Cardiol Young 2008; 18: 641–643
© Cambridge University Press
ISSN 1047-9511
doi:10.1017/S1047951108003156
First published online 7 October 2008

Brief Report

Creation of an intraatrial tunnel to produce a total cavo-pulmonary connection in a patient with dominant right ventricle, totally anomalous pulmonary venous connection, and pulmonary atresia with non-confluent pulmonary arteries

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Abstract The surgical strategy for patients having a functionally single ventricle associated with totally anomalous pulmonary venous connection and pulmonary atresia with non-confluent pulmonary artery has yet to be agreed. We created an intraatrial tunnel to produce a total cavo-pulmonary connection in such a patient, also creating a confluence for the pulmonary arteries. By minimizing the use of the GoreTex patch, the patient was able to discontinue the use of warfarin.

Keywords: Fontan circulation; functionally univentricular heart; double inlet ventricle

Since the first operation to treat tricuspid atresia by creation of the Fontan circulation was reported in 1971,[1] the surgical technique has been modified in various ways, with positive results. There are few reports, however, describing repairing of patients with functionally univentricular hearts associated with totally anomalous pulmonary venous connection and pulmonary atresia with non-confluent pulmonary arteries.[2,3] We describe here our approach to such a patient.

Case report

A cyanotic girl was referred to Chiba Children's Hospital shortly after birth, and was diagnosed with having a dominant right ventricle, totally anomalous pulmonary venous connection to the right superior caval vein, pulmonary atresia, bilaterally patent arterial ducts supplying non-confluent pulmonary arteries, and bilateral superior caval veins. She was only a month old when she underwent a left modified Blalock-Taussig shunt, followed by a stenting of the right-sided patent duct when she was two months old. When she was 15-months-old, she underwent a bilateral bidirectional Glenn procedure and pulmonary arterial plasty, the latter creating a confluent central pulmonary artery. The totally anomalous pulmonary venous connection was also repaired, the pulmonary venous orifice being augmented by cutback (Fig. 1). Concomitantly, the primary atrial septum was detached at its posterior attachment and shifted to the right in order to create a baffle separating the flow through the inferior caval vein from that entering through the pulmonary vein. The proximal stump of the left superior caval vein was closed with a GoreTex (Gore Medical, Flagstaff, AZ) patch. In the interim, a balloon angioplasty was used to relieve the stenoses at the centre of the anastomosed pulmonary artery and at the portion of the left Glenn shunt.

When she was two years old, catheterization study revealed a right ventricular end-diastolic volume of 142% of normal, right ventricular

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Accepted for publication 12 August 2008

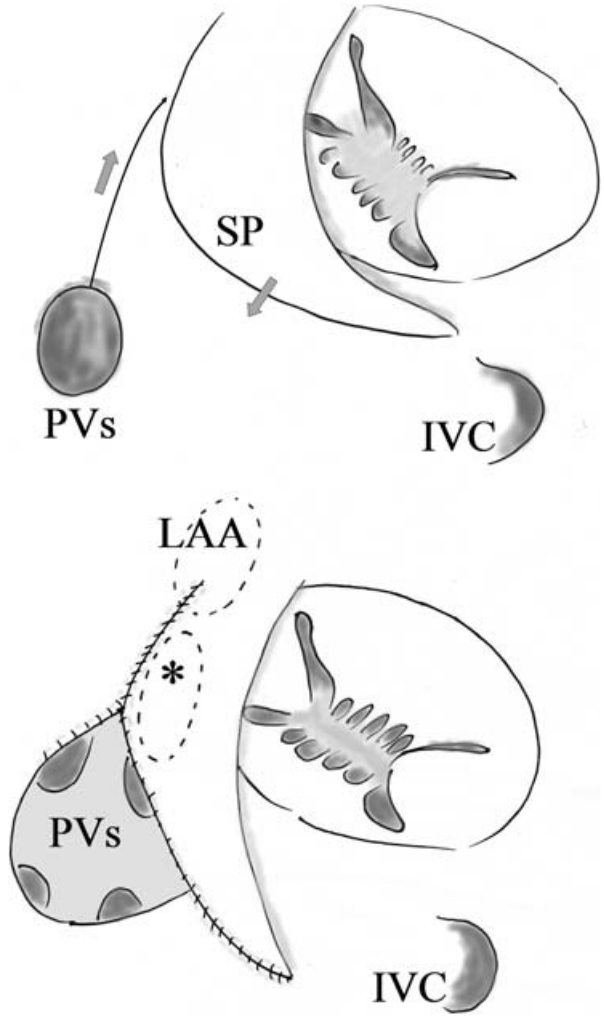

Figure 1.
The intraatrial procedure at the time of construction of the Glenn shunt. The pulmonary venous orifice was augmented by a cutback. The primary septum was detached and sutured anteriorly to the inferior caval venous orifice. Abbreviations: SP: primary septum, PVs: pulmonary veins, IVC: inferior caval vein, LAA: left-sided atrial appendage, asterisk: route to the pulmonary arteries.

ejection fraction of 51.5%, mean pulmonary arterial pressure of 10 mmHg, and pulmonary vascular resistance of 1.3 Woods Unit with a Nakata index of 140 mm^2/m^2.[4] Thus, a total cavo-pulmonary connection was planned.

A re-sternotomy was made, and cardiopulmonary bypass was achieved using the ascending aortic and the bilateral superior and the inferior caval veins. Using ventricular fibrillation, an incision was made to the right-sided atrial appendage parallel to the atrioventricular groove. The GoreTex patch at the roof of the atrium was removed. An incision was made to the lower aspect of the reconstructed central pulmonary artery, and anastomosed to the opening of the atrial roof. Subsequently, an intraatrial tunnel was

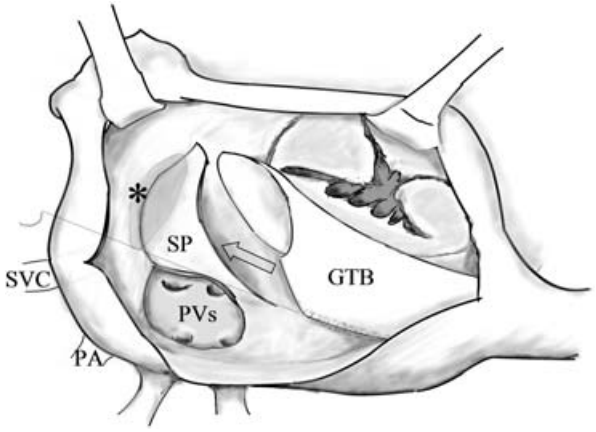

Figure 2.
The final route of the intraatrial total cavo-pulmonary connection created with a half circumferential GoreTex graft. The arrow indicates the flow from the inferior caval vein through the baffle. Abbreviations: PVs: pulmonary veins, SP: primary septum, SVC: superior caval vein, PA: pulmonary artery, GTB: GoreTex baffle, asterisk: route for the pulmonary arteries.

created to channel the inferior caval venous flow to the pulmonary arterial anastomosis site using an open GoreTex graft attached with a 5-0 continuous suture. The previously shifted primary atrial septum was used as a part of the route to reduce the usage of an artificial graft (Fig. 2). A 3.5 mm fenestration was punched out on the GoreTex baffle. Weaning from cardiopulmonary bypass was uneventful. Before the discharge, a Holter electrocardiogram did not show either any significant arrhythmia or heart block.

Postoperative catheterization revealed well balanced bilateral pulmonary arterial flow from both caval veins, with mean peripheral pulmonary arterial pressure of 11 mmHg and adequate right ventricular ejection fraction. The flow from the pulmonary veins to the atrioventricular valve was smooth without any obstruction. A veno-venous collateral channel running from the superior caval vein to the right-sided atrium was coiled. We were able to discontinue warfarin 3 months postoperatively, so now she is taking only aspirin and diuretics. Her recent saturation of oxygen as measured at the outpatient clinic was 94% in room air.

Discussion

Although many variations in the techniques used to create the Fontan circulation have contributed to improve surgical outcomes, the surgical strategy for patients with functionally single ventricles accompanied with totally anomalous pulmonary venous connection, pulmonary atresia, and non-confluent pulmonary artery remains inconclusive. Major concerns are, first, if balanced pulmonary

https://doi.org/10.1017/S1047951108003156 Published online by Cambridge University Press

arterial flow can be created bilaterally, and second, creation of a route for the total cavo-pulmonary connection from the inferior caval vein to the bilateral pulmonary arteries so as not to interfere with the pulmonary venous flow. A successful total cavo-pulmonary connection in almost this anatomy has been reported[2] using an extracardiac total cavo-pulmonary connection. In our case, because the pulmonary venous connection was to the right superior caval vein, we chose to stent a patent arterial duct rather than create a Blalock-Taussig shunt. Restenosis after construction of the confluence of the pulmonary artery is common.[5] Our strategy of replacing the Blalock-Taussig shunt with a stented duct proved successful in delaying the timing of construction of the confluence of the pulmonary arteries. In creating the total cavo-pulmonary connection, we used the primary atrial septum so as to reduce the use of artificial material. In consequence, about two-thirds of the circumference of the intracardiac lateral tunnel route was surrounded by the native tissues, so we were able to discontinue warfarin. The intraatrial tunnel connected to the centre of the newly created central pulmonary artery produced well-balanced flow to both pulmonary arteries. There was neither heart block nor any significant arrhythmia.

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