

A catheter-based interventional strategy redirects hepatic vein flows after Fontan procedure in left isomerism to treat severe hypoxemia

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ABSTRACT

Pulmonary arteriovenous malformations after the Kawashima procedure causing severe hypoxemia are treated by Fontan surgery that redirects hepatic venous blood to the pulmonary circulation. Alignment of the hepatic venous conduit toward both pulmonary arteries is crucial for their regression. Persistent hypoxemia due to nonregressed malformations in one lung often warrants a repeat surgery to redirect hepatic venous flows. Catheter-based redirection of Fontan flows is not commonly performed as metallic stents and devices may predispose to thrombus formation in the circuit. This report highlights a patient with persistent arteriovenous malformations in the left lung leading to severe disabling hypoxemia that was managed in the catheterization laboratory with an innovative strategy to redirect Fontan flows toward the affected left lung.

Keywords: Azygos continuation, extracardiac conduit Fontan surgery, heterotaxy, hypoxemia, left isomerism, redirection of Fontan flows

INTRODUCTION

Diversion of hepatic venous flow away from the pulmonary circulation is a key factor leading to pulmonary arteriovenous malformations (PAVMs) in patients with heterotaxy after a Kawashima procedure.^[1] Incorporation of the hepatic veins into the cavopulmonary circulation resolves these PAVM and improves hypoxemia.^[2] Ongoing search for the elusive hepatic factor has suggested a few candidate proteins such as Ang-1, CXCL 16, and LIF.^[3] Creating a surgical offset between superior vena cava (SVC) and conduit in a Fontan circuit reduces energy losses on computational models, but unfortunately leads to preferential hepatic vein flows toward one lung in post-Kawashima patients.^[4] Persistent hypoxemia in such

cases warrants additional surgeries to divert hepatic flows toward the affected lung.^[5] Individualized surgical plans designed from computational flow dynamic models may be needed after the Kawashima procedure, but are unfortunately not universally unavailable, especially in restricted economies.^[6] While revisions using a bifurcation “Y” graft to both pulmonary arteries from the hepatic veins or a conduit between hepatic vein and azygos vein are potential options, these repeat surgeries are morbid needing cardiopulmonary bypass.^[7] Extending the conduit superiorly toward the innominate vein without employing cardiopulmonary bypass is suggested as another way to divert hepatic veins toward the SVC.^[8,9] We report a patient who had resolution of right PAVM and persistence of left PAVM after a Fontan procedure managed with an extension conduit to the innominate vein to direct hepatic

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venous flows to the SVC. However, the extension conduit remained as a watershed zone and led to worsening PAVM on the left lung causing disabling hypoxemia.

CASE REPORT

A 5-year-old boy diagnosed to have heterotaxy, left isomerism, hepatic veins draining on the right side of the spine to the atrium, left azygos continuation of the inferior vena cava to a single left SVC, common atrioventricular valve, single ventricle, and valvar pulmonary atresia along with d-malposed dilated ascending aorta, underwent Kawashima procedure at 2 years of age resulting in improvement of oxygen saturations to 92% in room air. He developed progressive hypoxemia and oxygen saturation of 60% after 2 years. Rapid cardiac filling of agitated saline contrast injected from the arm veins suggested the development of PAVM [Figure 1]. After confirming the presence of bilateral PAVM on catheterization that demonstrated a mean pulmonary artery pressure of 10 mmHg across both pulmonary arteries, he underwent Fontan completion directing the hepatic veins to the right pulmonary artery (RPA) using a 16 mm conduit. The conduit could not be taken leftward closer to the previous cavopulmonary anastomosis to reduce the offset due to the dilated ascending aorta and need to cross across the midline. The same reasons precluded a “Y” graft too. The oxygen saturations worsened to 50%–60%, 6 months after the surgery with the persistent need for home oxygen and disabling symptoms. Lack of resolution of PAVM warranted a restudy that demonstrated a mean pressure of 14 mmHg in both pulmonary arteries and excluded any stenosis across the confluence. Selective

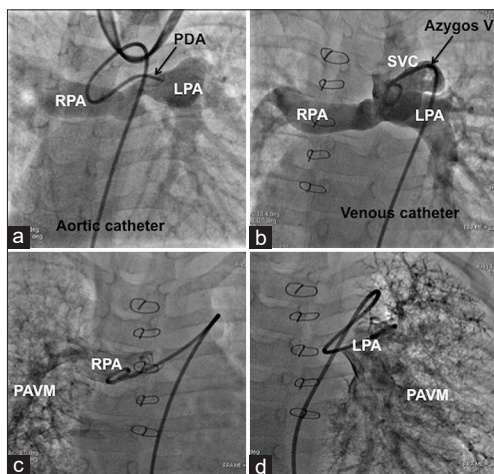


Figure 1: Angiogram (a) at 2 years of age through the arterial duct (patent ductus arteriosus) from the undersurface of aortic arch demonstrates adequate right pulmonary artery and left pulmonary artery. Two years following the Kawashima procedure, venous angiogram from the left azygos vein (b) demonstrates left superior vena cava connected to the pulmonary arteries. Selective injections in right pulmonary artery (c) and left pulmonary artery (d) shows extensive pulmonary arteriovenous malformations

distribution of hepatic venous blood to the right lung caused by a collision between the blood flows from the SVC and hepatic venous conduit at the confluence resulted in complete resolution of the right PAVM but worsened the PAVM on the left side explaining the hypoxemia [Figure 2]. Anastomosis of another 16 mm conduit extension from the anterior wall of the previous conduit to the right innominate vein was performed through a repeat sternotomy without cardiopulmonary bypass to direct hepatic vein flows toward the SVC without any relief of hypoxemia [Figure 3].

Hepatic vein angiogram showed preferential flows from the hepatic veins towards the right lung due to their anatomical proximity, rather than toward the innominate vein through the conduit extension [Video 1]. The extended conduit acted as a watershed zone with no significant flows through it. The sharp 90° angle between the conduit extension and the innominate vein could have been an additional contribution. The pressure in the entire Fontan circuit showed uniform mild elevation at 15 mmHg, possibly due to mild common atrioventricular valve regurgitation and hypoxia-induced subclinical ventricular dysfunction. As the conduit was 16 mm, the innominate vein that appeared smaller than the conduit was stented with a large 26 mm LD Max stent (Medtronic, Plymouth, MN, USA) using a 20 mm balloon to create a free flow of the hepatic venous blood towards the SVC [Figure 4]. We decided to close the flows towards the RPA as the small amount of hepatic venous return preferentially flowed to the RPA due to its proximity [Video 2]. The upper end of the hepatic vein conduit between the extension and the RPA was closed with a 18-16 Cera duct occluder (Lifetech Scientific, Shenzhen, PRC), ensuring that the device did

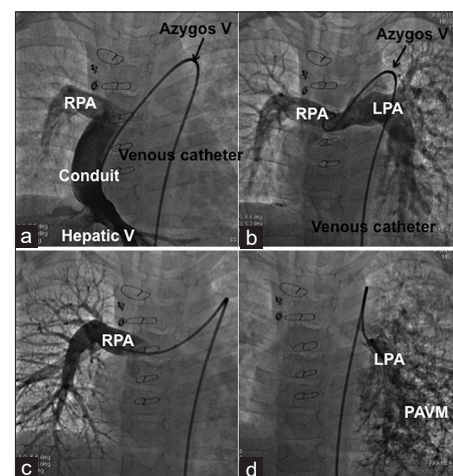


Figure 2: Angiogram in the Fontan conduit (a) from the left superior vena cava through the azygos vein shows preferential hepatic vein flows to the right pulmonary artery. Pulmonary angiogram (b) shows resolution of right and persistence of left pulmonary arteriovenous malformations. This is confirmed on selective right pulmonary artery (c) and left pulmonary artery angiography (d)

not protrude either into the RPA or into the 16 mm conduit extension [Figure 5]. The final hepatic vein angiogram showed a complete redirection of the flows towards the stented right innominate vein toward the left SVC [Videos 3 and 4]. He was discharged from the hospital on optimal doses of warfarin and aspirin. A rapid improvement of the hypoxemia followed this procedure

and oxygen saturation improved to 90% in room air after 1 month. However, the child succumbed after 2 months to viral pneumonia despite demonstration of patency of the conduit and stents during the terminal admission.

DISCUSSION

While an offset between SVC and conduit during Fontan surgery is desirable to reduce energy loss, avoiding an offset between the hepatic venous conduit to the SVC that drains the azygos vein is crucial after the Kawashima procedure to distribute blood evenly to both lungs.^[5] Persistent unilateral PAVM due to diversion of the hepatic venous blood created by such an offset was managed in one instance using a large bare stent between the SVC and the inferior conduit to realign and remove the offset.^[10] In our patient, we could not achieve an alignment between the SVC and hepatic vein conduit as the two vessels were far apart with an intervening dilated ascending aorta.

Extending the hepatic vein conduit to the innominate vein redirects the blood flow towards the SVC.^[8,9] This is not applicable as a primary solution for all patients as a very long conduit from hepatic veins to the innominate vein invites thrombus formation. Hypoxemia did not improve as the extended conduit acted as a watershed zone between the hepatic vein flows to the right lung below and the right innominate vein flows above. Stent angioplasty of the innominate vein to allow the additional hepatic venous flows along with occlusion of the initial

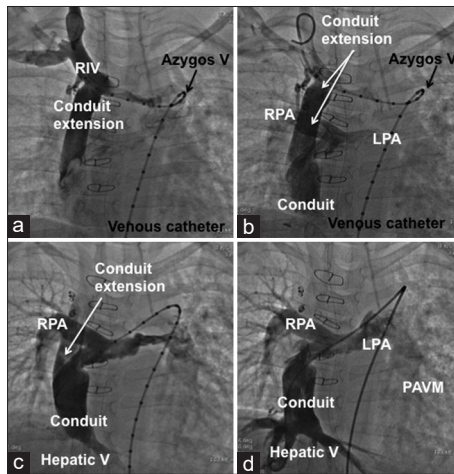


Figure 3: Right innominate vein angiography (a) shows reflux of contrast inferiorly into the extension of the conduit. The contrast reflux in the extended conduit later fills the lower hepatic vein conduit (b) and faintly opacifies the right pulmonary artery. Angiogram into the lower conduit (c) shows hepatic venous blood still preferentially flowing towards the right pulmonary artery. This resulted in the persistence of malformations (pulmonary arteriovenous malformation) in the left lung (d)

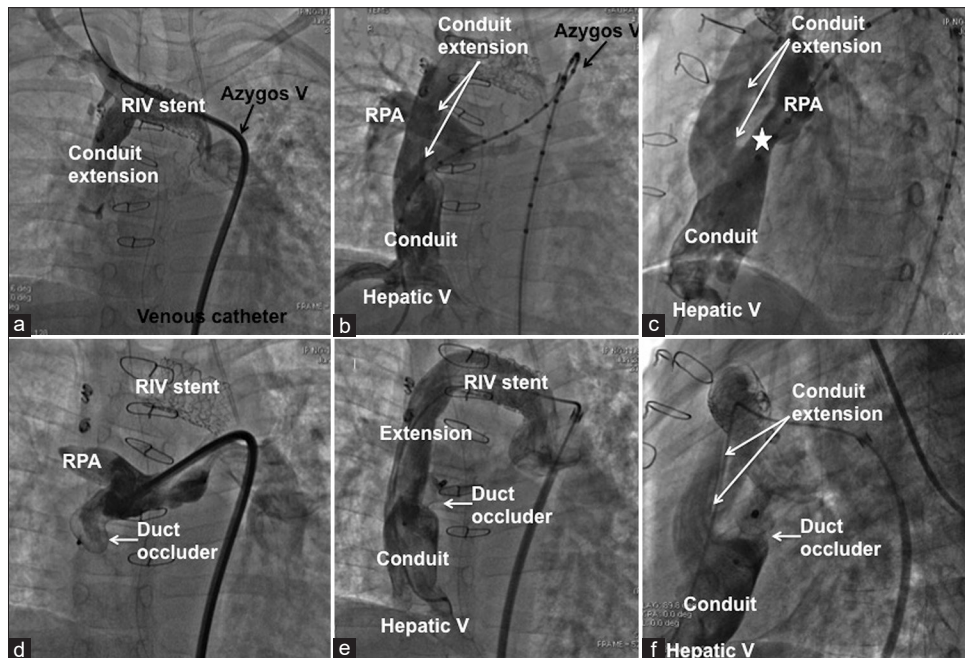


Figure 4: After stent angioplasty of the right innominate vein, angiogram (a) shows persistent reflux of contrast into the conduit extension rather than flowing upwards towards the superior vena cava. Hepatic vein angiogram in anteroposterior (b) and steep left anterior oblique projection (c) shows contrast flow into the right pulmonary artery rather than flowing into the superior conduit extension. After identifying the site of occlusion (*), a duct occluder was positioned in the distal conduit (d). Angiogram in hepatic veins in anteroposterior (e) and steep left anterior oblique projection (f) redirection of hepatic veins towards the conduit extension and superior vena cava

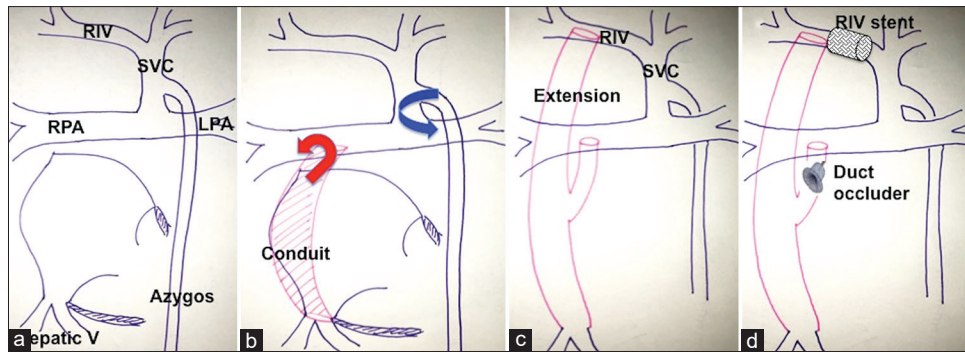


Figure 5: Cartoons explain the circulation after the first surgery-Kawashima procedure (a), second operation-Fontan surgery (b), third operation- conduit extension towards the right innominate vein without cardiopulmonary bypass (c) and after the closure of distal conduit with a duct occluder (d) and stenting of right innominate vein to redirect hepatic veins towards the superior vena cava

conduit near the RPA resulted in a redirection of flows towards the left lung. Catheter interventions to redirect hepatic venous flows in persistent PAVM were achieved by deploying transhepatic and transjugular multiple bare and covered stents within the Fontan conduit in three patients, but the authors cautioned about the heightened possibility of thrombosis due to the presence of 3-4 stents in the sluggishly flowing circuit.^[10] Despite minimal use of devices and stents within the Fontan circuit in our patient, meticulous monitored oral anticoagulation and aspirin are mandatory to avoid catastrophic thrombus formation.

CONCLUSIONS

Persistent PAVMs after Fontan surgery in patients with heterotaxy caused by the direction of hepatic venous flows towards one lung pose challenges for management. Careful understanding of the flow directions sometimes guided by computational imaging guides redirection of hepatic venous flows to the affected lung. Catheter intervention may play a role in such redirection in selected challenging patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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