Hepatoazygos venous shunt for Fontan completion after Kawashima operation

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ABSTRACT

Fontan completion after prior Kawashima repair for single ventricle with interruption of the inferior vena cava can be accomplished by various methods. We describe a patient who underwent the connection of hepatic to hemiazygos vein that we believe would be superior to the conventional cavopulmonary connection in our patient.

Keywords: Cavopulmonary connection, hemiazygos vein, hepatic vein, Kawashima

INTRODUCTION

Total cavopulmonary connection as definitive palliation for univentricular hearts is well established. With better understanding of Fontan physiology and its associated complications, the procedure itself has undergone continuous evolutions since its inception.

In those patients with the interruption of inferior vena cava (IVC) and azygos or hemiazygos continuation, The Kawashima operation is a bidirectional superior cavopulmonary shunt performed for first stage palliation of single ventricle patients with interruption of inferior vena cava(IVC). Here, except for the portal venous return, all systemic venous return is made to enter the pulmonary arteries, bypassing the heart.[1] The bypassing of pulmonary circulation by hepatic venous effluent is said to predispose to the development of pulmonary arteriovenous fistulae following Kawashima operation.[2,3] These arteriovenous malformations (AVMs) are responsible for progressive cyanosis. Studies have emphasized the need for Fontan completion following Kawashima operation to prevent/reverse the occurrence of pulmonary AVMs and cyanosis as their sequel.[3,4] The resorption of these malformations and hence the disappearance of cyanosis may take 6 months to 1 year following reestablishment of hepatic venous return to the lungs.[3,4] Here, it is important that hepatic venous flow must be directed to both lungs for complete resolutions of pulmonary AVMs.

Various methods for the surgical completion of routing hepatic veins (HVs) to the pulmonary arteries by means of intracardiac or extracardiac conduits have been described.

CASE REPORT

We report a case of an 8-year, 5-month-old boy who was diagnosed at the age of 18 months with complex congenital heart disease with left atrial isomerism. He had an unbalanced atrioventricular septal defect with a single ventricle (right ventricle) physiology, double outlet right ventricle with main pulmonary and right pulmonary artery (RPA) stenosis, and interrupted IVC with hemiazygos continuation with a single left superior vena cava (LSVC). The child underwent a Kawashima operation with RPA plasty at the age of 2 years, following which his oxygen saturation improved. The child developed restenosis of the RPA, for which for which a stent was deployed at the age of 3 years. This stent later needed dilatation for instent stenosis at 5 years of age. The child presented to us with dyspnea on exertion with progressive cyanosis. On examination, he had central cyanosis with oxygen saturation of 68% on room air with clubbing.

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Echocardiography confirmed the established diagnosis along with situs ambiguous, levocardia, and interrupted IVC with two HVs draining separately into the right-sided atrium. Laminar flow in the superior cavopulmonary anastomosis was documented.

Cardiac catheterization and angiography were performed to record the pulmonary artery pressures, ventricular function, and to identify any significant collateral. Diffuse pulmonary arteriovenous collaterals in both lung fields were noted. A Prominent collateral from the LSVC to the left upper pulmonary vein was occluded using a coil [Figure 1a and b]. The stented RPA was widely patent, and both pulmonary arteries were good-sized. Dye injection in the LSVC demonstrated an equal distribution of flow to both lungs. The LVEDP was 11 mm Hg and the mean pulmonary artery pressure was 15 mm Hg, respectively.

Surgery

The surgery was approached through a redo sternotomy. The cardiac mass and aorta were dissected. Two HVs in different planes were noted draining separately into the right-sided atrium, with the one on the right significantly smaller in size [Figure 2]. Before dissecting out the pulmonary arteries, the posterior mediastinum was exposed just above the diaphragm. The large hemiazygos vein (HAV) was identified to the left of the esophagus. A decision to connect the hepatic venous return to the HAV was taken in view of the proximity of the two vascular structures. The remote right hepatic vein did pose a problem but as it was much smaller in size, it was decided to ligate it. This would also preclude dissection for the pulmonary arteries behind the dilated ascending aorta. The HAV was dissected adequately, and a C-clamp was applied after administering unfractionated heparin at a dose of 1 mg/kg. HAV was opened between stays, and a 14 mm polytetrafluoroethylene tube graft was anastomosed in an end-to-side fashion using 6-0 prolene continuous suture. Initially, an off-pump procedure was contemplated for the connection of the graft to the hepatic vein. However, as there was recurrent hypotension upon repeated attempts to clamp the HVs, it was decided to do the rest of the procedure on cardiopulmonary bypass (CPB). On CPB between ascending aorta and the venous cannula in the right-sided atrium, the larger HV was separated from the atrium with a cuff of the right-sided atrium between clamps and the atrial end oversewn. Remaining anastomosis of the tube graft to the HV was performed in an end-to-end fashion with 6-0 prolene continuous suture after shortening the graft adequately and beveling the end to avoid kinking the HV. The smaller HV was allowed to drain the liver till the graft was opened. It was then ligated [Figures 2 and 3].

The patient was extubated in 24 h. Anticoagulation with warfarin was started to achieve a target international normalized ratio of 2–2.5. Postoperative course was uneventful. Liver function test were within normal limits. The patient was discharged on the 10th postoperative day with oxygen saturation of 68% on room air, as expected, because of diffuse pulmonary AVMs. The postoperative echo showed a good ventricular function with laminar flow in the conduit [Figure 4].

Comments

Options available for completing the Fontan in our case were (a) extracardiac tube from a cuff of atrium bearing both HVs to RPA,[5] (b) intra/extracardiac conduit type of connection, wherein the anastomosis of the tube to the atrium around the HVs entry would be intracardiac which

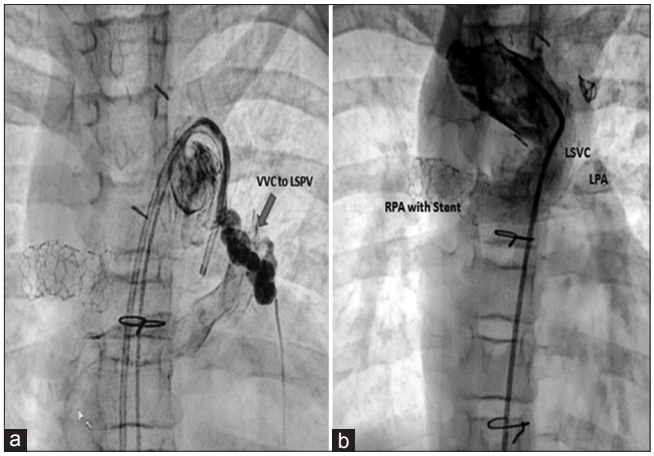

Figure 1: (a) Venovenous collateral draining into the left superior pulmonary vein. (b) Dye injection in the innominate vein showing bilateral equal distribution of contrast through bidirectional Glenn anastomosis into both pulmonary arteries. VVC: Venovenous collateral, LSPV: Left superior pulmonary vein, RPA: Right pulmonary artery, LPA: Left pulmonary artery, LSVC: Left superior vena cava

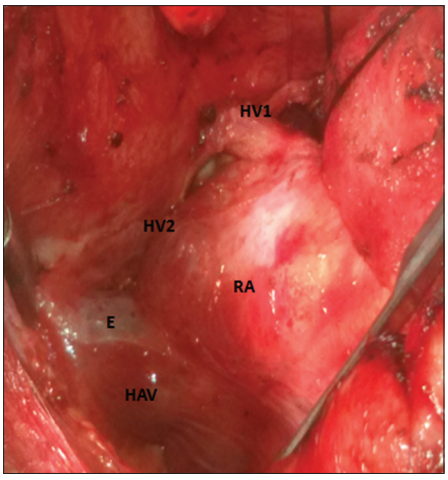

Figure 2: Intraoperative picture of dissection of the diaphragmatic surface of the heart showing the two hepatic veins (HV1, HV2) draining into the right atrium. In addition, the hemiazygos vein to the right of the esophagus (E) in the posterior mediastinum. RA: Right atrium, HV: Hepatic veins

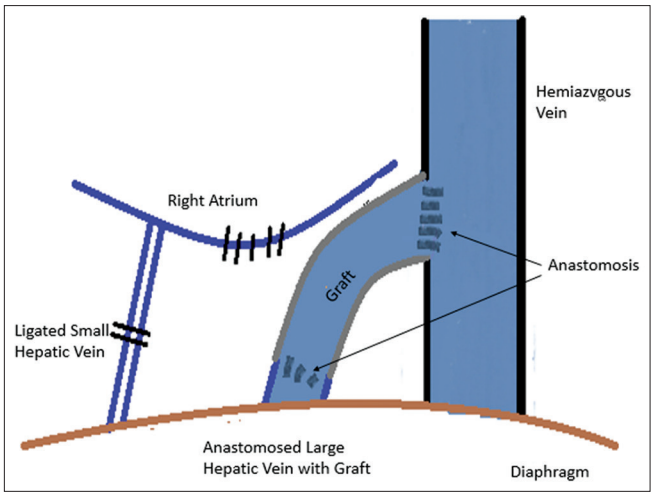

Figure 3: Schematic illustration of surgical procedure

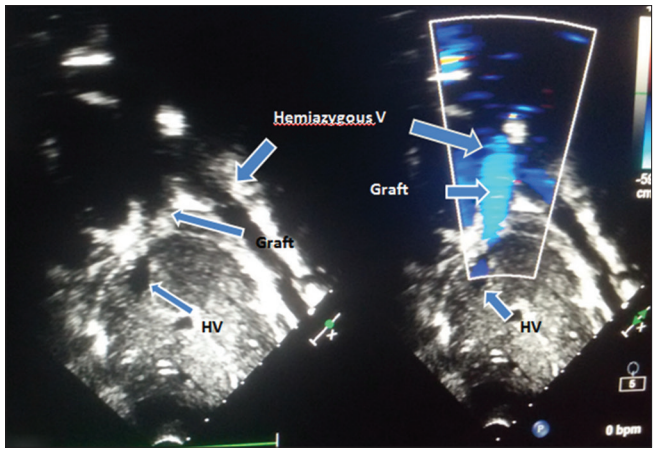

Figure 4: Postoperative ECHO pictures showing laminar flow in the conduit from hepatic (hepatic veins) to hemiazygos vein. HV: Hepatic veins

would then exit the right atrium (RA) and be connected to the RPA,[6] and (c) anastomosis between the HVs and the HAV rather then to the pulmonary arteries. Connecting the hepatic venous drainage to the pulmonary arteries has some inherent problems. The HVs may be multiple, and they may connect to the RA with a significant length of atrium between their separate sites of entry. Interposition of a tube between them and the pulmonary artery would necessitate a large diameter tube to accommodate all the veins. Having a disproportionately large tube would predispose to thrombosis, especially as the tube needs to be of sufficient length to reach the pulmonary arteries. Another method of connecting all veins to the tube would be to attach one of the veins separately onto the side of the tube. Here, a tube of smaller diameter could be selected but with the added risk of torsion of the implanted vein and anastomotic narrowing. Another problem with connecting to the pulmonary arteries is that the connection would be to ipsilateral pulmonary artery. In addition, as the ventricular mass was to the left, a standard extracardiac Fontan conduit would take a route to the previously stented RPA, a not so desirable option. This may not facilitate the resolution of AVMs in the contralateral lung.[4,7,8] The HV to HAV connection is short and therefore, less likely to suffer from thrombotic occlusion. In this case, as the superior vena cava was demonstrated to be draining to both the pulmonary arteries, having the HVs draining into the HAV eliminates the risk of unilateral perfusion.

The decision to ligate the smaller HV was taken because we believe that the size of a draining vein anywhere is a pointer to the size of the area being drained, and small size would indicate lesser importance or alternative channels of drainage for the particular area subtended. The likelihood of sacrificing some important issues such as tunnel lie, size, and construction, just to include a minor vein. prompted us to forgo attempts at including the smaller hepatic vein in the drainage to the hemiazygous connection. Leaving it draining in the RA would give rise to intrahepatic shunting and perpetuation of cyanosis. Hence, we decided to ligate it as we have done in the prior cases, once again without any consequence. If it was a reasonably-sized vein, it would have been included in the hepatopulmonary drainage.[9]

The need for anticoagulation following hepatic venous blood flow diversion remains the potential limitation to the procedure.[10,11]

HV to azygos/hemiazygos systems following Kawashima has been previously described.[12] Baskett *et al.* have described a similar method to ours where a cuff of RA containing the HV orifices was anastomosed directly to the azygos vein under total circulatory arrest.[13] Our concern regarding this method is that it would exert extra traction and thus, a risk of kinking and occlusion of one or both the HVs as in our case, both the HVs lay in different planes. In addition, excising a large atrial cuff could predispose to rhythm-related problems in the long run.

Kaneko *et al.* also described a similar method using direct posterior connection between atrial cuff and azygos vein, anteriorly augmented with autologous pericardium.[14] In our case, we felt an interposition graft would give the best lie for completion of the hepatoazygous connection.

CONCLUSION

The present experience highlights a method of connecting the hepatic venous egress to the pulmonary arterial circulation in patients with IVC interruption, which is both expedient and possibly meritorious secondary to the shorter length of the tunnel.

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Conflicts of interest

There are no conflicts of interest.

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