Total Right Ventricular Exclusion Improves Left Ventricular Function in Patients With End-Stage Congestive Right Ventricular Failure

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Background—We developed a total right ventricular (RV) exclusion procedure for the treatment of isolated congestive RV failure. The objective of the present study was to elucidate the effects of a complete removal of RV volume overload (RVVO) on the surgically created single left ventricle (LV).

Methods and Results—Three adults (2 arrhythmogenic RV dysplasia, 1 Ebstein) and 5 children (all Ebstein) in NYHA class IV underwent the procedure. The RV free wall was resected from the heart, and the tricuspid orifice was closed. Pulmonary blood supply was obtained by a cavopulmonary connection in 6 patients and a systemic-pulmonary shunt in 2. The LV function was evaluated by 2-dimensional echocardiography 1 month after the surgery. All patients are alive. The paradoxical movement of the interventricular septum and geometry of the LV expressed by its eccentricity (2.1 to 1.2, \( P<0.01 \)) were normalized after the operation in all 8 patients. LV end-diastolic volumes (59% to 109% of normal value, \( P<0.01 \)), indexed maximal left atrial area (6.5 to 10.5 cm\(^2\)/m\(^2\), \( P<0.01 \)), LV ejection fraction (27% to 62%, \( P<0.01 \)), and cardiac index (2.1 to 3.3 L/min/m\(^2\), \( P<0.05 \)) all significantly increased.

Conclusion—Removal of the RVVO by means of the total RV exclusion procedure provides effective volume loading, restores a cylindrical shape, and improves contractile function of the LV, thus leading to increased systemic output.

(Circulation. 2003;108[suppl II]:II-226-II-229.)

Key Words: heart failure ■ ventricular function ■ echocardiography ■ cardiac volume

Right ventricular volume overload (RVVO) is often associated with tricuspid regurgitation\(^1\)\(^–\)\(^7\) or a left-to-right shunt at the atrial level.\(^8\)\(^–\)\(^11\) Long-lasting RVVO gradually induces right ventricular (RV) and right atrial dilatation, leading to congestive RV failure. RV pump dysfunction coupled with compression of the left heart by an enlarged right heart reduces an effective preload on the left ventricle (LV). In addition, a decrease in the interventricular pressure gradient during the diastolic phase causes flattening and leftward displacement of the interventricular septum (IVS).\(^2\)\(^,\)\(^3\)

Therefore, RVVO may reduce systemic output and impair contractile function of a potentially normal LV.

We developed a total RV exclusion procedure for the treatment of isolated end-stage RV failure in 1996.\(^12\) This procedure is composed of the resection of the RV free wall and a cavopulmonary connection or systemic-pulmonary shunt. The objective of the current study was to elucidate the effects of a complete removal of RVVO on the surgically created single LV utilizing 2-dimensional (2D) and Doppler echocardiography.

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Circulation is available at http://www.circulationaha.org

DOI: 10.1161/01.cir.0000087382.12277.49
TABLE 1. Patients' Profiles

<table>
<thead>
<tr>
<th>Age</th>
<th>Gender</th>
<th>Diagnosis</th>
<th>Weight (kg)</th>
<th>Concomitant Procedure</th>
<th>V&lt;sub&gt;ED&lt;/sub&gt; (mL)</th>
<th>LVEF (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>27 y</td>
<td>M</td>
<td>ARVD</td>
<td>70</td>
<td>TCPC (Extracardiac)</td>
<td>81</td>
<td>14</td>
</tr>
<tr>
<td>44 y</td>
<td>F</td>
<td>ARVD</td>
<td>52</td>
<td>TCPC (Extracardiac)</td>
<td>42</td>
<td>121</td>
</tr>
<tr>
<td>16 mo</td>
<td>F</td>
<td>Ebstein</td>
<td>7.6</td>
<td>TCPC (Lateral tunnel)</td>
<td>3</td>
<td>9</td>
</tr>
<tr>
<td>5 y</td>
<td>M</td>
<td>Ebstein</td>
<td>10</td>
<td>TCPC (Extracardiac)</td>
<td>63</td>
<td>75</td>
</tr>
<tr>
<td>5 mo</td>
<td>M</td>
<td>Ebstein</td>
<td>5.5</td>
<td>BDG, PV-plasty</td>
<td>4</td>
<td>11</td>
</tr>
<tr>
<td>37 d</td>
<td>M</td>
<td>Ebstein</td>
<td>2.7</td>
<td>PV-tomy, SPS</td>
<td>4</td>
<td>9</td>
</tr>
<tr>
<td>50 d</td>
<td>M</td>
<td>Ebstein</td>
<td>4.0</td>
<td>SPS</td>
<td>3</td>
<td>11</td>
</tr>
<tr>
<td>51 y</td>
<td>F</td>
<td>Ebstein</td>
<td>48</td>
<td>TCPC (Extracardiac)</td>
<td>26</td>
<td>65</td>
</tr>
</tbody>
</table>

ALARA, arrhythmogenic right ventricular dysplasia; BDG, bidirectional Glenn anastomosis; Ebstein, Ebstein’s anomaly; F, female; M, male; LVEF, left ventricular ejection fraction; Pre and Post, pre- and post-operative; PV, pulmonary valve; SPS, systemic-pulmonary shunt; TCPC, total cavopulmonary connection; V<sub>ED</sub>, left ventricular end-diastolic volume.

A total cavopulmonary connection (TCPC) was constructed in 3 adults and 2 children; a fenestration was made in two ARVD patients. A bidirectional Glenn anastomosis (BDG) was performed in a 5-month-old boy, and systemic-pulmonary shunt was placed in 2 infants younger than 3 months. The heart was controlled with a DDD pacemaker in 3 patients.

Pulsed Doppler and 2D

Echocardiographic Examination

The Pulsed Doppler and 2D echocardiographic evaluations were performed at a mean of approximately 30 days before and after the surgery, respectively. Examinations were performed from the standard left parasternal and apical windows while the subjects were in sinus rhythm or paced in the left lateral recumbent position. All measurements were performed by an experienced pediatrician (S.O.) or echo technician (Y.T.).

Because some of the preoperative LV geometry was not circular, especially during end-diastole, we used a modified Simpson’s rule to calculate the LV end-diastolic (V<sub>ED</sub>) and end-systolic (V<sub>ES</sub>) volumes as previously reported. See Figure 1. End-diastolic and end-systolic cross-sectional areas (cm<sup>2</sup>) in the short-axis view, both at the mitral annulus (MA<sub>ED</sub> and MA<sub>ES</sub>) and the mid-papillary (PA<sub>ED</sub> and PA<sub>ES</sub>) levels, and LV length (cm), end-diastolic (L<sub>ED</sub>) and end-systolic (L<sub>ES</sub>) in the long-axis view were measured. V<sub>ED</sub> (ml), V<sub>ES</sub> (ml), and ejection fraction (EF) (%) were calculated by the equations shown below. V<sub>ED</sub> was expressed as a percentage using normal values calculated from the body surface area (BSA) as reported by Nakanawa et al.<sup>4,15</sup>

\[
V_{ED}(ml) = L_{ED}/3 \times [(MA_{ED} + MA_{ES} + PA_{ED})/2 + PA_{ES}/3]
\]

\[
V_{ES}(ml) = L_{ED}/3 \times [(MA_{ED} + MA_{ES} + PA_{ES})/2 + PA_{ES}/3]
\]

\[
EF(%) = (V_{ED} - V_{ES}) \times V_{ED} \times 100
\]

LV eccentricity was determined by the method of Schrieber et al.<sup>4,15</sup> in the short axis view at the level of mitral chordal transition at end-diastole as the ratio (B/A) of the two minor axes, B and A, of the LV (Figure 1). Maximal left atrial cavity area was measured from the apical four-chamber image<sup>1,16</sup> and indexed using the BSA. In 5 patients (3 adults and 2 children), the diameter of the LV outflow tract was measured from the parasternal long axis view, and its velocity was recorded using pulsed Doppler. Stroke volume was calculated as the product of the time velocity integral and cross-sectional area calculated from the diameter. The cardiac output was calculated from the stroke volume multiplied by the heart rate and then indexed using the BSA (cardiac index). In 4 patients (3 adults and 1 child), the trans-mitral flow velocity was obtained from the apical 4-chamber view by placing the sample volume at the tips of the mitral leaflets. The peak early (E) and late (A) diastolic trans-mitral flow velocity and their ratio (E/A) as well as the deceleration time of E wave were measured.

Representative changes of the LV shape in the short axis view. LV eccentricity was determined in the short axis view at the level of mitral chordal transition at end-diastole as the ratio (B/A) of the two minor axes, B and A, of the LV as shown in the left image. The length of the minor-axis diameter from the LV endocardium of the mid IVS to that of the lateral free wall was defined as A. The length of the orthogonal minor-axis diameter between the endocardial interfaces of the anterior and inferior LV free walls was defined as B. The marked LV compression at end-diastole in the left image was improved and the LV shape at end-diastole was almost circular in the short axis view after surgery in the image second to the right. The LV shape is circular at end-systole both pre- and post-operatively. IVS, interventricular septum; LV, left ventricular.

[Preoperative] [Postoperative]
**Results**

All data were expressed as mean value±SD. Changes in all of the parameters from pre- to post-operative values were analyzed using a paired t-test. Results with a probability value of less than 0.05 were considered statistically significant.

<table>
<thead>
<tr>
<th>TABLE 2. Echocardiographic Findings (N=8)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Preoperative</strong></td>
</tr>
<tr>
<td>Heart rate (beats/minute)</td>
</tr>
<tr>
<td>VES (mL)</td>
</tr>
<tr>
<td>VES (% of normal value)</td>
</tr>
<tr>
<td>VED (mL)</td>
</tr>
<tr>
<td>LVEF (%)</td>
</tr>
<tr>
<td>Indexed left atrial area (cm²/m²)</td>
</tr>
<tr>
<td>LV eccentricity index</td>
</tr>
<tr>
<td>†Stroke index (mL)</td>
</tr>
<tr>
<td>†Cardiac index (L/min/m²)</td>
</tr>
<tr>
<td>§E (m/sec)</td>
</tr>
<tr>
<td>§A (m/sec)</td>
</tr>
<tr>
<td>§E/A</td>
</tr>
<tr>
<td>§Deceleration time (msec)</td>
</tr>
</tbody>
</table>

* P<0.05; † P<0.01 compared with preoperative values. § N=5; § N=4 because of data availability.

**Statistical Analysis**

All data were expressed as mean value±SD. Changes in all of the parameters from pre- to post-operative values were analyzed using a paired t-test. Results with a probability value of less than 0.05 were considered statistically significant.

**Discussion**

The total RV exclusion procedure, comprised of RV free wall muscle resection, significantly increased the volumes at end-diastole, restored a cylindrical shape, and improved contractile function of the surgically created single LV. The resultant increase in systemic output led to rapid improvements in the postoperative patients’ condition. The results of the present study suggest that the severe LV pump failure associated with RV congestion is usually reversible, even in adults with long-lasting RVVO, and that the total RV exclusion procedure hold promise as a useful therapeutic option for isolated congestive RV failure.

The possible mechanisms for these rapid and remarkable improvements still need to be discussed. First and most importantly, the normalization of LV loading conditions was demonstrated as increases in the maximal left atrial area (160% of the preoperative value) in our study, which represents a value greater than those seen in normal subjects. Phoon et al observed that the “hypoplastic” and deformed LV resulting from RVVO in the patients with total anomalous pulmonary venous connection enlarged to a much larger size than its preoperative predicted potential volume after the total correction, suggesting that this small LV was not simply because of compression but rather it was a result of underfilling. Our data of increased left atrial area greater than normal subjects imply that the LV was filled enough to expand after the complete removal of the RV free wall. Consequently, the volumes at end-diastole increased to 109% of normal value. There is no doubt that the main component of the mechanisms responsible for the improvements of systemic output was this normalization of the loading conditions at end-diastole. Secondly, another important mechanism is the normalization of the IVS movement. RVVO is known to depress the LVEF mainly as a result of the paradoxical movement of the IVS as reported by Louie et al. In their study, even the volume-replete LV in patients undergoing valvulotomy for isolated tricuspid valve endocarditis exhibited a small but significant depression in LVEF when compared with normal subjects. Although the IVS normalization alone cannot explain such improvements of LVEF (27 versus
in our patients, we must consider its small but significant contribution to the LVEF.

Although the quality and amount of data regarding the LV inflow analysis are not adequate to draw definite conclusions, this procedure might have brought additional advantages to the LV compliance as shown in the increase of deceleration time and color M-mode Doppler flow propagation velocity. Because the diastolic function is a very important component in the prognosis after TCPC, it will be necessary to follow up with these patients periodically for the long-term. So far, all these patients are doing well and are being followed up as outpatients.

There are several limitations in this study. First, because the number of patients for this procedure is small, we included patients with a broad spectrum of age into our analysis. An immature heart may respond in a different way when compared with a mature heart. Second, a systemic-pulmonary shunt yields a greater volume load on the LV than a cavopulmonary connection. In fact, the postoperative VED in the number of patients for this procedure is small, we included patients with a broad spectrum of age into our analysis. An immature heart may response in a different way when compared with a mature heart. Second, a systemic-pulmonary shunt yields a greater volume load on the LV than a cavopulmonary connection. In fact, the postoperative VED in the patients are doing well and are being followed up as outpatients.

In conclusion, removal of RVVO by means of the total RV exclusion procedure provides effective volume loading, restores a cylindrical shape, and improves systolic performance of the LV, thus leading to increased systemic output. The physiological restoration of LV size, geometry, and function leads us to believe that this procedure holds promise as a useful therapeutic option for isolated end-stage RV failure caused by RVVO.

Acknowledgments
We thank Mr. Michael Kopcak for his excellent editorial assistance in the preparation of this article.

References
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_Circulation_. 2003;108:II-226-II-229
doi: 10.1161/01.cir.0000087382.12277.49

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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