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Successful Fontan Completion of a Single Ventricle with Floating Dysplastic Right Ventricle within the Left Ventricle

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Conflict of Interest
The authors have no financial conflicts of interest.

A full-term female neonate who had been diagnosed as tricuspid atresia with severely dysplastic right ventricle (RV) during the fetal period (**Movie 1**) was transferred to the neonatal intensive care unit at birth. Transthoracic echocardiography after birth revealed a dysplastic and hyperechogenic RV chamber partially obstructing the left ventricular outflow tract (LVOT) (**Movies 2** and **3**) and the absence of the pulmonary valve with severe pulmonary regurgitation (**Figure 1**). The mean pressure gradient of the LVOT was 7.8 mmHg and there was no pulmonary hypertension or coronary arteriovenous fistula. She underwent right modified Blalock-Taussig shunt with a 3.5-mm Gore-Tex tube graft (**Figure 2A**) and successful closure of the RV infundibular area 20 days after birth. At 4 months of age, the patient underwent bidirectional cavo-pulmonary shunt (**Figure 2B**) with ascending aorta reduction and pulmonary angioplasty with a tissue graft. Pre-Fontan cardiac catheterization showed good bidirectional cavo-pulmonary shunt flow and minimal LVOT obstruction (LV end-diastolic pressure; 13 mmHg, Qp/Qs; 0.59, Rp; 1.47 WU*m^2). Right coronary artery angiography demonstrated diffuse fistula tract to the RV lumen, enabling RV volume maintenance (**Movie 4**). After careful discussion, we proceeded with an extracardiac conduit Fontan operation with a 16-mm polytetrafluoroethylene graft and 3.5-mm fenestration between the Fontan tract and systemic atrium at 40 months of age (**Figure 2C**). Twenty months following the Fontan operation, the patient is doing well without any arrhythmia events.

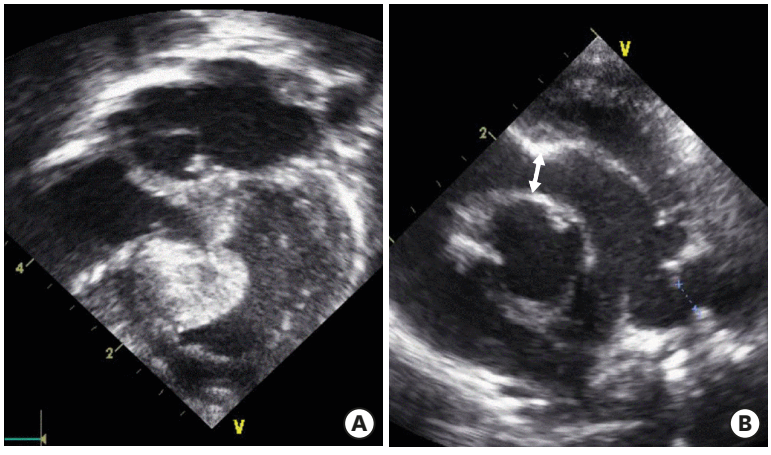

Figure 1. Echocardiography after birth. (A) Severely dysplastic and hyperechogenic right ventricle with tricuspid atresia. (B) Absent pulmonary valve with small pulmonary artery annulus (arrow 4.1 mm).

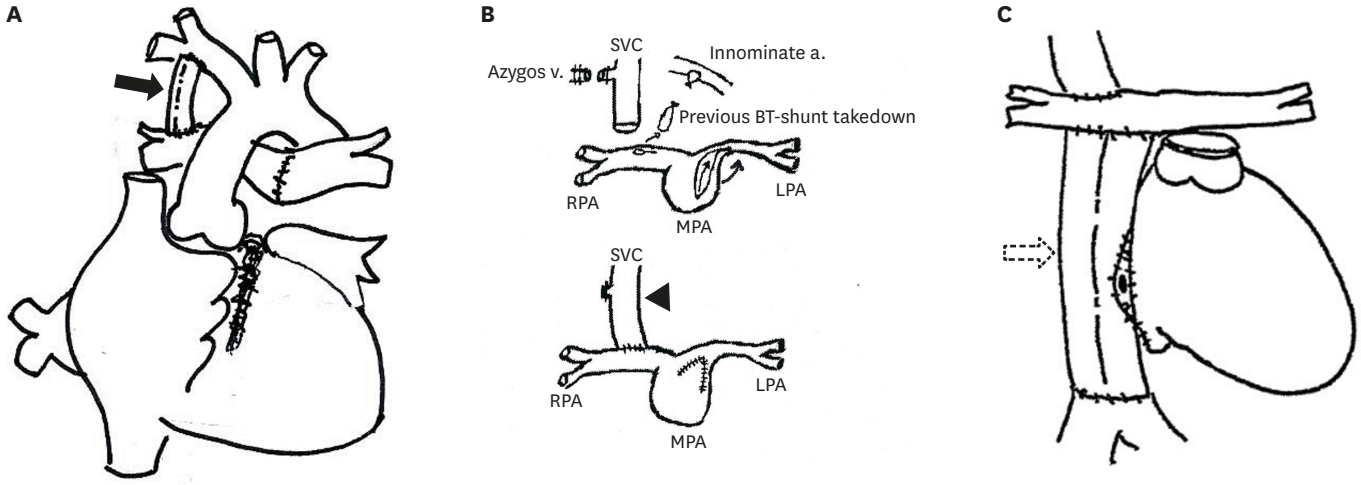

Figure 2. Sequential operation record. (A) Right modified BT shunt with a 3.5-mm Gore-Tex tube graft (arrow). (B) Bidirectional cavo-pulmonary shunt (arrow head) between the SVC and RPA. (C) Fontan operation with a 16-mm polytetrafluoroethylene graft (dotted area) and 3.5 mm fenestration.
SVC: superior vena cava, RPA: right pulmonary artery, LPA: left pulmonary artery, MPA: main pulmonary artery, BT: Blalock-Taussig.

Author Contributions

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Single ventricle is a severe congenital heart disease and may lead to ventricular volume overload.[1][2] LVOT obstruction increases left ventricle (LV) afterload, causing ventricular hypertrophy, which is known as a high risk of Fontan operation.[3][4] Dysplastic RV within a single LV may be fatal due to volume and pressure overload. Fortunately, the degree of left ventricular outflow tract obstruction was mild in this patient, so the patient did not show any symptom from it. Herein we describe a patient who was treated successfully toward Fontan operation, though patient had particularly high-risk for Fontan operation because of RV flotation in the LV partially obstructing LVOT.

SUPPLEMENTARY MATERIALS

Movie 1

Fetal echocardiography at a gestational age of 21 weeks. Severely dysplastic right ventricle and tricuspid atresia with obstructed left ventricular outflow tract are observed.

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Movie 2

Echocardiography after birth. Dysplastic and hyperechogenic right ventricle chamber partially obstructs the left ventricular outflow tract.

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Movie 3

Cardiac magnetic resonance imaging after birth. Functional single LV is present with a mass-like right ventricle chamber partially obstructing the LV outflow tract during diastole. Indexed LV end-diastolic volume: 155.3 mL/m^2, indexed LV end-systolic volume: 60.9 mL/m^2, ejection fraction: 60.8%, and cardiac output: 13.97 L/min/m^2.

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Movie 4

Right coronary artery angiography. Right coronary artery angiography reveals diffuse fistula tract to the right ventricular lumen enabling right ventricular volume maintenance.

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