Reconstruction of cavopulmonary pathway for the patient with persistent arteriovenous malformations due to offset flow from hepatic vein

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The hypoxemia caused by arteriovenous malformations after cavopulmonary shunt in patients with heterotaxy, an interrupted inferior vena cava and single ventricle physiology have been treated by incorporation of hepatic vein flow into the pulmonary circulation. However, some patients have persistent arteriovenous malformations because of offset hepatic venous flow to one pulmonary artery. Various approaches have been used to change offset flow to achieve balanced hepatic flow to the lungs in this patient population. This case report highlights the challenges that may be associated with anastomosis of the azygos vein to the inferior vena cava at the level of the diaphragm and illustrates an alternative technique to direct hepatic venous blood into an affected lung with arteriovenous malformations. The redirection of hepatic venous flow to the affected pulmonary artery resulted in resolution of symptoms within months of surgery.

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Keywords: Arteriovenous malformation, Hepatic vein

In patients with heterotaxy and azygous continuation of an interrupted inferior vena cava (IVC), the formation of pulmonary arteriovenous malformations (AVMs) is relatively common after bidirectional cavopulmonary anastomosis because of deficiency of hepatic vein flow into the pulmonary arteries.

AVMs have been treated by the inclusion of hepatic venous flow into the pulmonary circulation. However, some patients have persistent AVMs because of adverse streaming of hepatic venous flow to one pulmonary artery. Various procedures have been reported to establish balanced flow from the hepatic veins including anastomosis of the azygos vein to the IVC at the level of the diaphragm. However, this latter approach may not always be technically feasible. We report an alternative simple anterior approach for the redirection of hepatic vein flow.

A 13 year old patient underwent placement of a left Blalock shunt at 5 months of age following the diagnosis of heterotaxy with dextrocardia, double outlet right ventricle, hypoplastic right ventricle, pulmonary stenosis and interruption of inferior vena cava (IVC) with azygous continuation. This procedure was complicated by persistent left...
sided pleural effusions resulting in left sided pleurodesis. At 2 years of age a Fontan procedure was undertaken including a right sided bidirectional Glenn shunt and a left sided 16 mm extracardiac conduit with 5 mm fenestration from the hepatic veins to the left pulmonary artery. This procedure was also complicated by persistent pleural effusions lasting several months resulting in right pleurodesis. The patient did well for years, however, was referred to our institution for evaluation due to worsening cyanosis to \( \text{SaO}_2 \) of 65% and exercise intolerance. At cardiac catheterization mean pulmonary artery pressure was 16–17 mmHg with \( Qp/Qs \) of 0.6. The right pulmonary vein saturations varied between 72% and 76% and there was angiographic evidence of right sided AVMs (Fig. 1). There was obvious streaming of venous blood such that hepatic venous blood passed almost entirely to the left lung (Fig. 1). A redirection of hepatic blood flow was planned by connecting the hepatic veins to the azygous vein at the level of the diaphragm in an attempt to reverse the right lung AVMs. It was felt that approach through a thoracotomy, as has been previously described [1–4] was contraindicated because of previous bilateral pleurodeses.

At surgery, via a resternotomy, dissection was undertaken along the inferior surface of the heart, posterior to the hepatic veins with identification of the vertebral bodies. The esophagus was readily identified and was confirmed to lie between the azygous vein and the hepatic veins as noted on MRI scan (Fig. 2). It was felt that anastomosis at this level was not ideal because of the poor exposure with the azygous vein lying directly behind the esophagus. In addition there was concern regarding esophageal compression and erosion by the conduit. An alternative approach was therefore pursued. The superior mediastinum was dissected free. Following heparinization, cardiopulmonary bypass was commenced with the arterial cannula in the ascending aorta and venous return was established via the large left innominate vein and left sided atrium. The large confluence of the azygous vein and right sided superior vena cava was dissected free. A side biting clamp was applied and a 16 mm ring supported Gortex conduit was anastomosed to the right superior vena cava at its junction with the large azygous vein. The conduit was tunneled posterior to the ascending aorta. The previously placed extracardiac conduit was divided between clamps immediately inferior to its anastomosis to the left pulmonary artery. An end to end anastomosis was fashioned between the new Gortex conduit and the old.

Figure 1. MRI of chest prior to surgery. The esophagus was identified anterior to the azygous vein at the level of the diaphragm.

Figure 2. Angiography prior to surgery. Pulmonary angiography ((A) right PA and (B) left PA) demonstrated arteriovenous malformations on the right side. There was obvious streaming such that hepatic venous blood passed almost entirely to the left lung (C).
16 mm extracardiac conduit, oversewing the cranial stump. The postoperative course was uneventful. Chest tubes were removed 3 days after operation. The patient was discharged on the 6th postoperative day. SaO₂ four month after the operation was 95%. Postoperative cardiac catheterization showed an open conduit and resolution of the AVMs (Fig. 3).

Discussion

The hypoxemia caused by AVMs after cavopulmonary shunt in patients with heterotaxy, an interrupted IVC and single ventricle physiology can be reversed in many cases by incorporation of hepatic vein flow into the pulmonary circulation, presumably due to a combination of resolution of AVMs and elimination of hepatic venoatrial right-to-left shunting [5,6]. The resolution of hypoxemia supports the theory that there is a hepatic factor that is absent in the lungs that leads to the formation of AVMs, although the exact factor has not yet been identified [7].

Various approaches have been used and may be effective for changing offset flow to achieve balanced hepatic flow to the lungs in this patient population. This may be achieved by either (1) connecting the hepatic vein and SVC to the pulmonary artery to facilitate mixing of SVC and hepatic venous blood and consequently distributing hepatic venous flow to both pulmonary arteries or (2) directing hepatic blood into the systemic venous pathway upstream of the systemic venous insertion into the pulmonary artery. For the patients with central pulmonary artery hypoplasia, stent implantation into the stenosis site can be performed, though a previous report using this technique failed to show significant improvement [8]. Implantation of a graft from the existing hepatic vein to pulmonary artery conduit into the affected pulmonary artery is another possible option, though a previous report using a branched graft resulted in acute thrombosis of the new conduit in two patients, presumably due at least in part to stasis and competitive flow in the setting of a large effective conduit volume carrying low flow at low velocity and pressure [9]. In our case the original graft was divided and directly anastomosed to the junction of the SVC and azygous vein, which prevented competitive low flow and thrombosis formation.

Direct hepatic to azygous vein connection through a lateral thoracotomy without cardiopulmonary bypass at the level of diaphragm may provide the most reliable and predictable mixing and bilateral distribution of hepatic blood, regardless of the pulmonary artery, systemic venous, and hepatic vein anatomy [1–4]. In our case, anastomosis at this level was not possible because the esophagus was anterior to the azygous vein at the level of the diaphragm, which leads to difficulty with exposure as well as concern regarding esophageal compression and erosion by the conduit. In addition, previous bilateral pleurodeses contraindicated approach to these vessels through a thoracotomy approach. Therefore, an extension conduit was anastomosed between the previous conduit and the azygous vein in the superior mediastinum with routing posterior to the ascending aorta.

In conclusion, this case highlights the challenges that may be associated with anastomosis of the azygos vein to the IVC at the level of the diaphragm and illustrates an alternative technique.
to direct hepatic venous blood into an affected lung with AVMs. The redirection of hepatic venous flow to the affected pulmonary artery resulted in resolution of symptoms within months of surgery.

References


