

Case Report

Successful Staged Fontan Completion for Double-outlet Right Ventricle with Intact Ventricular Septum

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Double-outlet right ventricle with intact ventricular septum is an extremely rare heart malformation. To our knowledge, only 42 cases have been reported, and there is no previous case report achieving Fontan completion. The diagnosis was made by echocardiography and contrast-enhanced cardiac computed tomography. Our patient had additional interesting anomalies such as sinusoidal communications. We describe a successful case of staged Fontan completion for double-outlet right ventricle with intact ventricular septum.

Keywords: double-outlet right ventricle, intact ventricular septum, Fontan completion, sinusoidal communication, balloon atrial septostomy

Introduction

Double-outlet right ventricle (DORV) is characterized by the origin of both arteries from the morphologic right ventricle, and is almost always associated with a ventricular septal defect (VSD) which is necessary for transfer of oxygenated blood from the left ventricle (LV) to the right ventricle (RV). DORV with intact ventricular septum (DORV with IVS) was first described in 1964 by MacMahon and Lipa as “a pure case of double-outlet right ventricle,” in which both great arteries arise completely from the RV.¹ Only 42 cases have been reported according to a previous review by Vairo et al. from 1964 to 2000²) and our search on PubMed database from 2001 to 2015 (Table 1).^{2–8}) Before 2000, most cases had been diagnosed by autopsy. Only 4 patients survived the Glenn procedure,^{3–6}) and there is no previous case report achieving Fontan completion.

Case Report

A female infant was delivered via cesarean section at 37 weeks gestational age with a birth weight of 2,930 g. A fetal echocardiographic study had already diagnosed DORV associated with coarctation of the aorta (CoA). After birth, two-dimensional echocardiography and contrast-enhanced cardiac computed tomography (CT) demonstrated situs solitus, D-loop ventricles, D-side-by-side great arteries with complete origin from the RV (Fig. 1a–c), CoA with an 8.5 mm patent ductus arteriosus (PDA), and a 4.5 mm atrial septal defect (ASD). The ventricular septum was intact (Fig. 1d–f). The LV and mitral valve (MV) were severely hypoplastic; the LV diastolic dimension and MV diameter were 45% and 35% of normal value, respectively. Color and spectral Doppler ultrasound examination revealed little blood flow through the stenotic MV. We considered the infant a Fontan candidate because the LV volume and

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Table 1 Case reports in the literature from 2001 to 2015

Year/Author	Age at diagnosis	Sex	Diagnostic device	BAS	Course of treatment
2001 Vairo et al. ²⁾	3 days	F	echo + angio	+	mBT shunt
2001 Troise et al. ³⁾	2 days	F	echo + angio	+	BCPS
2003 Napoleone et al. ⁴⁾	0 day	No data	echo + angio	+	BCPS
2007 Sakurai et al. ⁷⁾	2 days	M	echo + angio	+	death
2013 Mullula et al. ⁸⁾	0 days	M	echo + angio	+	ASD stent, PA banding
2013 Özgür et al. ⁶⁾	9 days	M	echo + angio	-	BCPS
2015 Menon et al. ⁵⁾	16 months	F	echo	+	BCPS
2017 Okazaki et al. (this report)	0 day	F	echo + CT	+	TCPC

echo, echocardiography; angio, angiography; CT, computed tomography; BAS, balloon atrial septostomy; mBT shunt, modified Blalock-Taussing shunt; BCPS, bidirectional cavopulmonary shunt; TCPC, total cavopulmonary connection.

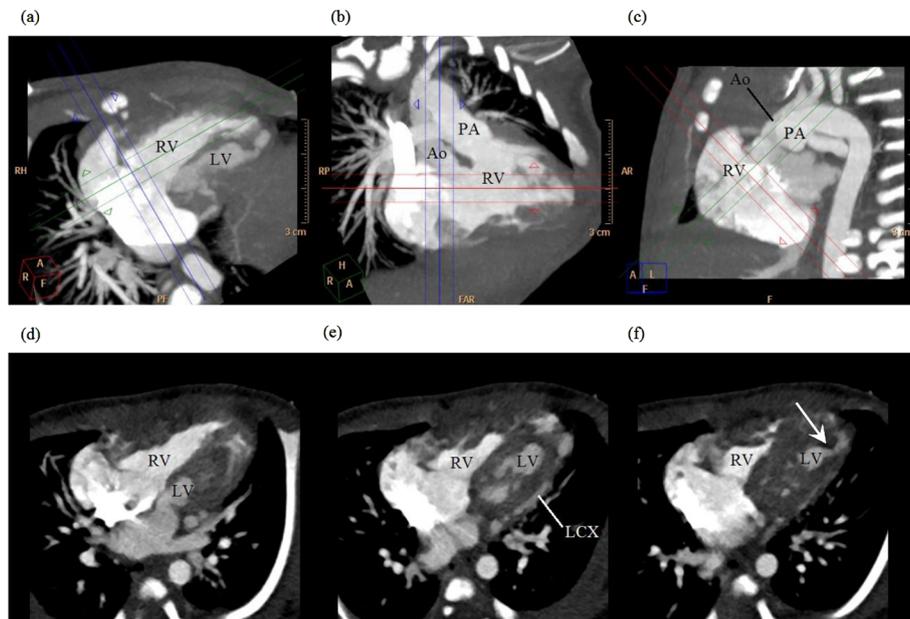


Fig. 1 Contrast-enhanced cardiac computed tomography

(a–c) Multiplanar reconstruction images showing both great vessels entirely arising from the right ventricle. (d–f) Axial images showing an intact ventricular septum and sinusoidal communications between the LV apex and dilated coronary arteries (arrow). RV, right ventricle; LV, left ventricle; PA, pulmonary artery; Ao, aortic artery; LCX, left circumflex coronary artery.

MV diameter were inadequate for biventricular repair. On the other hand, the RV showed adequate volume and contractility. In addition, sinusoidal communications (SC) between the LV apex and dilated coronary arteries were detected. In retrospect, LV myocardial sinusoids draining into the coronary arteries were apparent on fetal echocardiography (Fig. 1f, Fig. 2 and Movie 1).

Prostaglandin infusion was started soon after birth to maintain ductal patency and stabilize hemodynamics. At 6 days of age, surgical palliation consisting of division of the PDA, aortic arch reconstruction, and pulmonary artery banding was performed. At 1 month of age, balloon atrial septostomy (BAS) was carried out

for restrictive ASD that had gradually induced pulmonary congestion and respiratory failure with decreasing oxygen saturation. It was impossible to insert a catheter into the hypoplastic LV because of severe mitral stenosis.

Eight months after surgery, cardiac catheterization showed favorable hemodynamics with mean pulmonary artery pressure 12 mmHg, pulmonary artery index 314 mm²/m², and good RV systolic function. Spontaneous regression of SC and normalization of coronary circulation were also confirmed by angiography and echocardiogram.

At 19 months of age, a bidirectional cavopulmonary shunt (BCPS) was performed and total cavopulmonary

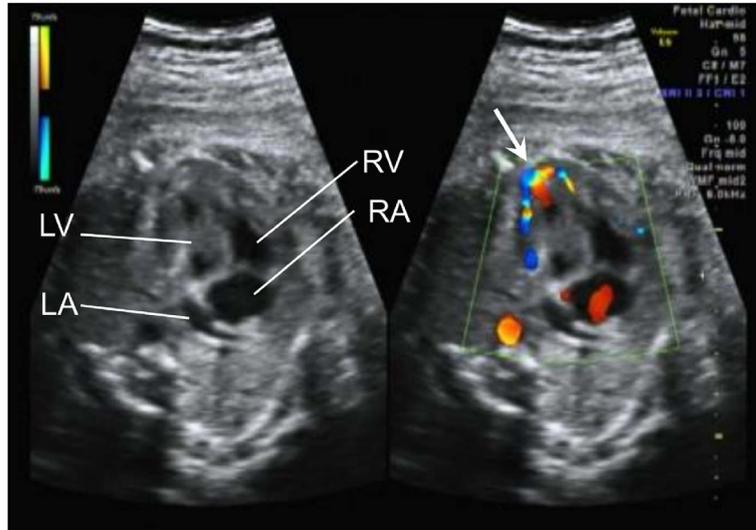


Fig. 2 Fetal echocardiography at 30 weeks of gestation demonstrating a hypoplastic left ventricle and ventriculo-coronary artery connections (arrow)

connection (TCPC) was completed 7 months after the BCPS. The postoperative course was uneventful. Ten months after the TCPC, she was in good clinical condition with 98% oxygen saturation is on room air.

Discussion

This is the first description of successful staged Fontan completion for DORV with IVS. In this very rare malformation, the atrial communication is the only way that oxygenated blood from the lungs reaches the systemic circulation. Despite being indispensable for survival, the ASD of DORV with IVS is usually restrictive and has a strong tendency to close. Therefore, DORV with IVS is a lethal cardiac abnormality if the restrictive ASD is not addressed. Previous reports revealed that almost all cases of DORV with IVS required either BAS or surgical atrial septectomy for survival in the early postnatal period.²⁾

The LV is always severely hypoplastic and nonfunctional owing to absence of the LV outflow tract. DORV with IVS is essentially a single ventricle disease, so the goal of treatment is establishment of a Fontan circulation. In the literature, only 4 patients survived the Glenn procedure.³⁻⁶⁾ Most patients with DORV with IVS might die soon after birth because the interatrial communications either were closed or were too small.

In three out of the four cases of successful palliation to BCPS, mitral avulsion or exclusion were concurrently performed to prevent the influence of LV movement

and mitral regurgitation.³⁻⁵⁾ We did not perform these procedures because the LV had already been excluded from the circulation and did not affect RV movement at the time of BCPS.

Our patient had the additional anomaly of left ventriculocoronary artery connections, also known as SC. The finding had already been detected by fetal echocardiography as the presence of anomalous dilated coronary arteries on the external wall of the heart. There is only one case report of DORV with IVS associated with SC.⁹⁾ It is well known that heart defects with both outflow tract obstruction and IVS—for example, pulmonary atresia with intact ventricular septum (PA/IVS) and hypoplastic left heart syndrome (HLHS),—can be associated with SC and are sometimes diagnosed in the prenatal period.¹⁰⁾ In our case, the disappearance of SC had been confirmed by cardiac CT and echocardiography before BCPS. Though we could not measure the LV pressure, it might already have been reduced at the time of BCPS.

To our knowledge, there is no previous case report of successful staged repair of DORV with IVS. In spite of an extremely rare and complex heart malformation, we succeeded in achieving Fontan completion without severe complications. Long-term follow-up of the patient will be required.

Conflicts of Interest

The authors declare that they have no conflict of interest.

Ethical Approval

This article does not contain any studies with human participants or animals performed by any of the authors.

Note

Supplementary movies are provided online for this article.

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