

Accessory Hepatic Vein Complicating Extra-cardiac Total Cavopulmonary Connection

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We encountered unexpected, severe hypoxia after the right heart bypass operation in a patient with isomerism. A 2-year-old girl with polysplenia had a complex cardiac anomaly consisting of a single atrium, single ventricle, pulmonary stenosis, absence of the right superior vena cava, hemiazygos continuation of the left inferior vena cava, and d-malposition of the great arteries. After a total cavopulmonary shunt, we performed an extra-cardiac total cavo-pulmonary connection with a 14 mm tube graft. The postoperative course was complicated by severe hypoxia. Angiography performed 20 days after the operation showed that contrast medium in the conduit poured into the hepatic vein, and through the intrahepatic communications, it passed into a left-sided accessory hepatic vein, which was connected directly to the left side of the aspect of the atrium. As the intrahepatic communication was adequate, we ligated the accessory hepatic vein within the pericardial cavity. The SpO₂ returned to normal and no hepatic dysfunction was detected. We conclude that surgeons performing extra-cardiac total cavopulmonary connection need to pay closer attention to the possibility that an accessory hepatic vein might exist. (*Ann Thorac Cardiovasc Surg* 2002; 8: 112–4)

Key words: accessory hepatic vein, bilateral hepatic veins, extra-cardiac TCPC, Fontan's operation, pitfall

Introduction

The use of extra-cardiac total cavopulmonary connection (TCPC) with vascular conduit in right heart bypass operations has become increasingly common. Its popularity is due to the fact that it results in better hemodynamics, has a lower incidence of atrial arrhythmias, and is simpler to perform than other procedures. We now routinely employ this method whenever possible. Recently, however, we encountered unexpected, severe postopera-

tive hypoxia in a patient with isomerism. The risk of this complication is an under appreciated pitfall of TCPC.

Case

A 2-year 3-month-old girl was found to have polysplenia at laparotomy for interruption of the jejunum at 1 month of age. She also had a complex cardiac anomaly consisting of a single atrium, single ventricle, pulmonary stenosis, absence of the right superior vena cava, hemiazygos continuation of the left inferior vena cava, which was connected to the left superior vena cava, and d-malposition of the great arteries. A modified right Blalock Taussig shunt was performed when the patient was 4 months old, and at 1 year 3 months of age, she underwent a total cavopulmonary shunt with takedown of the Blalock Taussig shunt. At 2 years 3 months of age, we performed

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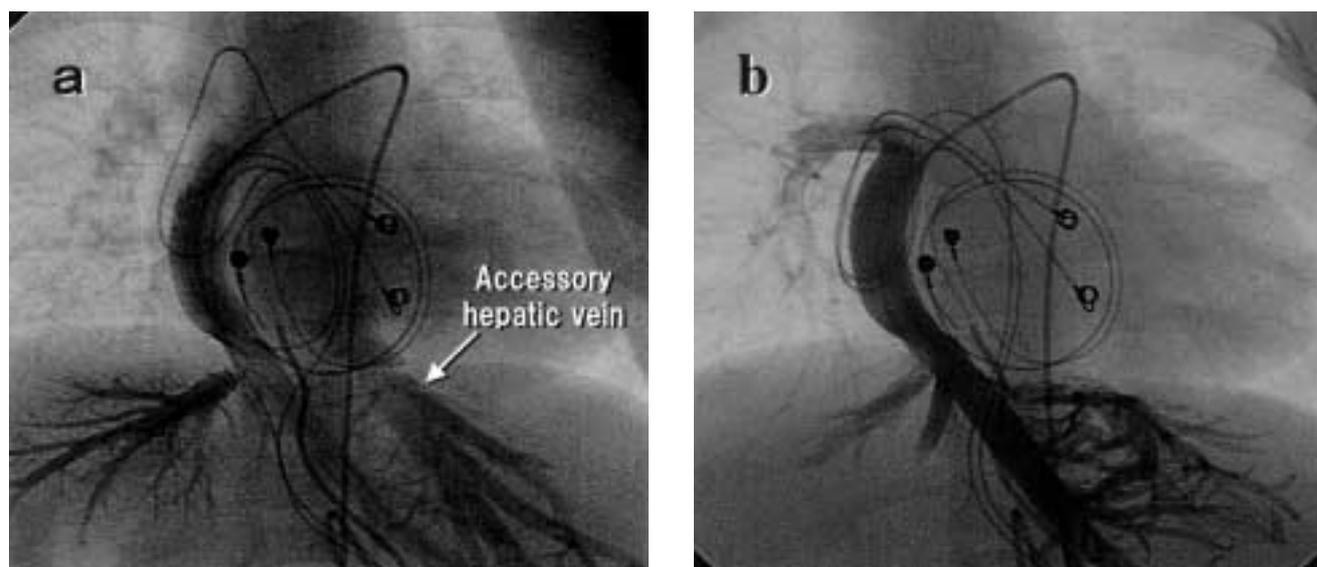


Fig. 1. Angiography showing an accessory hepatic vein following total cavopulmonary connection.

- a: Contrast medium in the conduit poured into the hepatic vein, and through intrahepatic communications, it passed into a left-sided accessory hepatic vein, which connected directly to the left side of the posterior aspect of the atrium.
- b: Postoperative angiography shows that all the venous return from the liver passes into the pulmonary artery.

a TCPC and implanted a permanent pacemaker for polysplenia-related arrhythmias. During this operation, we placed a 14 mm expanded polytetrafluoroethylene graft between the right-sided hepatic vein and a pulmonary artery. The operation was uncomplicated, but the postoperative course was complicated by severe hypoxia. The peripheral arterial oxygen saturation (SpO_2) was 98 to 100% immediately after the patient was weaned off cardiopulmonary bypass, but it gradually decreased to 90% by the end of surgery, and decreased to 80% in the intensive care unit, where it remained. Echocardiography and scintigraphy did not uncover any cause for the hypoxia. Angiography was performed 20 days after the operation. Contrast medium in the conduit poured into the hepatic vein, and through the intrahepatic communications, it passed into a left-sided accessory hepatic vein, which was connected directly to the left side of the aspect of the atrium (Fig. 1a). This shunt was the cause of the hypoxia. As the intrahepatic communication was adequate, we ligated the accessory hepatic vein within the pericardial cavity through a median sternotomy on the 26th postoperative day. The SpO_2 returned to normal after the ligation, and no hepatic dysfunction was detected. Postoperative angiography showed that all of the venous blood from the liver passed into the pulmonary artery (Fig. 1b). The patient is well 6 months after surgery.

Discussion

An accessory hepatic vein is considered rare. No description of an accessory hepatic vein exists in congenital heart surgery nomenclature and the database project of the Society of Thoracic Surgeons.¹⁾ The presence of an accessory hepatic vein alone does not indicate any disorder, and routine preoperative cardioangiography cannot detect this vessel. Recently, extra-cardiac conduit grafting between the inferior vena cava or hepatic vein and the pulmonary artery in TCPC has become common. Usually the inferior vena cava or hepatic vein is snared with a single tie, and an atriotomy is avoided. This means that the likelihood of discovering an accessory hepatic vein located behind the inferior vena cava or the hepatic vein is small. In our patient, we saw a protrusion in the posterior region of the pericardium, but we thought that it might have been a hemiazygos continuation of the inferior vena cava. However, the hemiazygos vein is unlikely to be located within the pericardial cavity.

Two similar cases were reported by Fernandez-Martorell et al. in 1996,²⁾ which involved TCPC with a lateral tunnel. One patient died without reoperation, and the other patient, with heterotaxy, recovered following ligation of the accessory hepatic vein. In 1999, Yoshimura et al.³⁾ reported the case of a patient with asplenia syndrome who also had complications related to an acces-

sory hepatic vein following TCPC by extra-cardiac tube grafting. Postoperatively, the patient's arterial oxygen saturation decreased to 73%. Aortography revealed multiple aortopulmonary collateral arteries arising from the descending aorta. Surgical ligation of a large accessory hepatic vein was performed after coil embolization of the collateral arteries.

In generally, an accessory hepatic vein is rare, but in cases of isomerism of the heart, its prevalence may be higher. Becker and Anderson⁴⁾ reported that "bilateral hepatic veins are frequent in left isomerism and connect isomerically to the atria." We conclude that surgeons performing extra-cardiac TCPC need to pay closer attention to the possibility that an accessory hepatic vein might exist.

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