Fontan completion in a patient with previous liver transplantation

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Abstract

We present the first case of a successful Fontan completion in a patient with previous liver transplantation. An infant with polysplenia syndrome with a functional single ventricle and biliary atresia had been surgically managed by pulmonary artery banding, Kasai operation and living donor liver transplantation. Subsequently, the patient successfully underwent bidirectional cavopulmonary shunt and total cavopulmonary connection with extracardiac conduit at 3 and 5 years of age, respectively.

Keywords: Congenital heart disease · Fontan · Liver · Abdominal organs

INTRODUCTION

Many patients with polysplenia syndrome have a single functional ventricle, for which Fontan track management is the gold standard. In addition, patients with polysplenia syndrome may have end-stage liver disease with biliary atresia [1], which is most often an indication for liver transplantation [2]. We here present the case of successful Fontan completion in a boy with polysplenia syndrome with a functional single ventricle and biliary atresia and who had previously undergone liver transplantation.

CASE REPORT

The patient had been detected with heterotaxy syndrome (polysplenia variety), with a midline liver, a right upper quadrant stomach and pancreatic head (dextrogastria), bilateral bilobed lungs, levocardia, unbalanced complete atrioventricular canal defect with a diminutive left ventricle, subaortic ventricular outflow tract stenosis, a common atrium receiving pulmonary venous and coronary sinus flow, bilateral superior vena cava (SVC), azygous vein continuation, a right-sided aortic arch, aortic coarctation and two separate hepatic veins draining directly into the atrium (Fig. 1, left).

The patient was born after 39 gestational weeks (birth weight, 2932 kg), and had severe congestive heart failure, which was successfully managed with pulmonary artery banding at 12 days of age and catheter balloon aortoplasty at 18 days of age. However, he had persistent neonatal jaundice even after undergoing Kasai operation at 45 days of age. At 8 months of age, he underwent living donor liver transplantation. During surgery, an over-sized donor liver (112% of the normal size) was placed in the left upper quadrant of the abdominal cavity, and only the left-sided hepatic vein was utilized for reconstruction of the donor liver. The patient’s total bilirubin level reached normal levels thereafter, and he was administered a tacrolimus-based immunosuppression regimen. The pulse oximetry oxygen saturation (SpO2) gradually decreased to 70–75% by 3 years of age, when a cardiac catheterization study showed a mean pulmonary arterial pressure of 14 mmHg and pulmonary arterial resistance of 1.73 unit/m².

At 3 years of age, he underwent total cavopulmonary shunt (Kawashima operation) and Damus–Kaye–Stansel anastomosis with cardiopulmonary bypass. During the 22-month postoperative period, the SpO2 decreased progressively from 90 to 75%. Subsequently, the donor liver index volume decreased to 87% of the normal value [3]. Repeated cardiac catheterization indicated a mean pulmonary arterial pressure of 9 mmHg and a pulmonary arterial resistance of 1.0 unit/m². The development of pulmonary arteriovenous malformations was not visualized.

At 5 years of age (body weight, 16.4 kg), he underwent Fontan completion with extracardiac conduit total cavopulmonary connection using a 12-mm ringed expanded polytetrafluoroethylene tube graft. During the operation, cardiopulmonary bypass with deep hypothermic circulatory arrest was performed to avoid a direct cannulation to the hepatic vein, which was individually connected to the atrium and associated with a previous end-to-end anastomosis at the liver transplantation. Only the left-sided hepatic vein was reconstructed with an end-to-end anastomosis to the tube graft, and the right-sided vein was doubly ligated and divided. The other end of the conduit was connected to the pulmonary artery exactly below the previous SVC anastomosis site (Figs 1, right and 2). The postoperative period was uneventful, as represented by peak lactate and bilirubin levels of 2.6 mmol/ml and 1.3 mg/dl, respectively, except with pleural effusion (total volume, 360 ml) on the left side that was aspirated during the first 2 days.
4 days. Cardiac catheterization 3 months after Fontan completion showed arterial oxygen saturation of 91.3%, and a Fontan conduit pressure of 13 mmHg. The patient was asymptomatic for 6 months after Fontan completion.

DISCUSSION

To our knowledge, this is the first report of a Fontan completion performed in a patient who had previously undergone liver transplantation. Thus far, open-heart surgery after liver transplantation has been limited to biventricular repair [3, 4], such as ventricular septal defect repair and Ross procedure.

In this case, a Kawashima operation and Fontan completion were indicated as life-saving surgeries. However, the transplanted liver, has several concerns on the prognosis. Late liver complications after the Fontan procedure [5] as well as late liver problems in children with polysplenia syndrome are not uncommon, but our follow-up is limited to only 6 months. The allograft should be subject to several different additional insults after transplantation, including hypoxia before Fontan completion, two cardiac operative insults using cardiopulmonary bypass with or without circulatory arrest, and high venous and portal pressures as well as low arterial perfusion after Fontan completion. All of these could lead to hepatic fibrosis. Hepatopulmonary syndrome, a triad of liver dysfunction, hypoxaemia and intrapulmonary vascular dilatation, can occur. Although the 12-mm Fontan conduit was selected based on the angiogram showing the 8-mm hepatic venous diameter of the transplanted liver, the optimal conduit size to minimize the incidence of thrombosis when connected to only hepatic vein(s) has not been determined. Therefore, a more meticulous follow-up from both cardiac and transplantation medicine perspectives is mandatory. Nevertheless, this case provides evidence that the Fontan completion could be effectively performed with a multidisciplinary approach even in a highly complex scenario.

Conflict of interest: none declared.
REFERENCES


