

An Unusual Cause of Cyanosis after Fontan Procedure in Right Atrial Isomerism



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INTRODUCTION

Heterotaxy syndrome is a complex developmental anomaly related to abnormal right-left axis patterning during embryogenesis.¹ Right atrial isomerism constitutes one of the two entities of heterotaxy syndrome and is characterized by mirror images of right-sided structures on two sides of the body.² Hence, patients with right atrial isomerism have the classic features of bilateral eparterial bronchial branching pattern, bilateral morphological right atrial appendages, and abnormal visceral arrangement with asplenia and a large central liver that wraps the stomach, although discordance among these features remains possible.³

The association of right atrial isomerism with complex congenital cardiac malformations is well documented, with the characteristic ones being complete atrioventricular septal defect, conotruncal anomalies including double-outlet right ventricle and transposition of the great arteries, and anomalous pulmonary venous drainage with or without obstruction.⁴ Fontan procedure is performed for patients with right atrial isomerism and a functional single ventricle. We⁵ and others⁴ have previously reported on unfavorable early and midterm outcomes, although recent reports suggest improved outcomes in these patients.^{6,7} In this report, we describe a child with right atrial isomerism who has an unusual cause of severe cyanosis early after the Fontan procedure.

CASE PRESENTATION

A 4-year-old boy with a cardiac diagnosis of right atrial isomerism, unbalanced atrioventricular septal defect with a dominant right ventricle, double-outlet right ventricle, subpulmonary and valvar pulmonary stenosis, and bilateral superior caval veins developed severe persistent cyanosis after the extracardiac Fontan procedure. Prior to the Fontan procedure, the patient had undergone bilateral bidirectional superior cavopulmonary anastomoses and transcatheter occlusion of the pulmonary outflow using an Amplatzer muscular VSD occluder (AGA Medical, Plymouth, MN) at, respectively, 3 and

10 months of age. During follow-up, his oxygen saturation in room air was about 80%. Pre-Fontan echocardiographic and catheterization assessments revealed nonobstructed bilateral superior cavopulmonary anastomoses, mild stenosis of the proximal right pulmonary artery, satisfactory systemic ventricular function, and moderate degree of atrioventricular valvar regurgitation. The inferior caval vein was noted to be right sided and joined by the hepatic veins before emptying into the right atrium.

Extracardiac Fontan procedure, with the use of a 19 mm fenestrated Gore-Tex (W.L. Gore & Associates, Newark, DE) conduit and repair of the common atrioventricular valve, was performed at 44 months of age. The postoperative course was complicated by low cardiac output syndrome, junctional rhythm, ventricular dysfunction, moderate degree of atrioventricular regurgitation, a relatively high Fontan circuit pressure of 20 mm Hg, and acute kidney injury. He was stabilized with inhaled nitric oxide, oral sildenafil, inotropic support, temporary atrioventricular sequential pacing, and renal replacement therapy and discharged from the intensive care unit 10 days after operation. The patient, however, developed progressive worsening of cyanosis with oxygen saturation decreasing from an initial 80% to 60%–70% at rest and 50%–60% during exertion at 2–3 weeks after the Fontan procedure. Despite increasing the dose of sildenafil, addition of bosentan, and optimization of heart failure medications including diuretics and angiotensin-converting enzyme inhibitor, the oxygen saturation did not improve.

Transthoracic echocardiographic assessment with color flow Doppler mapping showed reversal of flow in the right hepatic veins, venous communications between the right and left hepatic veins, and drainage of the left hepatic veins into the left-sided atrium (Figure 1A–C and Videos 1 and 2). Flow through the fenestration could also be demonstrated (Video 3). Echocardiographic recordings performed during the neonatal period (Video 4) were reviewed. The assessment then gave the impression that all of the hepatic veins join the inferior caval vein, which empties into the right atrium.

To define the hemodynamics of the Fontan circuit and the level and magnitude of right-to-left shunting of blood, cardiac catheterization was performed at 3 weeks after surgery. The mean Fontan circuit and pulmonary arterial pressure was 13 mm Hg at baseline and increased to 15 mm Hg after fluid challenge, the indexed pulmonary vascular resistance was 0.78 WU·m², and the systemic ventricular end-diastolic pressure was 17 mm Hg at baseline and increased to 20 mm Hg after fluid challenge. Angiography showed unobstructed bilateral superior cavopulmonary anastomoses, mild narrowing at the inferior caval vein–Gore-Tex conduit junction, satisfactory ventricular contractility, and communications between the right and left hepatic venous system (Figure 2). A significant degree of right-to-left shunting of contrast through the multiple venous channels that communicate with both the right and left hepatic venous system was demonstrated (Videos 5 and 6).

Computed tomography with coronal reformation showed a centrally located liver, three right hepatic veins joining the inferior caval

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VIDEO HIGHLIGHTS

Video 1: Transthoracic two-dimensional echocardiographic and color flow Doppler mapping assessment after Fontan procedure from the subcostal view showing reversal of flow in the inferior caval vein and right hepatic vein, right-to-left shunting through the venous communications between right and left hepatic veins, and drainage of the left hepatic veins into the left-sided atrium.

Video 2: Transthoracic two-dimensional echocardiographic and color flow Doppler mapping assessment after Fontan procedure from the subcostal sagittal view showing reversal of flow in the right hepatic vein.

Video 3: Transthoracic two-dimensional echocardiographic and color flow Doppler mapping assessment after Fontan procedure from the subcostal coronal view showing patency of the Fontan fenestration.

Video 4: Transthoracic two-dimensional echocardiographic and color flow Doppler mapping assessment from the subcostal coronal view with posterior to anterior panning of the ultrasound probe giving the impression of joining of all hepatic veins to the inferior caval vein, which empties into the right atrium.

Video 5: Angiogram in frontal projection performed with a pigtail catheter positioned in the extracardiac conduit showed mild narrowing at the inferior caval vein–Gore-Tex conduit junction and right-to-left shunting of contrast through the Fontan fenestration and communications between the right and left hepatic venous system. Another pigtail catheter was positioned in the right ventricle for measurement of systemic ventricular end-diastolic blood pressure.

Video 6: Angiogram in lateral projection performed with a pigtail catheter positioned in the extracardiac conduit showing mild narrowing at the inferior caval vein–Gore-Tex conduit junction, the patent Fontan fenestration, and absence of branch pulmonary arterial stenoses. Another pigtail catheter was positioned in the right ventricle for measurement of systemic ventricular end-diastolic blood pressure.

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vein that emptied into the extracardiac conduit, two left hepatic veins joining the inferior part of the left-sided atrium, and venous channels connecting the right and left hepatic venous systems (Figure 3).

Reoperation with placement of a 16 mm fenestrated Gore-Tex conduit between the left hepatic venous entry into the atrium and the previously placed extracardiac Gore-Tex conduit and further repair of the atrioventricular valve was performed. The fenestration, which drained into the left-sided atrium, was created given the mildly increased pulmonary arterial and Fontan circuit pressure. The postoperative course was smooth, and an oxygen saturation of about 85% was achieved. Predischarge echocardiography showed unobstructed forward flow in both left and right hepatic veins (Figure 4), a patent fenestration in the newly placed Gore-Tex conduit, and mild to moderate degree of residual atrioventricular valvar regurgitation. Medications

on discharge included furosemide, spironolactone, enalapril, sildenafil, bosentan, and warfarin.

DISCUSSION

Cyanosis may persist after Fontan procedure. Common causes include right-to-left shunting of systemic venous blood through the fenestration, veno-venous collaterals that drain into the pulmonary venous atrium, and/or existence of pulmonary arteriovenous fistulae.⁸ Furthermore, pulmonary arteriovenous malformations may develop after Fontan procedure to cause worsening of cyanosis. By contrast, the occurrence of right-to-left shunting of blood via the hepatic venous system is an uncommon occurrence found usually in the context of atrial isomerism. Uemura *et al.*⁹ described bilateral drainage of the hepatic veins to the atria via channels independent of the inferior caval vein in 18% of the 125 postmortem cardiac specimens from patients with right atrial isomerism. In 72 postmortem cases with visceral heterotaxy and asplenia suggestive of right atrial isomerism, Rubino *et al.*¹⁰ reported that 18 (25%) had some of the hepatic veins draining separately from the inferior caval vein into the contralateral atrium.¹⁰

In diagnosing the cause of persistent cyanosis in patients after Fontan procedure, agitated saline contrast echocardiography has been used to define the presence, type, and size of the right-to-left shunts.⁸ Nonetheless, to define the possible site of right-to-left shunt, the agitated saline has to be injected at different sites including the superior and inferior caval veins, innominate vein, Fontan circuit, and branch pulmonary arteries. In our patients, color flow Doppler mapping on transthoracic echocardiographic assessment has revealed hints of right-to-left shunting through hepatic venous communications and the Fontan fenestration, which are confirmed by angiography.

There are limited reports on the clinical consequence and management of residual hepatic venous drainage into the systemic circulation after Fontan procedure in patients with right atrial isomerism. Tofeig *et al.*¹¹ reported successful occlusion of a hepatic vein-to-pulmonary venous atrium fistula by an Amplatzer septal occluder in an 8-year-old boy with right atrial isomerism at 2 years after the Fontan procedure. Giamberti *et al.*¹² reported, on the other hand, ligation of the anomalous venous communication for the management of two Fontan patients with right atrial isomerism and significant right-to-left shunting of blood from the inferior caval vein to a hepatic vein that drained into the left-sided atrium. In a relatively large cohort of 17 patients with atrial isomerism, 16 having right and one having left atrial isomerism, with separate hepatic venous drainage, Nakata *et al.*¹³ reported the various surgical approaches to direct the separate hepatic venous flow to the pulmonary arteries. These include the use of extracardiac conduits, intra-extracardiac conduits, and lateral tunnels. Hence, the approach to management of this unusual cause of cyanosis after Fontan procedure in the setting of right atrial isomerism is either by surgical ligation or transcatheter device occlusion of the right-to-left hepatic shunts or reinclusion of the residual hepatic venous drainage into the Fontan circuit.

CONCLUSION

Our patient illustrates an unusual cause of cyanosis due to right-to-left shunting of blood from the inferior caval vein and right hepatic veins through hepatic venous communications into the left hepatic veins, the latter emptying into the left-sided atrium in

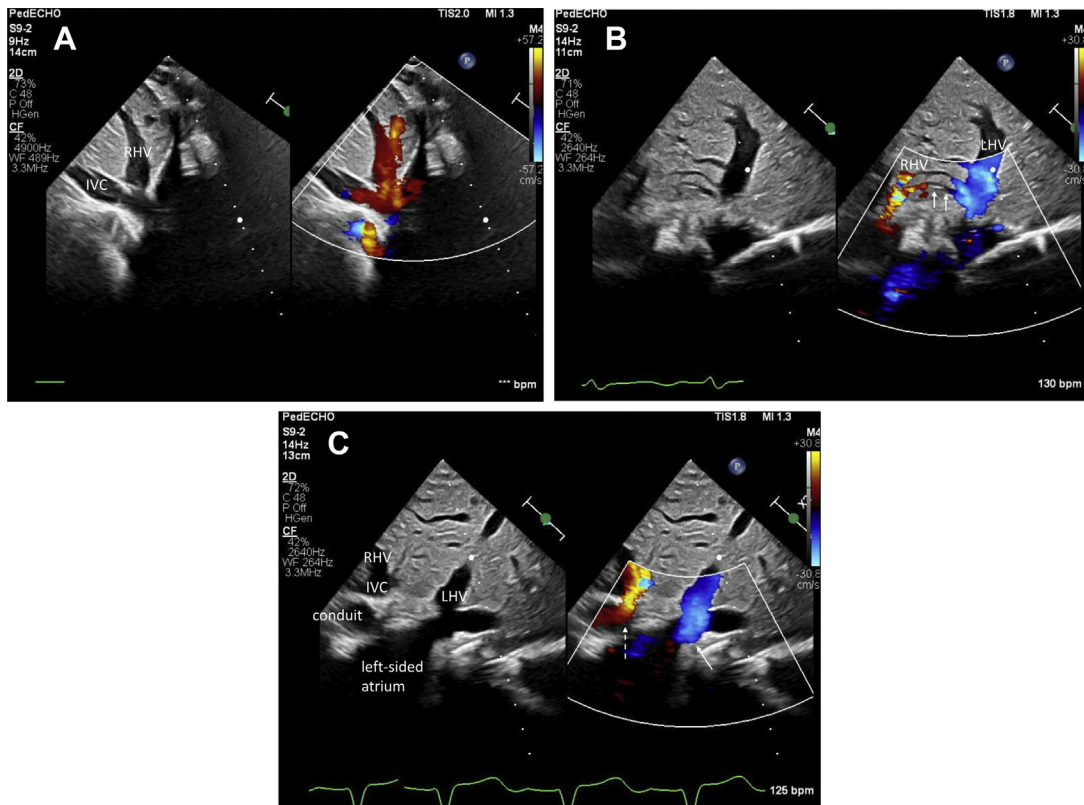


Figure 1 Transthoracic two-dimensional echocardiographic and color flow Doppler mapping assessment after Fontan procedure from the subcostal view showed (A) reversal of flow in the inferior caval vein (IVC) and right hepatic vein (RHV), (B) venous communications (arrows) between the RHV and left hepatic vein (LHV), and (C) reversal flow from the conduit into the IVC and RHV (dashed arrow) and drainage of the LHVs (solid arrow) into the left-sided atrium.

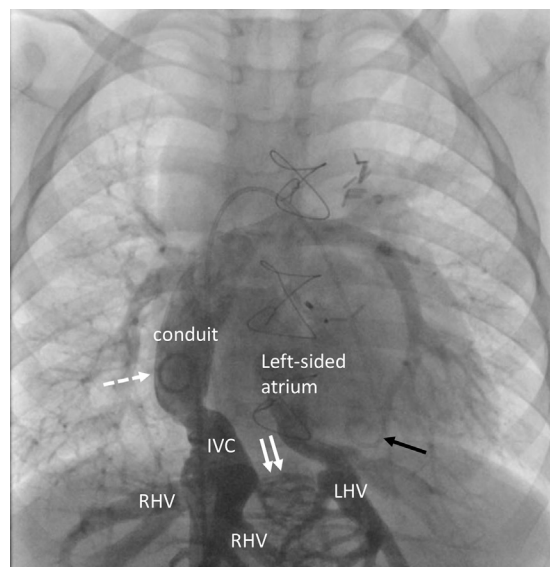


Figure 2 Angiography performed during cardiac catheterization in the frontal projection with a pigtail catheter (dashed arrow) positioned in the extracardiac conduit revealed mild stenosis at the inferior caval vein (IVC)-conduit junction, right hepatic veins (RHVs) draining into right-sided IVC, and left hepatic vein (LHV) receiving desaturated blood via multiple venous communications (white arrows) between the right and left hepatic venous system before emptying into the left-sided atrium. Another pigtail catheter (black arrow) was positioned in the right ventricle for measurement of systemic ventricular end-diastolic blood pressure.

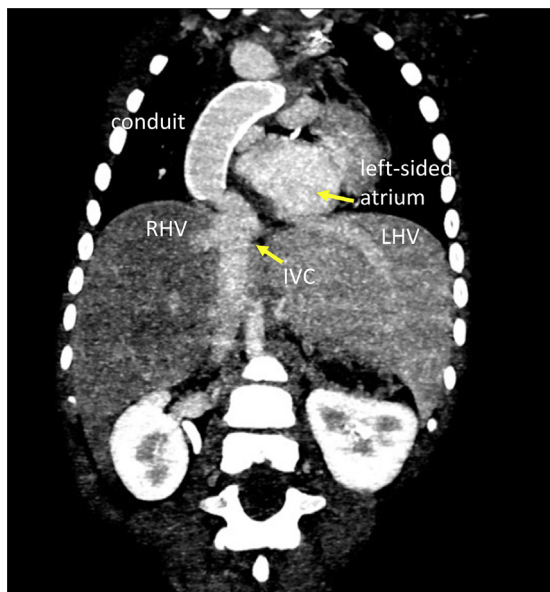


Figure 3 Computed tomography with coronal reformation confirmed a central liver with separate drainage of the right (RHV) and left (LHV) hepatic veins into the inferior caval vein (IVC) and left-sided atrium, respectively.

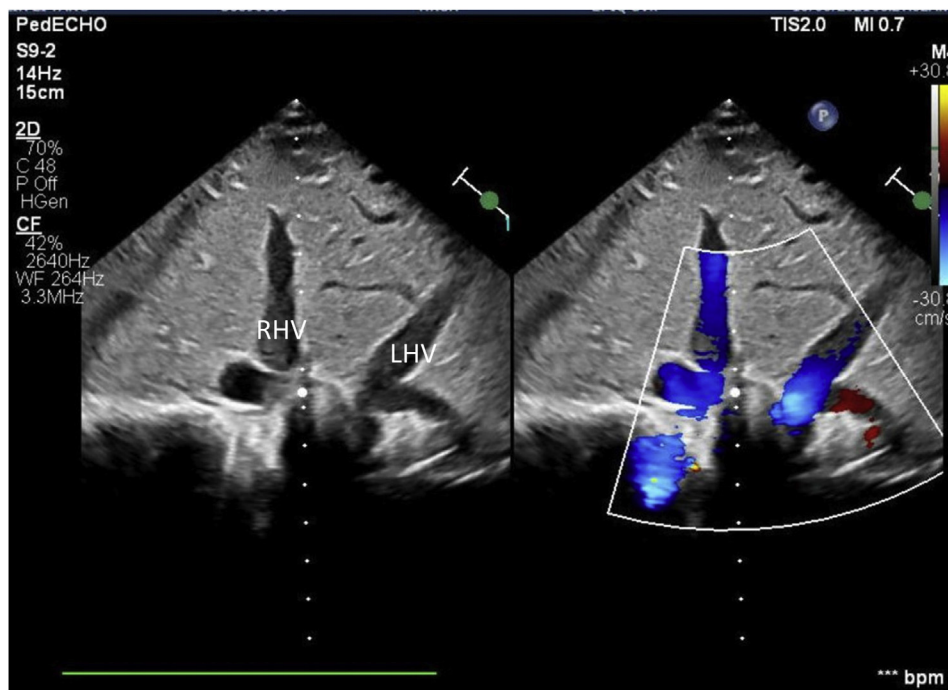


Figure 4 Repeated transthoracic two-dimensional echocardiographic and color flow Doppler mapping assessment after reoperation showed unobstructed flow of both the right (RHV) and left (LHV) hepatic veins in the same direction into the Fontan circuit.

the setting of right atrial isomerism. Detailed delineation of the pattern of hepatic venous drainage is warranted in patients with heterotaxy syndrome during preoperative assessment for Fontan procedure.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.case.2021.11.002>.

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