Outcome of symptomatic partial atrioventricular septal defect requiring repair in the first year of life.

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27th EDFSC, Paris, 11-14 January 2017
No conflict of interest
The prevalence - 0.11 per 1000 live births = 2.1% of children with congenital heart disease

- Patients typically asymptomatic, repair electively after infancy

- A small group of patients develop refractory heart failure during the first months of life and require earlier repair
Partial atrioventricular septal defect requiring repair in the first year of life

<table>
<thead>
<tr>
<th></th>
<th>Repair</th>
<th>N</th>
<th>Median age at repair</th>
<th>Mortality</th>
<th>Reoperations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Manning, 1994</td>
<td>1984-1992</td>
<td>11</td>
<td>8 m</td>
<td>36%</td>
<td>5 of 8 (12 operations) (5 subAS, 3 LAVV, 3 COA, 1 pace)</td>
</tr>
<tr>
<td>Giamberti, 1996</td>
<td>1982-1995</td>
<td>24</td>
<td>5 m</td>
<td>29%</td>
<td>5 LAVV, 3 COA</td>
</tr>
</tbody>
</table>
Partial atrioventricular septal defect requiring repair in the first year of life

Aims of the study

1) Document the outcome of infants with symptomatic pAVSD requiring early repair

2) Identify risk factors for mortality and reoperation
Study design

The inclusion criteria:

1) pAVSD (septum primum defect with common AV junction and separate AV valve orifices, patients with restrictive interventricular communication included

2) refractory heart failure

3) biventricular repair performed in infancy

4) repair between 1st January 2000 and 31st December 2015
### Results

**N=51 patients**

<table>
<thead>
<tr>
<th>Syndrome/ Genetic abnormality</th>
<th>N=28 (55%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trisomy 21</td>
<td>18</td>
</tr>
<tr>
<td>Trisomy 47XXX</td>
<td>1</td>
</tr>
<tr>
<td>Trisomy 14</td>
<td>1</td>
</tr>
<tr>
<td>Noonan</td>
<td>2</td>
</tr>
<tr>
<td>VACTERL</td>
<td>1</td>
</tr>
<tr>
<td>Mosaic Turner</td>
<td>1</td>
</tr>
<tr>
<td>Chromosomal translocation 6/9</td>
<td>1</td>
</tr>
<tr>
<td>CHARGE</td>
<td>1</td>
</tr>
<tr>
<td>Smith-Lemli-Opitz</td>
<td>1</td>
</tr>
<tr>
<td>Velo-Cardiac-Facial</td>
<td>1</td>
</tr>
</tbody>
</table>

### Concomitant diagnoses

<table>
<thead>
<tr>
<th>Concomitant diagnoses</th>
<th>N</th>
</tr>
</thead>
<tbody>
<tr>
<td>Secundum ASD/PFO</td>
<td>11</td>
</tr>
<tr>
<td>Ventricular component (spontaneously closed/small shunt)</td>
<td>21</td>
</tr>
<tr>
<td>Smaller LV, LAVV annulus</td>
<td>1</td>
</tr>
<tr>
<td>LVOTO</td>
<td>1</td>
</tr>
<tr>
<td>LAI</td>
<td>3</td>
</tr>
<tr>
<td>CoA/hypoplastic AoA (discrete CoA, isolated hypoplastic AoA)</td>
<td>9</td>
</tr>
<tr>
<td>PDA</td>
<td>11</td>
</tr>
<tr>
<td>RPA stenosis</td>
<td>2</td>
</tr>
<tr>
<td>LSVC/CS</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary vein stenosis</td>
<td>1</td>
</tr>
<tr>
<td>TAPVC supracardiac</td>
<td>1</td>
</tr>
<tr>
<td>AVB III congenital</td>
<td>1</td>
</tr>
<tr>
<td>Mild pulmonary stenosis</td>
<td>1</td>
</tr>
</tbody>
</table>
**Previous interventions**

<table>
<thead>
<tr>
<th>Cardiac surgeries</th>
<th><strong>Aortic coarctation repair</strong> (N=7) - end to end anastomosis (N=4) - subclavian flap repair (N=3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Catheterisation</td>
<td>Balloon angioplasty - <strong>re-coarctation of aorta</strong> (N=1) - <strong>branch PA stenosis</strong> (N=1)</td>
</tr>
</tbody>
</table>
| Non-cardiac surgeries | Duodenal atresia (N=2)  
Oesophageal atresia with tracheo-oesophageal fistula (N=2)  
Craniofacial synostosis (N=1) |
Surgical details

<p>| | |</p>
<table>
<thead>
<tr>
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</tr>
</thead>
<tbody>
<tr>
<td>Age at repair, days</td>
<td>179 (0-357)</td>
</tr>
<tr>
<td>Weight at repair, kg</td>
<td>5.0 (2.8-9.2)</td>
</tr>
<tr>
<td>Total bypass time, min</td>
<td>79 (37-280)</td>
</tr>
<tr>
<td>Cross clamp, min</td>
<td>55 (17-188)</td>
</tr>
</tbody>
</table>

LAVV procedure

“Cleft” closure
N=47

No LAVV repair
N=3

Double orifice LAVV
N=2

Small LAVV annulus
N=1

LAVV replacement (16 mm Carbomedics)
N=1
Left AV valve anatomy

Favourable morphology
N=35
- Thin leaflets, well developed mural leaflet
- Usual arrangement of the PM

Unfavourable morphology
N=16
- Dysplastic leaflets  N=7
- Double orifice  N=3
- Hypoplastic LAVV annulus/mural leaflet  N=3
- Short/poorly defined chordae  N=2
- Severely deficient LAVV tissue  N=1
Development of severe postoperative LAVV regurgitation and/or stenosis

No severe LAVVR in the early postoperative period
Severe LAVVR developed in 10 pts and severe stenosis in 1 pt, median 24 days (2 days-5.1 years)
Reoperations  
N=14

- LAVV replacement  
  N=1
  - LAVV re-repair  
    N=2
    - Pacemaker insertion  
      N=2

- LAVV repair  
  N=10

- LVOTO relief  
  N=2
  - LVOTO relief  
    N=1

- Relief of PV stenosis  
  N=1
Left AV reoperations

**Anatomy**

- Usual anatomy: 2
- Small AV valve annulus: 1
- Dysplastic: 4
- Hypoplastic leaflets: 1
- Underdeveloped subvalvar apparatus: 2

**Procedures**

- Closure of residual cleft: 10
- Commissural plication: 3
- Splitting of PMs: 1
- Posterior annuloplasty: 1

**2nd reoperation**

- Mitral valve replacement: 2
- Residual cleft closures: 2

**LAVV regurgitation**

- Usual anatomy: 2
- Small AV valve annulus: 1
- Dysplastic: 4
- Hypoplastic leaflets: 1
- Underdeveloped subvalvar apparatus: 2

**LAVV stenosis**

- Usual anatomy: 1
- Mitral valve replacement: 1

**Mitral valve replacement**

- 2
Kaplan-Meier survival analysis

3 in-hospital deaths after primary repair (5.9 %)
1 late death for viral gastroenteritis
## Risk factors of LAVV reoperation or death

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Univariate Analysis</th>
<th></th>
<th>Multivariable Analysis</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>HR (95% CI)</td>
<td>P value</td>
<td>HR (95% CI)</td>
<td>P value</td>
</tr>
<tr>
<td>LAVV anatomy</td>
<td>5.84 (1.75-19.48)</td>
<td>&lt;0.01</td>
<td>5.02 (1.41-17.80)</td>
<td>0.01</td>
</tr>
<tr>
<td>LAVV regurgitation before repair</td>
<td>3.29 (0.96-11.30)</td>
<td>0.06</td>
<td>1.79 (0.49-6.54)</td>
<td>0.38</td>
</tr>
<tr>
<td>Sex</td>
<td>0.55 (0.17-1.72)</td>
<td>0.30</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Associated syndrome</td>
<td>0.90 (0.29-2.86)</td>
<td>0.86</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>
### Risk factors of LAVV reoperation or death after adjustment for centre

<table>
<thead>
<tr>
<th></th>
<th>Univariate Analysis</th>
<th>Multivariable Analysis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>HR (95% CI)</td>
<td>P value</td>
</tr>
<tr>
<td><strong>LAVV anatomy</strong></td>
<td>5.97 (1.80-19.87)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td><strong>LAVV regurgitation before repair</strong></td>
<td>3.42 (1.02-11.46)</td>
<td>0.05</td>
</tr>
<tr>
<td><strong>Sex</strong></td>
<td>0.68 (0.25-1.88)</td>
<td>0.46</td>
</tr>
<tr>
<td><strong>Syndrome</strong></td>
<td>0.80 (0.29-2.22)</td>
<td>0.67</td>
</tr>
</tbody>
</table>
Last follow-up

Median follow-up 3.8 years (0.1-11.4 years)

Before repair

Severe
9
4
LAVV reoperation
4 Deaths

Moderate
15
4

Mild
18
6
2
10
3
4

None
9
5

Last follow-up

2 Mechanical valves
2

All pts symptom-free
Normal LV function

Medications:

Warfarin 2
Sildenafil 2
ACE inhibitors 4
Diuretics 5
Conclusion

Patients with pAVSD requiring repair in infancy:

- Significant morbidity and mortality
- Trend from LVOTO towards LAVVR as reason for reoperation
- High reoperation rate in patients with severe LAVVR due to unfavourable LAVV anatomy

Thank you for your attention
Partial and Transitional Atrioventricular Septal Defect Outcomes

L. LuAnn Minich, MD, Andrew M. Atz, MD, Steven D. Colan, MD, Lynn A. Sleeper, ScD, Seema Mital, MD, James Jaggers, MD, Renee Margossian, MD, Ashwin Prakash, MD, Jennifer S. Li, MD, Meryl S. Cohen, MD, Ronald V. Lacro, MD, Gloria L. Klein, MS, RD, and John A. Hawkins, MD [on behalf of the Pediatric Heart Network Investigators]
University of Utah, Salt Lake City, Utah; Medical University of South Carolina, Charleston, South Carolina; New England Research Institutes, Watertown, Massachusetts; The Hospital for Sick Children, Toronto, Ontario, Canada; Duke University Medical Center, Durham, North Carolina; Children's Hospital Boston, Boston, Massachusetts; and Children's Hospital of Philadelphia, Philadelphia, Pennsylvania

Reoperations After Repair of Partial Atrioventricular Septal Defect: A 45-Year Single-Center Experience

John M. Stulak, MD, Harold M. Burkhart, MD, Joseph A. Dearani, MD, Frank Cetta, MD, Roxann D. Barnes, MD, Heidi M. Connolly, MD, and Hartzell V. Schaff, MD
Division of Cardiovascular Surgery, Mayo Clinic and Foundation, Rochester, Minnesota
Ann Thorac Surg 2010;89:1352–9

Population-Based Perspective of Long-Term Outcomes After Surgical Repair of Partial Atrioventricular Septal Defect

Karl F. Welke, MD, Cynthia D. Morris, PhD, MPH, Emily King, BS, Christopher Komanapalli, MD, Mark D. Reiffel, MD, and Ross M. Ungerleider, MD
Division of Cardiothoracic Surgery, Department of Medical Informatics and Clinical Epidemiology, and Division of Pediatric Cardiology, Oregon Health and Science University, Portland, Oregon
Ann Thorac Surg 2007;84:624–9

Left Atrioventricular Valve Regurgitation After Repair of Incomplete Atrioventricular Septal Defect

Toshifumi Murashita, MD, PhD, Takehiro Kubota, MD, Jun-ichi Oba, MD, PhD, Toshihide Aoki, MD, PhD, Jun Matano, MD, and Keishu Yasuda, MD, PhD
Department of Cardiovascular Surgery, Hokkaido University School of Medicine, Sapporo, Japan

Specific issues after surgical repair of partial atrioventricular septal defect: Actuarial survival, freedom from reoperation, fate of the left atrioventricular valve, prevalence of left ventricular outflow tract obstruction, and other events


Ujwal K. Chowdhury, MCh, Diplomate NB, Balam Airan, MCh, Amber Malhotra, MCh, Akshay K. Bisoi, MCh, Mani Kalaiavani, MSc (Biostatistics), Raghu M. Govindappa, MS, and Panangipalli Venugopal, MCh

Congenital Cardiology Solutions

OUTCOMES AFTER REPAIR OF PARTIAL AND TRANSITIONAL ATRIOVENTRICULAR SEPTAL DEFECTS
Poster Contributions
Poster Sessions, Expo North
Saturday, March 09, 2013, 10:00 a.m.-10:45 a.m.

Session Title: Congenital Cardiology Solutions: Surgical Outcomes
Abstract Category: 13. Congenital Cardiology Solutions: Pediatric
Presentation Number: 1110-122

Authors: Jessica Brown, Harold Burkhard, Adile Goodfle, Joseph Dearani, Sabrina Phillip, Benjamin Edlin, Frank Cetta, Mayo Clinic, Rochester, MN, USA
Kaplan-Meier analysis of LAVV reoperation or death-free survival
Deaths
One newborn died intraoperatively after unsuccessful weaning from cardiopulmonary bypass. This patient had severe LV outflow tract obstruction and underwent emergency surgery with LAVV replacement on his first day of life. The other 2 patients required reoperation on their LAVV during the same admission as they developed severe regurgitation soon after primary repair. They died 36 and 68 days after primary repair (13 and 40 days after reoperation) from septic shock and enterococcal peritonitis, respectively. The LAVV regurgitation just before death was moderate in 1 patient and severe in the other one. All 3 deceased patients had unfavourable anatomy.
<table>
<thead>
<tr>
<th>Patient</th>
<th>Main LA VV features at primary repair</th>
<th>Mechanism of the LA VV regurgitation/stenosis at 1st reoperation</th>
<th>Surgical techniques at 1st LA VV reoperation</th>
<th>2nd reoperation of the LA VV</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Thickened superior bridging leaflet, accessory tissue from the superior leaflet</td>
<td>Residual ‘cleft’ + leakage along inferior bridging leaflet</td>
<td>Closure of residual ‘cleft’ + Commissural plication</td>
<td>Posterior annuloplasty</td>
</tr>
<tr>
<td>2</td>
<td>Accessory tissue from the Inferior bridging leaflet on the left side, very poorly defined subvalvar apparatus</td>
<td>Residual ‘cleft’</td>
<td>Splitting of the PM</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Usual anatomy of the LA VV*</td>
<td>Residual ‘cleft’ with suture dehiscence</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Generalized very short chordae</td>
<td>Residual ‘cleft’ with suture dehiscence</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Hypoplasia of the bridging leaflets</td>
<td>Residual ‘cleft’ with suture dehiscence</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Usual anatomy of the LA VV*</td>
<td>Residual ‘cleft’ with suture dehiscence</td>
<td>Residual ‘cleft’ closure + Posterior annuloplasty</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>Usual anatomy of the LA VV*</td>
<td>Significant stenosis precluding repair</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Dysplastic valve</td>
<td>Residual ‘cleft’</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Very dysplastic valve leaflets with retraction and hypomobility, very poorly defined subvalvar apparatus</td>
<td>Residual ‘cleft’, tethering of the mural leaflet due to abnormal chordal attachments of the mural leaflet</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Usual anatomy of the LA VV: Rationale for the use of mitral valve repair in the patient 6.
<table>
<thead>
<tr>
<th>Study</th>
<th>N</th>
<th>Median age at repair</th>
<th>Survival rate</th>
<th>Reoperations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bowman, 2014</td>
<td>105</td>
<td>7.9y</td>
<td>at 1 year 97%</td>
<td>12% - most common LAVVR</td>
</tr>
</tbody>
</table>
| Minich, 2010          | 87  | 1.8y                 | 1 in-hospital death | no reop at 6 months  
20% had significant LAVVR |
| Minich, 2010          | 87  | 1.8y                 | 1 in-hospital death | no reop at 6 months  
20% had significant LAVVR |
| Welke, 2007           | 133 | 3.4y                 | at 30 days 95% at 10 years 87% | 11% for LAVV pathology |
| Chowdhury, 2009       | 132 | 54 m (2.5m- 43y)     | 84% at mean follow-up 107m | 5.8% - most common LAVVR + resid ASD |
| Stulak, 2010          |     |                      |               | 1962-2006 - 96 pts - LAVV repair/LAVV replacement -38/35 pts  
Subaortic stenosis relief- 22 pts |
The whole patient cohort
N=51

Favourable anatomy
N=35

Unfavourable anatomy
N=16

N=3 (9%)

1 death

N=8 (50%)

Reoperations of LAVV
N=11

1 death

2 deaths
(Unfavourable anatomy)