right Atrial and right ventricular free walls removed, demonstrating a large septal defect.

The cleft in the anterior leaflet of the left AV valve oriented toward the midportion of the ventricular septum (arrow) along the anterior-inferior rim of the septal defect.

Introduction


Goose-neck appearance in angiographic imaging.
Introduction

Echo: Long axis view showing the distance from the LV apex to the posterior left AV valve annulus 20% - 25% < the distance from the apex to the aortic annulus.


The ideal timing for repair of CAVC remains controversial. Some authors advocate primary repair at the age of six months or earlier.

Others suggest two-stage repair consisting of pulmonary artery banding as a first stage followed by complete repair thereafter.
Introduction

- CAVSDs are associated with high-flow systemic pressure in the pulmonary vasculature leading to fibrosis and intimal hyperplasia.

- This eventually leads to a reduction in the total cross-sectional area of the pulmonary vascular bed and development of pulmonary vascular obstructive disease (PVOD).

Introduction

- The pulmonary vascular resistance along with the amount of AV valve regurgitation determine the onset of symptoms.

- Early congestive heart failure develops within the first few weeks of life that might be not controlled with medical therapy alone. About half of these patients, if left untreated, will die within the first year of life, usually from heart failure or respiratory tract infections.
Two-stage Repair?

- Although the two stage repair was carried out to reduce in-hospital mortality and technical difficulty from operating on smaller hearts with delicate tissues, yet this approach turned out to bear all long-term complications of pulmonary artery banding.

PA Banding??

- Tight banding can cause severe pulmonary stenosis with myocardial hypertrophy, and loose banding can result in irreversible PVOD.
- Palliation with pulmonary artery banding is now seldom indicated and has been abandoned for a single-stage definitive surgical repair.
Why do we prefer early single stage repair?

Stellin G., Vidaa V.L., Milanesi O., Rizzolia G., Rubinoa M., Padalino M.A., Bonatoc R., Casarotto D.
Surgical treatment of complete A-V canal defects in children before 3 months of age.

- 119 consecutive patients underwent repair of CAVCD from January 1985 to March 2001. 58 patients (49%) underwent correction before 3 months of age (Group A), and 61 patients (51%) after 3 months (Group B).
**Early Repair of Complete Atrioventricular Septal Defect is Safe and Effective.** 

- The mean age was 2.14 months in group A (39.3%) who underwent repair on or before 3 months of age (26 patients) and 16.76 months in group B (59.6%) who underwent repair after 3 months of age (39 patients).

Mariko Kobayashi, Yukihiro Takahashi and Makoto Ando 
**Ideal timing of surgical repair of isolated complete atrioventricular septal defect** 

- The trend towards earlier intervention has decreased the incidence of pulmonary hypertensive crises.

- The early surgical intervention before the development of pulmonary vascular obstructive disease is the optimal approach.
Single stage repair?

- The chronic volume overload from left-to-right shunting increases common AV valve annular dilatation resulting in increased left AV valve regurgitation and promotes secondary pathological changes in the AV valve tissue including the cleft area, leading to difficulty in achieving good coaptation after repair.

Single stage repair?

- The hypothesis is that CAVSD should be repaired before the onset of irreversible pulmonary hypertension especially in the presence of Down syndrome. This is preferably scheduled before the sixth month of life especially with improvements in anesthetic, and intensive care as well as surgical techniques.
Aim of Work

The aim of this study was to evaluate early results of primary repair of complete AV canal malformations in the first 6 months of life.

Patients and Methods
Patients and Methods

- **Study Group**: 20 patients < 6 months of age with CAVC undergone primary total repair.
- **Control group**: 20 patients > the age of 6 months with CAVC undergone primary total repair.
- *In the period between November 2009 and August 2011, at Cairo university hospitals*

- **Inclusion Criteria**
  - Patients with isolated complete AV canal defects below & above 6 months of age.

- **Exclusion Criteria**
  - Cases associated with complex anomalies such as Tetrology of Fallot.
  - Redo cases or cases underwent previous pulmonary artery banding.
Patients and Methods

I-Preoperative Parameters:

- History taking
- Clinical examination
- Investigations:
  1. *Routine laboratory investigations.*
  2. *Radiological examination:* Chest X-ray
  3. *Echocardiography:* PAP was measured. Rastelli type was delineated. Cardiac valve functions were evaluated.
  4. *Cardiac catheterization & angiography:* if pulmonary artery pressure was systemic or near systemic.

II-Intraoperative Parameters:

- *Intra-operative time parameters:* Cross-clamp time, total bypass time and operative time.
- *Technique used for repair:* single-, double-, or modified single-patch.
- *Use of inotropes* after weaning from cardiopulmonary bypass.
- *Patient rhythm* on going off bypass and on discharge from the operating theatre.
Patients and Methods

Single-patch technique
Patients and Methods

Single-patch technique
Patients and Methods

Double-patch technique
Patients and Methods

Double-patch technique
Patients and Methods

Double-patch technique

Patients and Methods

Modified single-patch technique
Patients and Methods

Modified single-patch technique
Patients and Methods

Modified single-patch technique

- The main advantage of not using a VSD patch is that by lowering the level of the left AV valve implantation at the crest of the septum, the area of coaptation is increased resulting in better competence.
- Another advantage is that it simplifies the procedure reducing both ischemic and total pump times. Wilcox and colleagues reintroduced this method of repair for patients with small VSDs.


- Now progression is going from direct suture of the VSD component, to avoiding the Atrial patch as well, aiming at reducing operating time more and getting normal sized atria that were dilated preoperatively which may help in preventing the occurrence of postoperative arrhythmias, the so-called No-patch technique.

Nicholson and colleagues advocated that the VSD patch could be avoided in most cases of complete AV canal with moderate and large VSDs.

Patients and Methods

III-Postoperative parameters

- ICU course:
  1. Mechanical Ventilation time.
  2. Incidence of PH crisis.
  3. Inotropic support.

- Total ICU stay & Hospital stay.
- Morbidities (complications).
- In-hospital mortality.
## Results

### Preoperative Demographic data

<table>
<thead>
<tr>
<th></th>
<th>STUDY GROUP</th>
<th>CONTROL GROUP</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age at operation</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(months)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Range</td>
<td>4-5.5</td>
<td>8-36</td>
<td></td>
</tr>
<tr>
<td>Mean ±SD</td>
<td>5.3±0.49</td>
<td>12± 8.5</td>
<td></td>
</tr>
<tr>
<td><strong>Body weight (Kg)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Range</td>
<td>4-5.5</td>
<td>7-15</td>
<td></td>
</tr>
<tr>
<td>Mean ±SD</td>
<td>4.7 ± 0.45</td>
<td>9± 2.5</td>
<td></td>
</tr>
<tr>
<td><strong>Sex</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Females</td>
<td>11(55%)</td>
<td>12(60%)</td>
<td></td>
</tr>
<tr>
<td>Males</td>
<td>9(45%)</td>
<td>8(40%)</td>
<td></td>
</tr>
<tr>
<td><strong>Down syndrome</strong></td>
<td>18</td>
<td>17</td>
<td></td>
</tr>
</tbody>
</table>

Non significant
**Results**

➢ Pre-operative Echo-cardio graphic data

<table>
<thead>
<tr>
<th></th>
<th>STUDY GROUP</th>
<th>CONTROL GROUP</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Rastelli type</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>A</td>
<td>11 (55%)</td>
<td>13 (65%)</td>
<td>Non significant</td>
</tr>
<tr>
<td>B</td>
<td>2 (10%)</td>
<td>1 (5%)</td>
<td></td>
</tr>
<tr>
<td>C</td>
<td>7 (35%)</td>
<td>6 (30%)</td>
<td></td>
</tr>
<tr>
<td><strong>Mean PAP</strong></td>
<td>60± 9 mmHg</td>
<td>65± 9 mmHg</td>
<td>Non significant</td>
</tr>
</tbody>
</table>

**Intraoperative Results**

➢ Intraoperative Time Results

<table>
<thead>
<tr>
<th></th>
<th>STUDY GROUP</th>
<th>CONTROL GROUP</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Total operative time</strong></td>
<td>182.5± 8min.</td>
<td>190± 8.5min.</td>
<td>Non significant</td>
</tr>
<tr>
<td><strong>Cardiopulmonary bypass time</strong></td>
<td>122 ±12min.</td>
<td>125 ±10min.</td>
<td>Non significant</td>
</tr>
<tr>
<td><strong>Cross-clamp time</strong></td>
<td>88± 7.5min.</td>
<td>82± 8min.</td>
<td>Non significant</td>
</tr>
</tbody>
</table>
Intraoperative Results

Study group

- Single-patch 20%
- Double patch 40%
- Modified single-patch 40%

Control group

- Single-patch 10%
- Double-patch 90%
# Intraoperative Results

## Inotropic Support

<table>
<thead>
<tr>
<th></th>
<th>Study Group</th>
<th>Control Group</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adrenaline infusion (100-200 ngm/kg/h)</td>
<td>18(90%) patients</td>
<td>19(95%) patients</td>
<td>Non significant</td>
</tr>
<tr>
<td>Milrinone infusion (0.5 μg/kg/min)</td>
<td>11 (55%) patients</td>
<td>15(75%) Patients</td>
<td>Non significant</td>
</tr>
</tbody>
</table>

## Patients Rhythm

<table>
<thead>
<tr>
<th></th>
<th>Study Group</th>
<th>Control Group</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sinus Rhythm</td>
<td>18(90%) Patients</td>
<td>19(95%) patients</td>
<td>Non significant</td>
</tr>
<tr>
<td>2nd H.B</td>
<td>2(10%) Patients Resolved within 24h</td>
<td>1(5%) Patients Resolved within 24h</td>
<td>Non significant</td>
</tr>
</tbody>
</table>
### Postoperative Results

#### ICU course:

<table>
<thead>
<tr>
<th></th>
<th>Study Group</th>
<th>Control Group</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mechanical ventilation</td>
<td>26 ± 10 hours</td>
<td>48 ± 20 hours</td>
<td>(.03)</td>
</tr>
<tr>
<td>Frequency of PH crises</td>
<td>4 (20%)</td>
<td>8 (40%)</td>
<td>(.01)</td>
</tr>
<tr>
<td>Inotropic support</td>
<td>48 ± 12 hours</td>
<td>72 ± 12 hours</td>
<td>significant</td>
</tr>
<tr>
<td>Chest tube drainage</td>
<td>60±29cc</td>
<td>80± 35 cc</td>
<td>Non significant</td>
</tr>
<tr>
<td>Re-exploration</td>
<td>-</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

#### Total ICU stay (Days)

<table>
<thead>
<tr>
<th></th>
<th>Study Group</th>
<th>Control Group</th>
<th>Significant</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean ± SD Range</td>
<td>3±1 days (2-6)Days</td>
<td>4±0.88 days (3-6)Days</td>
<td>significant</td>
</tr>
</tbody>
</table>

#### Total Hospital stay (Days)

<table>
<thead>
<tr>
<th></th>
<th>Study Group</th>
<th>Control Group</th>
<th>Significant</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean ± SD Range</td>
<td>15± 2.8 days</td>
<td>18.5± 2.3 days</td>
<td>Insignificant</td>
</tr>
</tbody>
</table>
Results
Morbidity & Mortality

<table>
<thead>
<tr>
<th></th>
<th>Study Group</th>
<th>Control Group</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Morbidity</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Superficial wound infection</td>
<td>3 (15%) patients</td>
<td>4 (20%) patients</td>
<td>Non significant</td>
</tr>
<tr>
<td>Pericardial collection</td>
<td>1 (5%) patient</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td><strong>Mortality</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>No Mortality</td>
<td>1 (5%) Patient due to postoperative PH crisis</td>
<td>Non significant</td>
</tr>
</tbody>
</table>

Conclusion

- The current risk factors for repair of CAVSD in patients younger than 6 months and in those older than 6 months are equal.
- No difference in operative mortality and early morbidity between patients undergoing surgical repair of CAVSD at 6 months of age or younger.
Recommendations

- **Repair** should be done at any age.
- Elective repairs for CAVSD are best performed at age 4 to 6 months.

Thank you