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Fontan Revision with Y-Graft in a Patient with Unilateral Pulmonary Arteriovenous Malformation

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The extracardiac conduit Fontan procedure is the last surgical step in the treatment of patients with a functional single ventricle. An acquired pulmonary arteriovenous malformation may appear perioperatively or postoperatively due to an uneven hepatic flow distribution. Here we report a case of a bifurcated Y-graft Fontan operation in a 15-year-old male patient with a unilateral pulmonary arteriovenous malformation after an extracardiac conduit Fontan operation.

Key words: 1. Congenital heart defects
2. Fontan
3. Conduits
4. Vascular disease
5. Y-graft

Case report

A 15-year-old male patient with a history of complete atrioventricular septal defect, pulmonary atresia, right isomerism, asplenia, and separate hepatic venous drainage was treated with a left modified Blalock-Taussig shunt at 50 days after birth, a bidirectional cavopulmonary shunt at 21 months, and an external cardiac conduit Fontan operation at 5 years of age. His oxygen saturation was maintained at approximately 93%–95% after the Fontan procedure. During follow-up, mild desaturation at about 90%–93% was observed. In the cardiac catheterization, there were no anomalous findings regarding ventricular function or systemic venous pressure; however, preferential flow was observed. Blood from the superior vena cava showed a tendency to flow toward the right lung, while blood from the inferior vena cava (IVC) showed a tendency to flow toward the left lung. The right lung had a diffuse pulmonary arteriovenous malformation (pAVM) (Fig. 1). A lung perfusion scan (LPS) was performed by a 99mTc albumin-aggregated (99mTc-MAA) injection in the lower limb and the shunt fraction was reported as 15%.

Given the diagnosis of a unilateral pAVM, a Fontan revision was planned. A bifurcated Y-graft was used for the redistribution of the hepatic blood flow. Under general anesthesia, a redo median sternotomy and adhesiolysis were performed. After aortic/bicaval cannulation, the operation was performed under cardiopulmonary bypass. In the operative findings, the previous Fontan graft was offset to the left side, and

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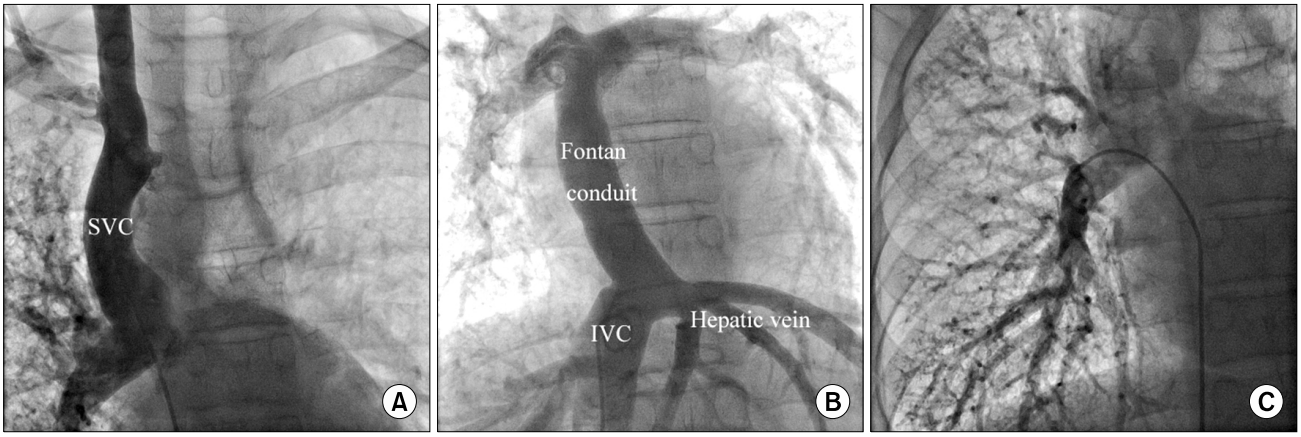

Fig. 1. A selective angiogram showing that (A) the right lung received almost all blood flow from the SVC, (B) while the blood from the IVC mainly flowed to the left lung. (C) The right lung displays a reticulonodular density representing a diffuse pulmonary arteriovenous malformation. SVC, superior vena cava; IVC, inferior vena cava.

the previous bidirectional cavopulmonary shunt site was very wide. The previous Fontan conduit was removed, and a 22×11×11-mm Y-graft conduit was connected to the left pulmonary artery and the right pulmonary artery lower branch. The IVC side was connected to the previous remnant graft. At the previous anastomosis site of the graft to the pulmonary artery, a direct closure and angioplasty using GORE-TEX Vascular Grafts (W. L. Gore & Associates Inc., Newark, NJ, USA) had been performed (Fig. 2). The total cardiopulmonary bypass time was 100 minutes.

The patient was transferred to the general ward on postoperative day 1 and discharged on postoperative day 8 without any complications. In the postoperative LPS, performed by using a right foot vein, the shunt fraction was found to have increased by 20% as compared to the preoperative LPS. Three months after surgery, the patient underwent a magnetic resonance imaging (MRI) scan of the heart, and oxygen saturation was still 88%. In the MRI quantification, the total blood flow per stroke volume of the Y-graft was 25.9 mL. The blood flow volumes in the right and left limbs of the Y-graft were 14.0 mL (54%) and 11.9 mL (46%), respectively. Considering that this patient had a preoperative preferential flow of the Fontan graft to the left lung, greater IVC flow, including hepatic venous return, was directed to the right lung postoperatively. One year after the Fontan revision, LPS was performed again. It showed an increased blood flow to the right lung and oxygen saturation amounting to 95%, considerably greater than the results of the previous test (Table 1).

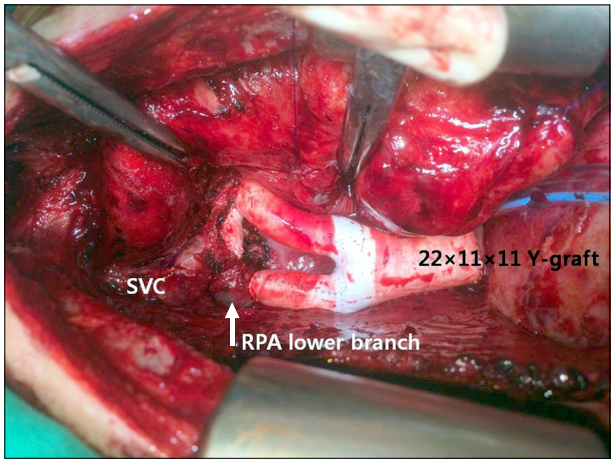

Fig. 2. The Y-graft conduit connected to the left pulmonary artery and the lower branch of the RPA (arrow). SVC, superior vena cava; RPA, right pulmonary artery.

Discussion

There are various factors that cause pAVMs, but these do not account for the details of how pAVMs are formed. A congenital heart defect, such as a left isomerism or exclusion of the hepatic vein blood flow to the pulmonary circulation, could be a risk factor for a pAVM. Even after the completion of the Fontan operation in patients with a functional single ventricle (FSV), an unbalanced distribution of the hepatic flow might result in a pAVM.

Table 1. Changes in preoperative and postoperative blood flow, shunt fraction, and oxygen saturation

	Right	Left	Shunt fraction	Peripheral oxygen saturation
Preoperative LPS	30.6%	69.4%	15%	91%
Postoperative LPS	34.4%	65.6%	20%	85%
Y graft flow in postoperative heart MRI 3 months after operation[a)]	14.0 mL (54%)	11.9 mL (46%)		88%
LPS 1 year after operation	37.1%	62.9%	20%	95%

LPS, lung perfusion scan, using Technetium 99mTc albumin aggregated injection to the lower limb; MRI, magnetic resonance imaging.
[a)]Blood flow per stroke volume.

It has not been clearly established how lung angiogenesis is inhibited. The precursors of angiotensin II, an agonist of angiogenesis, and endostatin, an antagonist of angiogenesis, are known to be produced primarily in the liver. For instance, in a review of the occurrence and mechanisms of pAVMs, Hoffman [1] showed that, in the heart systems of FSV patients, unlike in the normal heart system, the lungs are exposed to a high concentration of angiotensin II. Furthermore, Field-Ridley et al. [2] found that collagen XVIII, a precursor of endostatin, is degraded in the lungs. In FSV patients, the collagen XVIII levels increase and the endostatin levels decrease, resulting in angiogenesis, since the pulmonary circulatory system inhibits endostatin production from collagen XVIII.

It is essential to plan a surgical strategy to achieve a better distribution of the hepatic flow. When a pAVM is diagnosed in the pre-Fontan state, most cases of pAVMs regress after Fontan completion with hepatic vein inclusion [3]. In this case, to avoid complications, we performed the first Fontan procedure with a separate drainage of the hepatic vein connecting to the IVC stump. Nevertheless, a pAVM occurred and it was necessary for us to devise ways to provide a more even and effectively distributed hepatic flow. Imoto et al. [4] reported good results from treating a pAVM by changing the position of the Fontan conduit. In addition, McElhinney et al. [5] reported that only a direct hepatic vein–azygous vein connection may provide the most reliable mixing and bilateral distribution of hepatic venous blood in patients with heterotaxy.

The case reported here was somewhat different; thus, we considered using a Y-graft conduit to redesign the Fontan pathway. Kanter et al. [6] reported largely successful results from the Fontan operation using a Y-graft. Likewise, some patients who underwent a Y-graft Fontan revision showed increased oxygen saturation at discharge. For example, Haggerty et al. [7] reported positive hemodynamic results in patients who underwent a Y-graft Fontan operation using computed tomography and MRI in a quantitative simulation. Yang et al. [8] also found that asymmetric blood flow distribution was reduced after the Y-graft Fontan operation.

In the reported case, oxygen saturation, in terms of clinical improvement, did not increase immediately after the surgery. Rather, the shunt fraction increased to 20% in the postoperative LPS. However, this may have reflected the increased flow from the IVC to the right lung after the Y-graft Fontan operation. Considering that pAVMs will gradually disappear, it is evident that this reflects the increasing flow into the right pulmonary artery, which increases flow via the pAVM and causes a consequent increase in the shunt ratio.

In this case, all of the preoperative and postoperative LPSs used a lower extremity vein to inject the radionuclide (99mTc-MAA), thus showing the regional uptake ratio in each lung from the Fontan graft flow. The serial uptake ratio of radionuclide in the right lung increased over time from 30.6% preoperatively and 34.4% immediately postoperatively to 37.1% at 1 year after the operation. In the presence of a unilateral pAVM, this ratio represents not the perfusion ratio, but the captured radionuclide in the pulmonary capillaries of each lung. Therefore, the serial uptake ratio in the right lung was below 50%, but gradually increased, even though, in the postoperative MRI flow measurement, flow in the right limb of the Y-graft (14.0 mL, 54%) was greater than in the left limb (11.9 mL, 46%). This means that the captured radionuclide in the right lung increased over time and the pAVM started to gradually disappear. However, a residual pAVM in the right lung still needs to be considered. This is the reason why the oxygen saturation improved to 95% while the shunt fraction was 20% at 1 year postoperatively.

Therefore, much more time is needed to show radionuclide uptake over 50% in the right lung.

As in other reported cases, oxygen saturation improved over time in our patient. Thus, the Y-graft Fontan operation may be a promising option for reducing the risk of pAVMs by solving the problem of unbalanced distribution in the hepatic flow.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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