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Staged Kawashima operation with cavopulmonary connection

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We herein present our technique of staging the Kawashima procedure in a child with single ventricle and interruption of the inferior vena cava (IVC) with fair-sized pulmonary arteries by creating a bidirectional cavopulmonary anastomosis first, followed by Fontan completion as the second stage.

CLINICAL SUMMARY

A 3.5-year-old girl weighing 10 kg with tricuspid atresia, ventricular septal defect, and pulmonary stenosis with fair-sized pulmonary arteries was taken for bilateral bidirectional cavopulmonary anastomosis on the basis of echo-

cardiography in 2001. At surgery, absent right superior vena cava (SVC) and interrupted IVC with hemiazygos continuation to a large left SVC and joining of hepatic veins to the right atrium were noted. On cardiopulmonary bypass, the left SVC was divided above the hemiazygos vein and anastomosed to the left pulmonary artery as a left bidirectional Glenn shunt, leaving the IVC drainage unchanged. Because the pulmonary artery mean pressure was 18 mm Hg, the pulmonary trunk was banded to allow a limited antegrade flow (Figure 1, A). She made a good recovery but was lost to follow-up after 2 years.

She returned at the age of 13 years (2010) with progressive effort dyspnea and worsening cyanosis (arterial oxygen saturation, 72%). A cardiac catheterization showed a well-flowing left-sided Glenn shunt and IVC drainage via the hemiazygos vein into the coronary sinus through the proximal left SVC (Figure 2). There were no pulmonary arteriovenous malformations or venovenous collaterals.

During Fontan completion, the hemiazygos vein was disconnected from the left SVC stump and anastomosed end to side to the left pulmonary artery, completing the Kawashima procedure. The hepatic veins were connected to the right pulmonary artery with an 18-mm

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FIGURE 1. A and B, Kawashima completion by direct hemiazygos to left pulmonary artery connection and extracardiac hepatopulmonary connection to the right pulmonary artery completing total cavopulmonary connection. The pulmonary trunk was interrupted. IVC, Inferior vena cava; LSVC, left superior vena cava; RPA, right pulmonary artery; LPA, left pulmonary artery; HEP. VEIN, hepatic vein.

FIGURE 2. Angiogram showing a well-flowing left-sided Glenn shunt and inferior vena cava draining through the proximal left superior vena cava to the coronary sinus.

polytetrafluoroethylene tube extracardiac conduit (Gore-Tex; W. L. Gore & Associates, Inc, Flagstaff, Ariz) with a 4-mm fenestration, thus completing the Fontan circuit. The main pulmonary artery was interrupted (Figure 1, B). Her postoperative recovery was uneventful.

At 1 year's follow-up, she was asymptomatic with an arterial oxygen saturation of 96% on oral anticoagulation; echocardiography showed good Fontan flows from the left SVC, hemiazygos vein, and hepatic veins. The fenestration could not be demonstrated.

DISCUSSION

Although the classic Kawashima operation needs a second-stage procedure for hepatic vein inclusion,¹ single-stage cavopulmonary connection with hepatic vein inclusion has been reported with good results.² In Infants with less satisfactory Fontan criteria, a high-risk Kawashima procedure with fenestration has also shown good results.³

In our patient, who had fair-sized pulmonary arteries, we did a simple staging of a high-risk Kawashima procedure by doing only the superior cavopulmonary anastomosis, leaving the IVC drainage intact. We kept the antegrade flow open through a pulmonary artery band to prevent the development of pulmonary arteriovenous malformations.

Even after 10 years our patient did not have pulmonary arteriovenous malformations owing to the limited antegrade flow.⁴ She did not have venovenous malformations (between the hemiazygos and hepatic veins) because the hemiazygos system was not pressurized as in a high-risk

Kawashima procedure, in which it would be draining to the pulmonary circulation.⁵ At the time of the second-stage procedure, the hemiazygos vein could be connected to the left pulmonary artery without much technical difficulty.

To the best of our knowledge, the division of the SVC above the azygos or hemiazygos entry into the SVC to create a bidirectional Glenn shunt, first with controlled antegrade blood flow and then by a second-stage Fontan completion, has not been reported so far in the setting of Kawashima procedure. This technique allows the surgeon to plan a stepwise approach in Kawashima patients who need palliation early, avoiding pulmonary vascular overload, especially in young infants whose pulmonary vasculature may not yet be ready to passively accept the majority of systemic venous flow.

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