

## Brief Report

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**Abstract**

Combined tricuspid atresia and absent pulmonary valve with dysplasia of the right ventricular myocardium is a very rare congenital heart anomaly with a poor prognosis. We present three cases of this rare disease that reached the Fontan operation without prior surgical intervention of the right ventricle or pulmonary artery; no right ventricular dilation was detected. All patients had uneventful post-operative courses.

Combined tricuspid atresia and absent pulmonary valve, together with an intact ventricular septum and right ventricular dysplasia, is extremely rare. Surgical management, especially intervention for the non-functional right ventricle and pulmonary artery, has been discussed. In previous reports, right ventricular exclusion and pulmonary artery division or banding were performed concomitantly with the Fontan operation<sup>1,2</sup>. Conversely, we avoid intervention because this could cause right ventricular hypertension and dilation, resulting in left ventricular outflow tract obstruction. We present three cases underwent the Fontan operation with good outcome.

**Case report**

We present three cases of combined tricuspid atresia and absent pulmonary valve detected on fetal echocardiography. All cases underwent right-modified Blalock–Taussig shunt, bidirectional Glenn procedure, and Fontan operation. The right ventricular end-diastolic volume index was measured through cardiac catheterisation. Table 1 summarises the details of the cases. In Case 1, a right ventricular aneurysm was diagnosed during the first echocardiography. The series of right ventriculograms in Case 2 are shown in Figure 1. In all cases, no coronary-to-right ventricular fistulas were detected. Regarding the Fontan operation, an 18-mm polytetrafluoroethylene graft was used. Aspirin was administered as an anti-coagulant. The follow-up periods for the three cases were 10, 8, and 3 years after the Fontan operation. In Case 1 and 2, the latest cardiac catheterizations showed no right ventricular dilation (Table 1). Until now, no left ventricular outflow tract obstruction was noted nor were thromboembolic events.

**Discussion**

Cases of tricuspid atresia and absent pulmonary valve with an intact ventricular septum and right ventricular dysplasia are extremely rare.<sup>1</sup> Only a few cases undergo Fontan operation because of the rarity of the disease and the poor prognosis.<sup>1</sup>

Characteristically, the right ventricular myocardium, similar to that in Uhl's disease, gradually increases in size gradually and affects the left ventricle.<sup>2</sup> Another morphological feature is asymmetrical ventricular septal hypertrophy, which could also lead to left ventricular outflow tract obstruction. Furthermore, pulsatile flow from the right ventricle to the pulmonary artery causes unnatural backward flow in systemic veins, which is a limitation for Fontan circulation.<sup>3</sup> Thus, surgical interventions for right ventricle and pulmonary artery remain controversial.

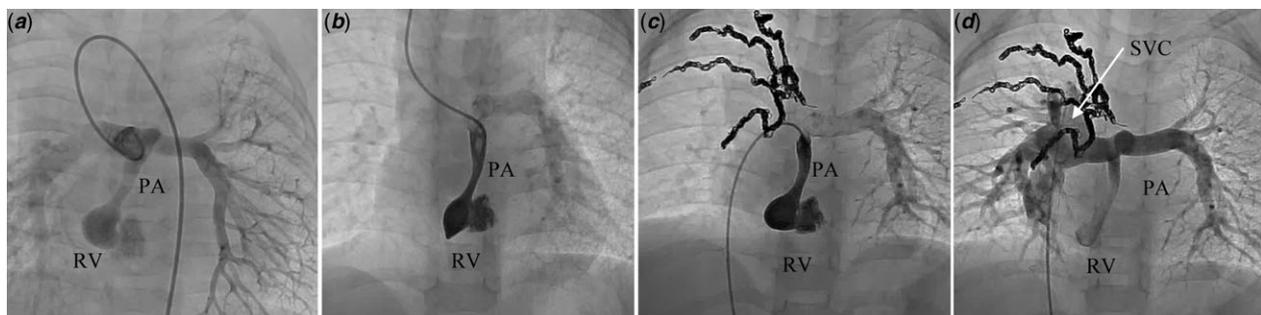
In our practice, they are not manipulated because right ventricular dilation may cause right ventricular hypertension. The inflow to the right ventricle, such as from the Thebesian veins, could contribute to the increased pressure. The presence of significant coronary-to-right ventricular fistulas would have further increased the inflow. We believed that right ventricular hypertension would have exacerbated right ventricular hypertrophy, and pressure needs to be reduced by opening the tricuspid valve or retaining pulmonary artery forward flow. This is a completely different approach to those mentioned in previous reports. To date, the volume of the right ventricle has not increased to such a point so as to cause left ventricular outflow tract obstruction. Regarding the flow in the systemic veins after the Fontan operation, cardiac catheterisation detected no significant pulse flow in the superior or inferior vena cava pressure line. This indicates that the pulmonary artery forward flow does not affect Fontan circulation.

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**Table 1.** Details of the cases

		Case 1	Case 2	Case 3
Right modified Blalock–Taussig shunt	Age	24 days	43 days	31 days
Cardiac catheterisation	Age	17 months	3 months	3 months
	RVEDVI (mL/m <sup>2</sup> )	–	4.0	–
Bidirectional Glenn procedure	Age	19 months	4 months	4 months
	Concomitant procedure	Ventricular aneurysm resection	ASD creation left PA plasty	Left PA plasty
Cardiac catheterisation	Age	4 years	2 years	9 months
	RVEDVI (mL/m <sup>2</sup> )	27	5.8	20
Fontan operation	Age	4.5 years	2.5 years	1 year
Cardiac catheterisation	Age	5.5 years	3.5 years	2 years
	RVEDVI (mL/m <sup>2</sup> )	29	9.5	25
Latest cardiac catheterisation	Age	13 years	8 years	–
	RVEDVI (mL/m <sup>2</sup> )	28	5.6	–

PA = pulmonary artery; RVEDVI = right ventricle end-diastolic volume index.



**Figure 1.** Right ventriculogram of Case 2 during cardiac catheterisation before bidirectional Glenn procedure (a), before Fontan operation (b), and 1 year after the Fontan operation (c). Superior vena cava angiogram after the Fontan operation (d). PA, pulmonary artery; RV, right ventricle; SVC, superior vena cava.

It is difficult to apply this strategy to all cases. From our experience, we believe that this approach is suitable for cases in which right ventricular end-diastolic volume index is less than 30 mL/m<sup>2</sup>. We will continue to monitor for right ventricular dilation, left ventricular function, and the overall condition of these patients.

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**Conflicts of interest.** None.

**Ethical standards.** This article does not contain any studies with human participants.

**Informed consent.** All participants provided informed consent.

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