Successful Permanent Resynchronization for Failing Right Ventricle After Repair of Tetralogy of Fallot

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A fter repair of tetralogy of Fallot with significantly decreased exercise tolerance, a 17-year-old symptomatic boy underwent successful permanent resynchronization of a failing right ventricle (RV). The patient had surgical repair including pulmonary valvulotomy, infundibulectomy, and pericardial right ventricular outflow tract (RVOT) plasty at 1 year of age. At 3 years of age, he underwent resection of RVOT aneurysm, pulmonary valvuloplasty, and patch enlargement of the left pulmonary artery. Finally, a balloon pulmonary valvuloplasty of a mild residual pulmonary valve stenosis was performed at 17 years of age, resulting in a gradient reduction from 18 to 4 mm Hg and mild (grade I) pulmonary and tricuspid regurgitation. However, the patient continued to show significant RV dilation and systolic/diastolic dysfunction with signs of major RV dyssynchrony (right bundle-branch block; QRS duration of 200 ms and paradoxical interventricular septal motion with a late systolic right to left septal flash) by echocardiography and MRI. As a result of reports on successful acute subpulmonary RV resynchronization, the patient underwent resynchronization testing in the catheterization laboratory with the aim to identify the RV pacing site associated with maximum acute increase in RV dP/dt max from 432 to 624 mm Hg/s when pacing in complete fusion with spontaneous activation. One month later, an MRI-compatible permanent transvenous dual-chamber pacemaker was implanted, with ventricular pacing lead inserted as close as possible to the previously identified optimal site at the RV free wall (Figure 1), resulting in a Q-RV interval of 140 ms, proving late electric activation of this area. Atrio-ventricular delay was programmed to achieve complete fusion with spontaneous ventricular depolarization, as subsequently confirmed by 24-hour Holter ECG.

Six months later, a major decrease in RV size, along with improvement in RV as well as LV function, functional class, and exercise capacity were noted (Table, Figures 2 through 6, and Movies I and II in the online-only Data Supplement). We conclude that permanent resynchronization of a failing RV is an effective method to achieve hemodynamic improvement after repair of tetralogy of Fallot. Patients with right bundle-branch block and significant RV dilation and dysfunction might be considered for this procedure in isolation or along with other indicated surgical or transcatheter interventions. RV electromechanical dyssynchrony may be, in addition to volume overload, a major factor in RV failure development. The precise role and indications for this novel treatment option remain to be studied. This is, to our knowledge, the first report of successful permanent resynchronization of a failing subpulmonary RV in congenital heart disease.

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Disclosures
None.

References
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Figure 1. Chest x-ray film. Antero-posterior (A) and lateral (B) view of the pacing system with ventricular lead inserted to the site of late electric activation at the right ventricular free wall. CAUD indicates caudal view; LAO, left anterior oblique view; and RAO, right anterior oblique view. Arrows indicate the ventricular lead insertion site.

Table. Electrocardiographic, MRI, and Functional Parameters Before and 6 Months After RV Resynchronization

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<tr>
<th></th>
<th>RV</th>
<th>LV</th>
<th>VO₂max</th>
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<tbody>
<tr>
<td>QRS</td>
<td>ms</td>
<td>mL/m²</td>
<td>mL/kg/min</td>
</tr>
<tr>
<td>Before</td>
<td>200</td>
<td>212</td>
<td>172</td>
</tr>
<tr>
<td>After</td>
<td>140</td>
<td>141</td>
<td>87</td>
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BSA indicates body surface area; EDVi/ESVi, end-diastolic/end-systolic volume index; EF, ejection fraction; NYHA, New York Heart Association Class; LV, left ventricle; RV, right ventricle; and VO₂ max, maximum oxygen uptake during exercise stress testing.

Figure 2. Twelve-lead ECG. A. Before pacing: sinus rhythm, spontaneous ventricular activation with complete right bundle-branch block morphology. B. After resynchronization: major decrease in QRS complex duration during dual mode, dual chamber, dual sensing (DDD) pacing with an atrio-ventricular delay adjusted to achieve complete fusion with spontaneous ventricular depolarization. Leads aVR, aVL and aVF indicates augmented limb leads.
Figure 3. Echocardiography (speckle-tracking analysis). A, Before pacing: asynchronous right ventricular (RV) contraction; interventricular septum (yellow arrow) is the earliest contracting segment, whereas RV free wall (red arrow) is the latest. B, After resynchronization: synchronous RV contraction.
Figure 4. Echocardiography (tricuspid valve Doppler): increase in the right ventricular filling time. A, Before pacing. B, After resynchronization. CL indicates cycle length; and RVFP, right ventricular filling period.
Figure 5. Echocardiography (tricuspid valve Doppler): increase in right ventricular $dP/dt$ max.  
A, Before pacing.  
B, After resynchronization.

Figure 6. MRI (short axis image): decrease in right ventricular end-diastolic dimensions.  
A, Before pacing.  
B, After resynchronization.
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