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Brief Report

Intrahepatic right-to-left shunting after the Fontan operation

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Abstract We describe two patients with right isomerism, corrected with a fenestrated Fontan operation, who suffered severe progressive cyanosis. Cardiac catheterisation in both revealed a massive right-to-left shunt from the inferior caval vein, through the liver, to a hepatic vein draining directly to the left side of the intra-atrial baffle. The anomalous vein was successfully ligated in both patients.

Keywords: Congenital heart defects; isomerism; Fontan operation; cyanosis

Progressive cyanosis after the Fontan operation is often due to pre-existing or newly developed pulmonary arteriovenous fistulas, or intrahepatic venovenous channels producing shunting in the liver.[1–5] As there are relatively few reports of anomalous hepatic venous malformations,[2–5] the aim of our study is to draw attention to this complication, and to discuss its avoidance and management.

Patients and methods

Our first patient was a 7-year-old boy admitted to our hospital because of increasing cyanosis. He was known to have right isomerism, a right-sided heart, left-sided superior and inferior caval veins, atrioventricular septal defect, double outlet right ventricle, pulmonary atresia, and supracardiac totally anomalous pulmonary venous connection to a right vertical vein draining to the brachiocephalic vein.

A central shunt had been constructed when he was a neonate. The totally anomalous pulmonary venous connection was corrected, and a new systemic-to-pulmonary arterial shunt created when he was 2. He then underwent a fenestrated total cavo-pulmonary connection when he was 7.

Following this procedure, he remained cyanosed. The fenestration was closed, and an attempt was made to ligate the vertical vein. In spite of this, he remained cyanosed, and cardiac catheterisation was performed 6 months later. This showed an aortic saturation of 76%, and revealed that the vertical vein was still patent. In addition, there was a massive shunt within the liver between a left-sided hepatic vein connected to the inferior caval vein and a right-sided hepatic vein entering directly the atrial chamber (Fig. 1). The inferior caval vein, and the pathway

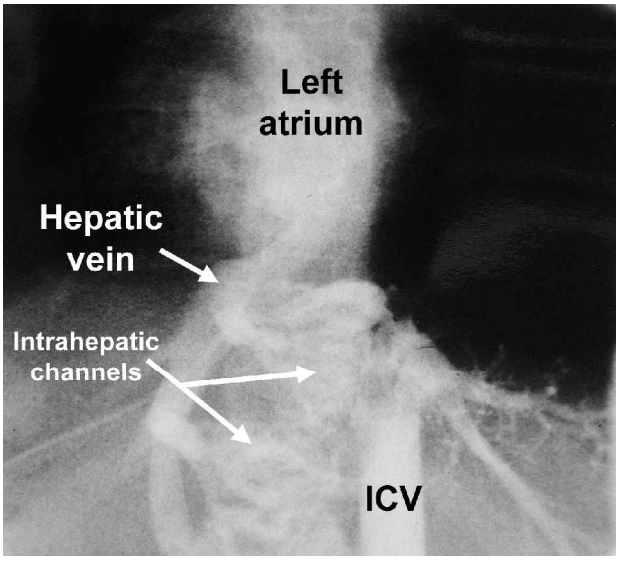

Figure 1.
Intrahepatic collateral channels in our first patient underscoring a right-to-left shunt from the left hepatic vein to the left atrium.

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to the pulmonary arteries, were patent. The pulmonary arterial pressure was measured at a mean of 13 mmHg.

Through a midline sternotomy, and without cardiopulmonary bypass, the vertical vein was snared without the expected improvement of the aortic saturation. Next, the right-sided hepatic vein was identified and snared. The saturation immediately rose from about 75% to 95%, with a rise of about 2–3 mmHg in the systemic venous pressure. The vein was therefore ligated. The postoperative course was uneventful.

Our second patient was a 3-year-old boy who was admitted to our hospital because of increasing cyanosis following construction of a fenestrated total cavo-pulmonary connection. He had right isomerism with supracardiac totally anomalous pulmonary venous connection to a left vertical vein draining to the brachiocephalic vein, complete atrioventricular septal defect, discordant ventriculo-arterial connections, severe pulmonary stenosis, right aortic arch, and a small right-sided patent arterial duct. He had previously undergone construction of a left modified Blalock-Taussig shunt at the age of 5 months, repair of the totally anomalous pulmonary venous connection, and patch enlargement of the origin of the right pulmonary artery. A bidirectional cavo-pulmonary anastomosis was constructed 1 year later. When he was three, a total cavo-pulmonary connection was constructed using an intra-extracardiac fenestrated Gore-tex tube. Desaturation became gradually more severe over the period of follow-up, and we planned to proceed with interventional closure of the fenestration 5 months later.

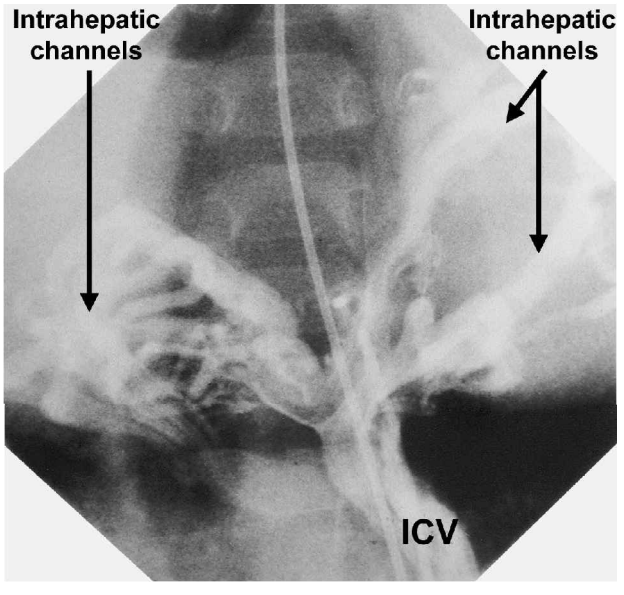

Figure 2.
Similar intrahepatic collateral channels in our second patient permitting a right-to-left shunt into the left atrium.

Cardiac catheterisation showed the presence of a small fenestration, but closure of this communication with a balloon did not change the low saturation. At the same time, the investigation showed the presence of several intrahepatic channels, connected with an anomalous left hepatic vein which drained into the left-sided atrium (Fig. 2).

Through a midline sternotomy, the anomalous hepatic vein was identified and controlled, and the systemic arterial saturation immediately rose to 85%. The venous channel was therefore ligated. The patient had an uncomplicated post-operative course.

Discussion

Normally, two or three large hepatic veins drain the portal circulation from the liver to the inferior caval vein just below the diaphragm. Another 10–15 small veins usually exist, and they open to the inferior portion of the inferior caval vein. A peculiar feature of all these veins is the absence within them of any valves. Very rarely, one of these veins, or an accessory hepatic vein, probably a persisting proximal segment of the embryological left vitelline vein,[3] can drain directly into the right atrium. Direct atrial connection of the hepatic veins, however, is much more common in patients with isomeric atrial appendages.

In left isomerism, the inferior caval vein is interrupted in more than three-quarters of cases. The hepatic veins then always drain directly into the atriums.[6] In right isomerism, the inferior caval vein is hardly ever interrupted, but some hepatic veins can drain separately from the inferior caval vein, either into the same atrium receiving the inferior caval vein or the opposite atrium.[7] The presence of such anomalous or accessory hepatic veins draining directly into the right atrium is of no haemodynamic significance in the normal heart, or prior to the Fontan operation. After a Fontan operation, in contrast, if such a vein continues to drain to the left side of the atrial baffle, the higher systemic venous pressure compared with the pulmonary venous atrium can stimulate the formation of intrahepatic channels. The result is a right-to-left intrahepatic shunt from the inferior caval vein to the left atrium. Pre-operative echocardiographic[8] or angiographic identification of separate drainage of such an anomalous hepatic vein is essential to avoid this complication, especially in patients with right and left isomerism.

Some groups[4] use the physiologic presence of right-to-left intrahepatic shunting as an alternative to fenestration in the Fontan operation. They position the atrial baffle so that one or more of the hepatic veins drain to the pulmonary venous side of

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the baffle. The recruitment of additional channels, nonetheless, can cause progressive cyanosis and, as reported,[4] the need for further operations.

If an anomalously connected hepatic vein has not already been identified, it can be difficult to diagnose in the presence of the Fontan circulation for two reasons. First, the accessory hepatic veins and the intrahepatic channels can be small initially, becoming dilated and physiologically important only with the increase in the flow of blood leading to progressive cyanosis. Second, the desaturation produced by fenestration can initially hide the intrahepatic right-to-left shunt. Progressive severe cyanosis after the Fontan operation, nonetheless, should alert physicians to the likelihood of this complication. The mortality rate is very high in its presence, more than 50%, as reported in the literature.[2–5] When the diagnosis is made, prompt treatment is necessary. Surgical intervention, or transcatheter occlusion, can be considered. We prefer surgical intervention because the procedure is technically simple, and because some attempts at coil occlusion have proved unsuccessful.[2,3] The potential risk of ligation of a hepatic vein is hepatic necrosis. We believe that the collateral intrahepatic channels between the two venous systemic circulations are such that this is unlikely to be a problem. This is confirmed by the marked improvement in systemic arterial saturation immediately after the procedure.

The presence of an accessory hepatic vein joining the pulmonary venous atrium, therefore, should always be considered in patients developing progressive cyanosis after the Fontan operation, particularly in those with right and left isomerism. Based on our experience, surgical ligation of the accessory hepatic vein is safe, technically simple, and effective.

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