Determinants of pulmonary valve replacement indication in repaired Tetralogy of Fallot patients: a multicenter experience


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AEPC Meeting, Helsinki, May 23, 2014
NO CONFLICT OF INTEREST TO DECLARE
The surgical repair of Tetralogy of Fallot (ToF) leads to various degrees of pulmonary regurgitation (PR) according to the different surgical approaches (infundibular or transannular patch).

The long-term volume load of PR leads to progressive RV dilation and dysfunction and deterioration of LV function associated with adverse patient outcome (*).

*Geva T, JACC 2004
*Ghai A, JACC 2002
Background

Pulmonary valve replacement (PVR) has been shown to improve RV hemodynamics and patients’ symptoms.

At present, PVR is univocally indicated in rToF patients, when symptoms or decreased exercise tolerance are attributable to PR.

In rToF asymptomatic patients, PVR timing is only based on CMR data. Nonetheless, definition of the ideal RV size for PVR timing is not yet established.
Understand more in depth the relationship between RV dilatation and function and:

- Age
- Severity of PR
- Type of surgical repair: Transannular patch (TP) Infundibular patch (IP)

To identify which factors are involved in the progression of RV dilatation and consequential RV dysfunction in order to optimize the follow-up management strategy of these patients
Methods

We retrospectively reviewed CMR studies of rToF patients performed at several paediatric centers in Italy.

The following CLINICAL and CMR data were collected:

- age at repair
- type of correction
- era of surgery
- Pulmonary regurgitation (PR)
- RVEDVi and RVESVi (ml/m²)
- RVEF (%)
- LVEDVi and LVESVi (ml/m²)
- LVEF (%)
- Tricuspid regurgitation (TR)
- RVOT and pulmonary arteries anatomy
CMR indication for PVR in asymptomatic patients was defined in presence $\geq 2$ of the following criteria (*):

- $\text{RVEDVi} > 150 \text{ ml/m}^2$
- $\text{RVESVi} > 80 \text{ ml/m}^2$
- $\text{RVEF} < 47\%$
- $\text{LVEF} < 55\%$

However, due to variability in clinical workflows among centers, the RV size at which PVR was actually performed was different in each participating paediatric center.

(*) Tal Geva, Circulation 2013;128:1855-1857
R.M. Martinez, Cardiology in the Young 2013;23,921-932
Results

Tot. 495 PTS (2008 – 2013)

- Bambino Gesù Children’s Hospital, Rome
- IFC-CNR Institute, Massa-Pisa
- Pediatric Institute Giannina Gaslini, Genoa
- Serv Cardiologia pediatrica, Osp Regionale, Bolzano

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Results

Age at enrollment (CMR) [years]

- ALL PTS: 30 ± 13
- TP: 21 ± 11
- IP: 18 ± 9

Age at surgical repair [years]

- TP: 1.8 ± 3
- IP: 4.9 ± 7

383 total patients

112 patients with IP

TP vs IP: 2:1 ratio
### Results: CMR parameters

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Transannular patch (age 18±9 years)</th>
<th>Infundibular patch (age 30±13 years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RVEDVi (ml/m2)</td>
<td>149 ± 37</td>
<td>122 ± 43</td>
</tr>
<tr>
<td>RVESVi (ml/m2)</td>
<td>122 ± 43</td>
<td>72 ± 25</td>
</tr>
<tr>
<td>EFRV (%)</td>
<td>72 ± 25</td>
<td>59 ± 28</td>
</tr>
<tr>
<td>LVEDVi (ml/m2)</td>
<td>53 ± 7</td>
<td>53 ± 8</td>
</tr>
<tr>
<td>LVESVi (ml/m2)</td>
<td>81 ± 14</td>
<td>80 ± 17</td>
</tr>
<tr>
<td>EFLV (%)</td>
<td>81 ± 14</td>
<td>80 ± 17</td>
</tr>
<tr>
<td>PR (%)</td>
<td>34 ± 90</td>
<td>32 ± 10</td>
</tr>
<tr>
<td></td>
<td>59±7</td>
<td>61±7</td>
</tr>
<tr>
<td></td>
<td>61±7</td>
<td>24±18</td>
</tr>
</tbody>
</table>
Results CMR parameters for PVR indication

- RVEDVi > 150 ml/m$^2$
  - Transannular patch (age 19±9 years): 48%
  - Infundibular patch (age 28±11 years): 22%

- RVESVi > 80 ml/m$^2$
  - Transannular patch (age 19±10 years): 16%
  - Infundibular patch (age 29±12 years): 13%

- RVEF < 47%
Results – Correlation of RV growth

**TRANSSANNUULAR PATCH**

\[ P<0.02, \; r=0.166 \]

**INFUNDIBULAR PATCH**

\[ p=NS \]

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Results - Correlates of PVR indication in all population

The determinants of the need for PVR were:

- DEGREE of PR
- RV VOLUMES
- TYPE of SURGERY
- GENDER
## Results - Correlates of PVR indication in relation to the type of surgery

<table>
<thead>
<tr>
<th>TRANSANNULAR PATCH</th>
<th>INFUNDIBULAR PATCH</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Significantly correlated with:</strong></td>
<td><strong>Significantly correlated with:</strong></td>
</tr>
<tr>
<td>• The grade of pulmonary regurgitation ( p=0.008 )</td>
<td>• The grade of pulmonary regurgitation ( p=0.022 )</td>
</tr>
<tr>
<td>• Time elapsed from surgical repair to CMR examination ( p=0.017 )</td>
<td>• The grade of tricuspid regurgitation ( p=0.032 )</td>
</tr>
<tr>
<td><strong>Not correlated with:</strong></td>
<td><strong>Not correlated with:</strong></td>
</tr>
<tr>
<td>• Time elapsed from surgical repair to CMR examination</td>
<td>• Time elapsed from surgical repair to CMR examination</td>
</tr>
</tbody>
</table>
Conclusions

Many factors play a role in different ways in the development and progression of RV dilatation and dysfunction.

The type of surgical repair and severity of PR/TR are among the most crucial factors involved in RV dilation leading to the need for PVR.

The different types of corrective surgery may result in substantial differences in the follow-up management strategy of these patients.
Gain further insight on the impact of various factors related to RV dilatation and dysfunction may help personalize the follow-up of rToF patients and fine-tune PVR timing, with the ultimate result of reducing the adverse outcome associated with this disease.
Thank you very much for your kind attention
# Table on CMR parameters

<table>
<thead>
<tr>
<th></th>
<th>RVEDVi &gt; 150 ml/m²</th>
<th>RV EF &lt; 47%</th>
<th>RVESVi &gt; 80 ml/m²</th>
<th>LV EF &lt; 55%</th>
<th>RVEDVi / LVEDVi &gt; 2</th>
<th>RVESVi &gt; 80 ml/m² + RV EF &lt; 47%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>TP</strong></td>
<td>182</td>
<td>95</td>
<td>124</td>
<td>103</td>
<td>149</td>
<td>65</td>
</tr>
<tr>
<td><strong>IP</strong></td>
<td>25</td>
<td>23</td>
<td>19</td>
<td>18</td>
<td>27</td>
<td>16</td>
</tr>
<tr>
<td><strong>TP</strong></td>
<td>48%</td>
<td>25%</td>
<td>32%</td>
<td>27%</td>
<td>39%</td>
<td>17%</td>
</tr>
<tr>
<td><strong>IP</strong></td>
<td>22%</td>
<td>21%</td>
<td>17%</td>
<td>16%</td>
<td>24%</td>
<td>14%</td>
</tr>
</tbody>
</table>

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