Repair of Parachute and Hammock Valves in Infants and Children and its Long-Term Outcome

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Background

- The **rarity** and **complexity** of parachute and hammock valves as well as their **occurrence in infants and children** has stimulated great interest and fascination in its surgical management.

Objective

- We aim to **report our entire experience** with valve-preserving repair techniques and the **long-term outcome** in parachute and hammock valves and their variations.
**Profile of 20 children with parachute and hammock valves**
January 1990-June 2014

<table>
<thead>
<tr>
<th></th>
<th>Parachute valve (n=12)</th>
<th>Hammock valve (n=8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median (range) age</td>
<td>2.5 years (2 months-13.0 years)</td>
<td>7 months (1 month-14.9 years)</td>
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<tr>
<td>Median (range) weight, kg</td>
<td>10 (range 2.8-37.4)</td>
<td>8 (3.0-42)</td>
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<td>Associated diseases</td>
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<tr>
<td>Patent foramen ovale</td>
<td>1</td>
<td>2</td>
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<tr>
<td>Patent ductus arteriosus</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td>1</td>
<td>2</td>
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<tr>
<td>Tricuspid insufficiency</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Previous surgeries</td>
<td></td>
<td></td>
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<tr>
<td>Patch closure of sinus venosus defect</td>
<td>1</td>
<td></td>
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<tr>
<td>Resection of patent ductus arteriosus</td>
<td>4</td>
<td></td>
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<tr>
<td>Resection of vascular ring</td>
<td>1</td>
<td></td>
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<tr>
<td>Closure of ventricular septal defect</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Severity of mitral insufficiency</td>
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<tr>
<td>III°</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>IV°</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>Severity of mitral Stenosis</td>
<td></td>
<td></td>
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<tr>
<td>moderate</td>
<td>7</td>
<td>2</td>
</tr>
<tr>
<td>severe</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>Ross/NYHA Functional Class</td>
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</tr>
<tr>
<td>3</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>7</td>
<td>7</td>
</tr>
</tbody>
</table>
Not included are parachute valves in Shone’s anomaly (series of 45 children published in 2013).

- Coarctation of aorta
- Supravalvar mitral ring
- Parachute mitral valve
- Subaortic stenosis
Indications and timing of surgery

- Neonates and infants with parachute and hammock mitral valves were considered for surgical intervention based on the severity of symptoms.

- Clinical status, presence of moderate to severe mitral insufficiency and stenosis, with echocardiographic confirmation of abnormal MV anatomy are considered surgical indications.
Surgical procedures
Mitral valve repair is performed through a **median sternotomy** (optimal in presence when associated congenital heart lesions are present).

Aortic and bicaval cannulation
Left atriotomy via a direct incision along the interatrial groove

Mitral valve is assessed with a forceful injection of saline with a bulb syringe through the valve.
Assessment of leaflet and subvalvar apparatus

Using a nerve hook, the posterior leaflet is drawn out near the anterolateral commissure and the opposing position of the anterior leaflet to determine if sufficient tissues along the coaptation plane are present and to inspect the subvalvar apparatus.

Measurement of the valve diameter with a Ziemer-Hetzer valve sizer, after surgical opening of the valve.
Spectrum of anomalies seen intraoperatively

normal mitral valve
parachute-like asymmetric valve

parachute valve
hammock valve
Repair techniques for parachute valve
A and B. Commissurotomy and fenestration:
The most appropriate site for leaflet-splitting incision is defined on both sides from the common papillary muscles towards the assumed trigones.

C and D. These incisions are extended into the body of the papillary muscle, which is split towards its base, assuring sufficient thickness of both new “papillary muscle heads.”

E. Appearance after commissurotomy, fenestration and papillary muscle splitting.

Parachute valve has the usual two MV leaflets and commissures, but all the chordae tendinae merged into only one major papillary muscle. It presented as a funnel-type structure with some distinct fibrous lines at the sites of commissural fusion. The valve naturally was deformed, the chordae were short and thick; these, coupled with their convergent papillary insertion, allowed restricted leaflet mobility, thus creating a stenotic MV since the leaflets were closely apposed, greatly reducing the effective mitral orifice area.
Repair techniques for parachute-like asymmetric valve
**Parachute-like asymmetric mitral valve**

- has a large anterior papillary muscle directly fused with the leaflets, absence of chordae and presence of only fenestrations.

- However, the posterior papillary muscle was very small with just few short chordae, causing an asymmetric location of the valve orifice (A).

- To increase leaflet mobility, bilateral commissurotomy was performed and the anterior papillary muscle was incised (B) and split to a length which allows greater coaptation of the anterior and posterior leaflet (C).
Operative and long-term outcome of repair of parachute valve

No patients were lost to follow-up. The end of the follow-up study was June 2014.
Repeat MV repair (other than previously discussed in late mortality) was performed 1 year postoperatively on a 14 month-old infant at the time of initial surgery. He is now on his 12th year follow-up.

A 23-month old child at the time of initial repair likewise underwent repeat repair 10 years postoperatively. This patient had been now on his 15-year follow-up without any untoward event.
Repair techniques for hammock valve
- **Hammock valve** was seen in this series as an enormously deformed valve, dysplastic, with shortened chordae directly inserted in a muscular mass of the posterior LV wall resulting in tethering of both leaflets (A).
- The valvar orifice is partially obstructed by intermixed chordae and abnormal papillary muscles, characteristically implanted underneath the posterior leaflet.
- The chordae tendinae of the anterior leaflet cross the orifice toward the posteriorly implanted papillary muscles, producing the hammock appearance.

A suitably thick part of the posterior left ventricular wall carrying the rudimentary chordae is carved off the wall (B).

Precautions must be observed to ensure that both the remaining LV wall and the "new papillary muscles" maintain sufficient muscle thickness to maintain their function (C).
Operative and long-term outcome of repair of hammock valve

No patients were lost to follow-up. The end of the follow-up study was June 2014.
Repeat mitral valve repair (other than those described in the section of mortality) was performed on a **4-month old infant** a month after the initial surgery. She has been having regular follow-ups for 10 years since then.

- A **9-month old infant** underwent repeat mitral valve repair 19 months after the initial surgery and having a regular follow-up for 7 years since that time.
- A **6-month old infant** underwent repeat repair 5 years postoperatively and is now on her 19th year follow-up.
Overall functional outcome of MV repair

Mitral valve function

There was a marked absence of MS (mean MV orifice area of 5.2 ± 0.8 cm² without mean resting end-diastolic pressure gradient) and MI after MV repair (p<0.001).

In the course of follow-up, 2 PV patients have developed significant MS (mean resting end-diastolic pressure gradient 5.7 ± 1.3 mm Hg warranting repeat intervention).

Change in functional class

There was a significant improvement in NYHA functional class postoperatively (p<0.001) sustained until the late follow-up period.

3 HV patients had progressive MI which was 40%-60% at the time of repeat repair.
Limitations of the study

- The retrospective nature of this study imposed several inherent limitations.

- This series of children have been treated in the span of 21 years.

- Surgical and perfusion techniques have been modified over time.

- Whether this could have influenced the results is beyond the scope of this retrospective analysis.
Conclusions

- In children with parachute and hammock valves, surgical repair offered a satisfactory functional outcome during the long-term follow-up.

- **Repeat** MV repair may be necessary during the course of follow-up.

- **Infants** have a greater risk for reoperation and mortality.
Thank you for your attention!