Heterotaxy syndrome is a complex developmental anomaly related to abnormal right-left axis patterning during embryogenesis. 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. 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.

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. Fontan procedure is performed for patients with right atrial isomerism and a functional single ventricle. We and others have previously reported on unfavorable early and midterm outcomes, although recent reports suggest improved outcomes in these patients. 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.

From the Department of Paediatrics and Adolescent Medicine, LKS Faculty of Medicine, University of Hong Kong (C.K.-m.Y., K.-t.C., Y.-f.C.); and Paediatric Cardiothoracic Surgery Unit (B.A.R.) and Department of Radiology (W.-k.C.N.), Hong Kong Children’s Hospital, Hong Kong, People’s Republic of China.

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Correspondence: Professor Yu-fai Cheung, MD, Department of Paediatrics and Adolescent Medicine, University of Hong Kong, Hong Kong Children’s Hospital, 1 Shing Cheong Road, Kowloon Bay, Kowloon, Hong Kong, People’s Republic of China. (E-mail: xfcheung@hku.hk)

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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. 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. 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, Rubin et al. reported that 18 (25%) had some of the hepatic veins draining separately from the inferior caval vein into the contralateral atrium.

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. 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. 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. 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. 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 reinclution 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.
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.

**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.

**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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