

# Residual Restrictive Right Ventricular Physiology after One-and-a-Half Ventricular Repair Conversion in Pulmonary Atresia with Intact Ventricular Septum



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## INTRODUCTION

Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare congenital heart defect. Among various surgical procedures that can be performed in early childhood, our institutional strategy is to aim for biventricular repair (BVR) on the basis of evidence of potential right ventricular (RV) growth after adequate creation of RV and pulmonary artery continuity<sup>1-3</sup> to avert abnormal hemodynamics and late complications in univentricular repair. Despite the relatively good long-term survival in adult patients with PA-IVS who have undergone BVR,<sup>4</sup> one-and-a-half ventricular repair (1.5VR) conversion concomitant with pulmonary valve intervention is required in some adult patients undergoing BVR because of pulmonary regurgitation (PR) and elevated central venous pressure followed by end-organ damage and heart failure. Restrictive RV physiology is a unique feature in patients with PA-IVS after BVR<sup>5</sup> and can be assessed on pulsed Doppler echocardiography. Although restrictive RV physiology is a potential culprit of BVR failure,<sup>5</sup> changes in Doppler and other parameters after 1.5VR conversion from BVR remain unknown.

## CASE PRESENTATION

A 20-year-old woman with PA-IVS who had undergone BVR during infancy presented with progressive shortness of breath on exertion and pedal edema. She was diagnosed with PA-IVS at birth. Her tricuspid valve (TV) Z score from echocardiography was  $-6$ , RV end-diastolic volume (RVEDV) from cine angiography was 24% of the predicted normal value, and RVEDV indexed to body surface area (RVEDVi) was  $9.5 \text{ mL/m}^2$ . Balloon atrioseptostomy was performed at birth, followed by pulmonary valvotomy, left modified Blalock-Taussig shunt, and ligation of the patent ductus arteriosus at 2 weeks of age. At the age of 5 months, cardiac catheterization revealed valvular pulmonary stenosis with a peak pressure gradient

of 46 mm Hg and 30% of the predicted normal value of RVEDV (RVEDVi  $14 \text{ mL/m}^2$ ). Therefore, balloon pulmonary valvuloplasty was performed, with the expectation of RV growth by means of relief of the pulmonary stenosis. At the age of 1.7 years, the TV Z score from echocardiography was  $-1$ , RVEDV from cine angiography was 58% of the predicted normal value, and RVEDVi was  $30 \text{ mL/m}^2$ . Although the right ventricle was shown to be relatively small, it was considered to be large enough to proceed to BVR upon inspection of the right ventricle and TV at surgery. BVR including RV overhaul, RV outflow tract reconstruction with a Gore-Tex monocusp valve, semiclosure of the atrial septal defect, and take-down of a modified left modified Blalock-Taussig shunt were performed at the age of 1.8 years. After definitive surgery, she had been doing well without any symptoms of heart failure or arrhythmia, and RVEDV had enlarged to 73% of the normal predicted value (RVEDVi  $42 \text{ mL/m}^2$ ) at the age of 3 years.

The patient presented with signs of heart failure at the age of 20 years. Electrocardiography showed normal sinus rhythm with complete right bundle block (Figure 1). Cardiac magnetic resonance imaging revealed an enlarged right ventricle, enlarged right atrium, mild tricuspid regurgitation, and severe PR with a regurgitant fraction of 27% (Figure 2A). Echocardiography showed preserved left ventricular ejection fraction of 65%, normal inferior vena cava diameter of 13 mm with poor respiratory change of 38%, and end-diastolic forward flow in the pulmonary artery with a peak velocity of 44 cm/sec (Figure 2B; Videos 1 and 2), suggesting the presence of restrictive RV physiology.<sup>6</sup>

Cardiac catheterization showed an elevated right atrial (RA) pressure of 11 mm Hg. Liver ultrasound revealed severely dilated hepatic veins, an irregular surface, and a coarse parenchymal texture (Figure 2C). Serum B-type natriuretic peptide level was elevated to 51.0 mg/dL. Therefore, the patient was diagnosed as having severe PR and right heart failure followed by chronic liver damage. A decision was made after multidisciplinary discussion to convert to 1.5VR.

Intraoperatively, the Gore-Tex monocusp pulmonary valve was severely calcified, and it was replaced with a bioprosthetic valve (25-mm Epic; St. Jude Medical, St. Paul, MN). The RV endocardium was severely scarred as a result of muscle resection at the time of RV overhaul and BVR, and this finding was considered to be closely associated with the presence of restrictive RV physiology. The postoperative course was uneventful. Three months after 1.5VR and pulmonary valve replacement, although restrictive RV physiology was still observed by echocardiography (Figure 3B; Videos 3 and 4), the patient's symptoms were dramatically reduced, RV and RA sizes were decreased (Figure 3A), RA pressure was decreased to 5 mm Hg, and serum B-type natriuretic peptide level was decreased to 14.8 mg/dL. Moreover, abdominal ultrasound showed that liver congestion had diminished (Figure 3C).

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Keywords: Adult congenital heart disease, Pulmonary atresia with intact ventricular septum, Restrictive right ventricular physiology

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**VIDEO HIGHLIGHTS**

**Video 1:** Apical four-chamber window on preoperative transthoracic echocardiography demonstrating a dilated right heart.

**Video 2:** Parasternal short-axis window on preoperative transthoracic echocardiography demonstrating diastolic flow reversal in branch pulmonary arteries indicating severe PR.

**Video 3:** Apical four-chamber window on postoperative transthoracic echocardiography demonstrating decreased RA and RV volumes compared with those on preoperative transthoracic echocardiography.

**Video 4:** Parasternal short-axis window on postoperative transthoracic echocardiography demonstrating diminished PR.

View the video content online at [www.cvcasejournal.com](http://www.cvcasejournal.com).

**DISCUSSION**

Surgical strategies used for patients with PA-IVS vary among institutions because of a lack of evidence for achieving optimal physiology in this unique anatomy. Because our institutional strategy is to aim for BVR, our choice of initial palliation includes a modified Blalock-Taussig shunt with pulmonary valvotomy to achieve growth of right-sided heart structures. After the initial surgical palliation, an RV overhaul procedure is performed for patients with RVEDV < 50% of the predicted normal value.<sup>1,2</sup> Finally, we generally consider a TV Z score > -3 as an indication for BVR. If the TV Z score is < -8, we consider univentricular repair favorable. Patients with TV Z scores between -8 and -3 are considered

candidates for 1.5VR. In the present case, because RVEDV was a borderline value for BVR, RV overhaul was performed before establishing biventricular circulation.

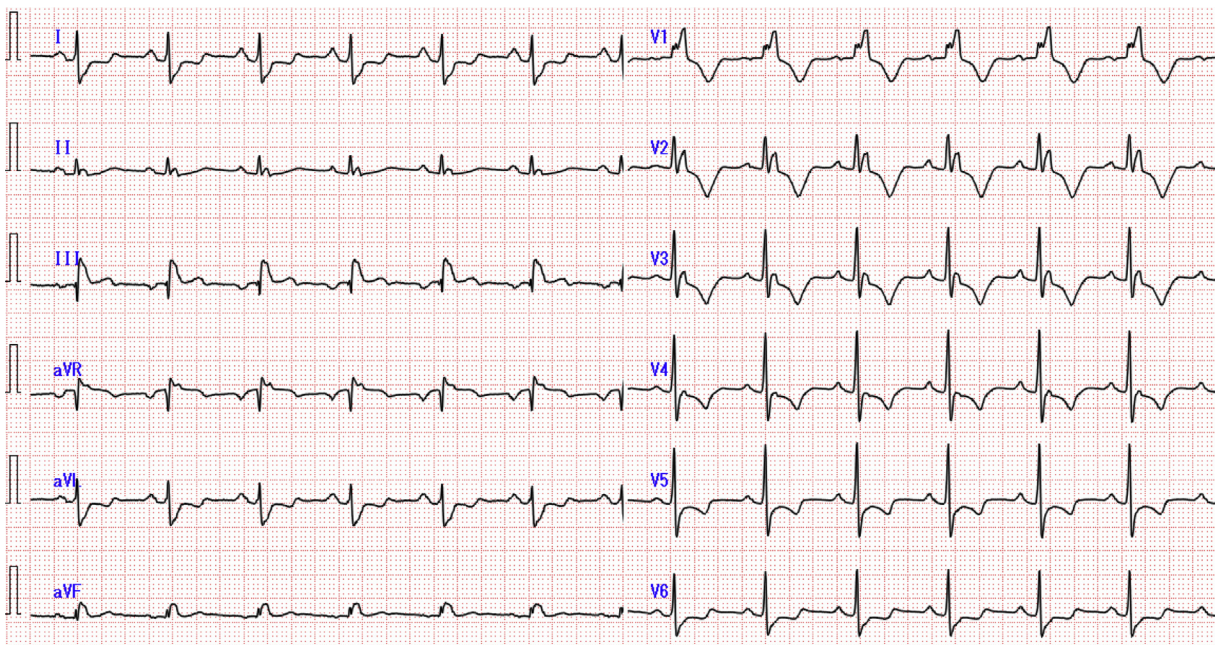
Although long-term survival is relatively good in BVR patients,<sup>4</sup> restrictive RV physiology after BVR is often observed,<sup>5</sup> and it is associated with elevated RA pressure, RA dysfunction,<sup>7</sup> and increased central venous pressure, leading to end-organ damage such as liver congestion. At our institution, according to our accumulated experiences<sup>1</sup> and previous reports,<sup>8</sup> 1.5VR conversion is performed for patients with PA-IVS with failed BVR who fulfill the following criteria: (1) heart failure symptoms, (2) significant PR, (3) elevated RA pressure (>10 mm Hg), and (4) liver damage assessed by ultrasound. In the present case, after successful 1.5VR conversion concomitant with pulmonary valve replacement, RA pressure and serum B-type natriuretic peptide were significantly decreased, and liver congestion was markedly improved. However, restrictive RV physiology was still observed after 1.5VR conversion, which may be related to either the RV overhaul procedure or unique RV anatomy in patients with PA-IVS. Therefore, close follow-up is mandatory even after 1.5VR conversion from BVR.

**CONCLUSION**

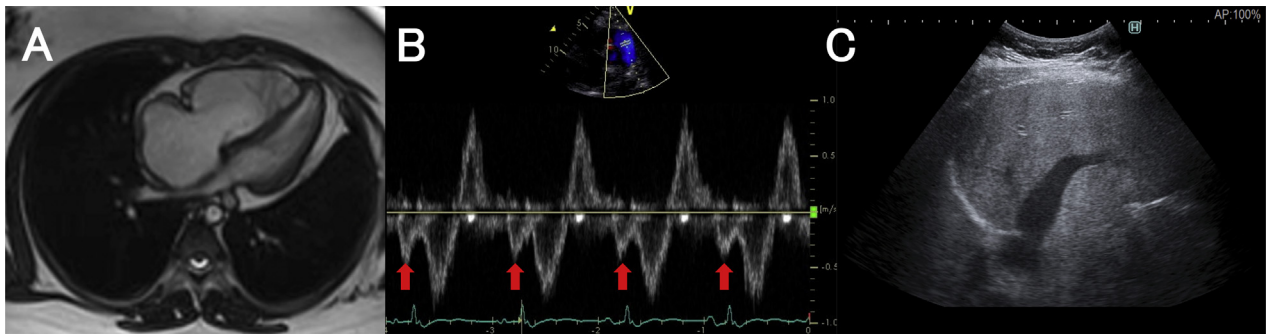
Restrictive RV physiology is a unique finding in patients with PA-IVS who have undergone BVR. This case demonstrated that restrictive RV physiology persisted even after 1.5VR conversion from BVR.

**SUPPLEMENTARY DATA**

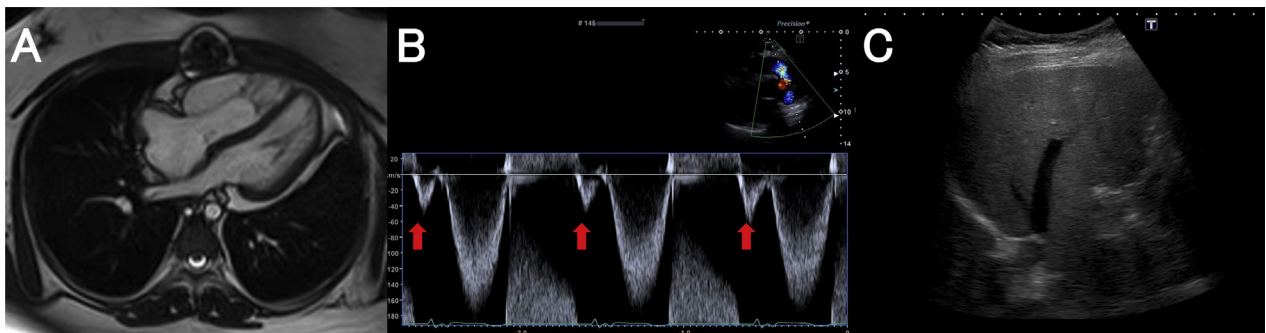
Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.08.002>.



**Figure 1** Electrocardiogram obtained at cardiology visit for heart failure symptoms, showing normal sinus rhythm with complete right bundle block and prolonged QRS duration of 138 msec.



**Figure 2** Preoperative images. **(A)** Cardiac magnetic resonance imaging showed a dilated right heart, and RVEDVi was  $65 \text{ mL/m}^2$ . **(B)** On Doppler echocardiography, an end-diastolic forward flow of  $44 \text{ cm/sec}$  to the pulmonary trunk (red arrow) was observed, suggesting the presence of restrictive RV physiology. **(C)** Liver ultrasound revealed severely dilated hepatic veins, an irregular surface, and a coarse parenchymal texture.



**Figure 3** Postoperative images. **(A)** Right heart size became smaller after 1.5VR conversion, and RVEDVi was decreased to  $54 \text{ mL/m}^2$  on cardiac magnetic resonance imaging. **(B)** An end-diastolic forward flow of  $45 \text{ cm/sec}$  (red arrow) was still observed even though 1.5VR conversion concomitant with pulmonary valve replacement was successfully performed. **(C)** The hepatic vein diameter was significantly reduced, indicating that liver congestion was diminished.

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