

Case report

Heart transplant in an adult patient with dysfunctional Fontan circulation at a high-complexity cardiovascular center in Peru: A case report

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ABSTRACT

Fontan circulation failure represents a progressive condition associated with multiorgan dysfunction and increased morbidity and mortality. In this context, heart transplantation has become the definitive treatment for this subgroup of patients. Reported survival rates after heart transplantation in patients with Fontan circulation are approximately 80% at 1 year, 74% at 5 years, and 59% at 10 years. We present the case of a 46-year-old patient with a history of Ebstein anomaly who underwent intracardiac Fontan surgery, which progressed to progressive Fontan circulation failure, and who underwent orthotopic heart transplantation with reconstruction of the pulmonary branches.

Keywords: Heart Transplantation; Fontan Procedure; Pulmonary Artery (Source: MeSH-NLM).

RESUMEN

Trasplante cardíaco en paciente adulto con circulación de Fontan disfuncionante en un centro de alta complejidad cardiovascular en el Perú: reporte de caso

La insuficiencia de la circulación de Fontan es una condición progresiva, asociada a disfunción multiorgánica y elevada morbimortalidad. En este contexto, el trasplante cardíaco se ha consolidado como el tratamiento definitivo para este subgrupo de pacientes. Las tasas de supervivencia posteriores al trasplante cardíaco en pacientes con circulación de Fontan se sitúan en aproximadamente el 80% al año, 74% a los cinco años y 59% a los 10 años. Presentamos el caso de un paciente de 46 años con antecedente de anomalía de Ebstein, sometido a cirugía de Fontan intracardiaca, que evolucionó con insuficiencia progresiva de la circulación de Fontan, siendo sometido a trasplante cardíaco ortotópico con reconstrucción de las ramas pulmonares.

Palabras clave: Trasplante de Corazón; Procedimiento de Fontan; Arteria Pulmonar (Fuente: DeCS-BIREME).

Introduction

Currently, 5-10% of patients with Fontan circulation die or require heart transplantation during childhood, and approximately 50% experience the same outcome by the age of 40 years ⁽¹⁾. Although overall survival at 20 years after Fontan surgery reaches 90%, 20-30% of patients develop Fontan circulatory failure. Heart transplantation is the definitive treatment for this subgroup. Right ventricular dominance has been shown to be an independent risk factor for Fontan circulatory failure ⁽²⁾. Patients with a failing Fontan circulation require comprehensive assessment by a multidisciplinary team with expertise in adult congenital heart disease, mechanical circulatory support, and heart transplantation ^(3,4). However, 40% are referred late for transplant evaluation, adversely affecting clinical outcomes and post-transplant survival ⁽³⁾.

Published experience with heart transplantation in patients with Fontan circulation in Latin America remains limited. A case involving an 18-year-old patient in Argentina was recently reported, representing one of the few published cases from the region ⁽⁵⁾. However, the challenges associated with surgical planning, vascular reconstruction, and perioperative management in this complex patient population remain poorly described. We present the case of an adult with a failing Fontan circulation who underwent orthotopic heart transplantation, highlighting the technical and surgical aspects of the procedure.

Case report

A 46-year-old man with Ebstein's anomaly had undergone a fenestrated intracardiac Fontan procedure in 1998, at the age of 19 years, in Boston, United States. He subsequently underwent embolisation of systemic-to-pulmonary collateral vessels in 2002, 2005, and 2016.

The patient reported dyspnoea on moderate exertion, with no angina or syncopal episodes over the preceding 10 years. In November, 2025, he was hospitalised with cardiorenal syndrome and required placement of a high-flow haemodialysis catheter for thrice-weekly haemodialysis. His comorbidities included persistent atrial fibrillation, liver cirrhosis, hypothyroidism, and progressive Fontan circulatory failure.

Pretransplant assessment

Cardiac catheterisation in June, 2025, showed a left pulmonary artery diameter of 12.4 mm (Z score: -1.6) and a right pulmonary artery diameter of 15.1 mm (Z score: -0.8), with no evidence of hypoplasia. The Z score indicates the deviation of the pulmonary artery diameter from the expected value for body surface area and age.

Systemic venous pressures were elevated, with pressures of 24 mmHg in both the superior and inferior venae cavae. Pulmonary vascular resistance was 2.2 Wood units, the pulmonary-to-systemic vascular resistance ratio was 0.27, and the pulmonary-to-systemic blood flow ratio (Qp/Qs) was 0.76. Left ventricular pressure was 103/15 mmHg. Echocardiography showed a left ventricular ejection fraction (LVEF) of 85%, a left ventricular outflow tract velocity-time integral (VTI) of 12.9 cm, and a dilated right ventricle with a fractional area change (FAC) of 25%. The right ventricular-to-right atrial pressure gradient was 28 mmHg, and moderate tricuspid regurgitation was present.

The patient was receiving digoxin, spironolactone, furosemide, valsartan, bisoprolol, sildenafil, levothyroxine, and warfarin. Given the progressive Fontan circulatory failure, multiorgan involvement, and absence of palliative surgical options, orthotopic heart transplantation was selected.

Before surgery, cardiac computed tomography (CT) angiography (**Figure 1**) and venography (**Figure 2**) were used to assess surgical approaches and plan reconstruction of the pulmonary artery branches.

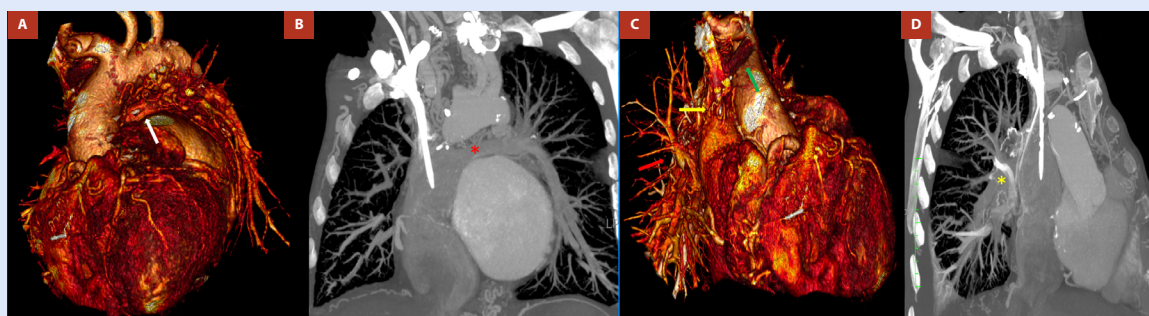


Figure 1. **A)** Three-dimensional cardiac reconstruction in a left oblique view showing the left pulmonary artery branch (white arrow), venoarterial collateral vessels at the level of the lobar branches, and pulmonary artery atresia. **B)** Coronal section corresponding to the view in panel A, confirming the absence of stenosis or thrombosis in the left pulmonary artery branch (red asterisk). **C)** Three-dimensional reconstruction showing the intracardiac Fontan circuit, with drainage from the superior vena cava (yellow arrow), persistence of the innominate vein (green arrow), and a patent right pulmonary artery branch (red arrow). **D)** Patency of the Fontan circuit and the right pulmonary artery branch (yellow asterisk).



Figure 2. **A)** Angiography showing a patent intracardiac Fontan circuit (red asterisk) associated with pulmonary artery atresia. **B)** Patent right pulmonary artery branch (yellow arrow), measuring 15.15 mm in diameter. **C)** Anteroposterior view showing a patent left pulmonary artery branch (blue arrow), measuring 12.4 mm in diameter. **D)** Multiple collateral vessels occluded with coils (white arrows), together with occlusion devices for aortopulmonary and venoarterial fistulas.

Surgical procedure

Bicaval orthotopic heart transplantation was performed, together with bovine pericardial patch reconstruction of both pulmonary artery branches and mediastinal packing. As a precaution, the right subclavian artery was exposed and an 8-mm polytetrafluoroethylene (PTFE) graft was anastomosed to it to provide access for emergency circulatory support if required. The left femoral vein was also cannulated.

After initiation of cardiopulmonary bypass, repeat sternotomy and extensive adhesiolysis were performed, followed by complete takedown of the Fontan circuit. Both pulmonary artery branches were reconstructed using a bovine pericardial patch (**Figure 3A**). The donor heart was then implanted by anastomosing the left atrium, venae cavae, aorta, and pulmonary artery. The innominate vein was reconstructed with a bovine pericardial patch because it had been used for superior venous cannulation earlier in the procedure (**Figure 3C**).

Substantial bleeding associated with multiple collateral vessels and hepatic dysfunction required multiple

blood transfusions and mediastinal packing. The total cardiopulmonary bypass time was 6 h, and the aortic cross-clamp time was 3 h 27 min. The packing was removed after 48 h without complications.

Postoperative course

In the immediate postoperative period, the patient developed haemorrhagic and cardiogenic shock with primary right ventricular graft dysfunction, requiring dobutamine, noradrenaline, vasopressin, and inhaled nitric oxide at 30 ppm, despite normal pulmonary vascular resistance. Initial echocardiography showed an LVEF of 71%, a right ventricular FAC of 20%, and severe tricuspid regurgitation.

He subsequently developed acute respiratory distress syndrome and septic shock caused by extended-spectrum β -lactamase (ESBL)-producing *Escherichia coli*. He received meropenem and vancomycin, with doses adjusted for haemodialysis, followed by linezolid. Inhaled nitric oxide was discontinued on postoperative day 5. Echocardiography on

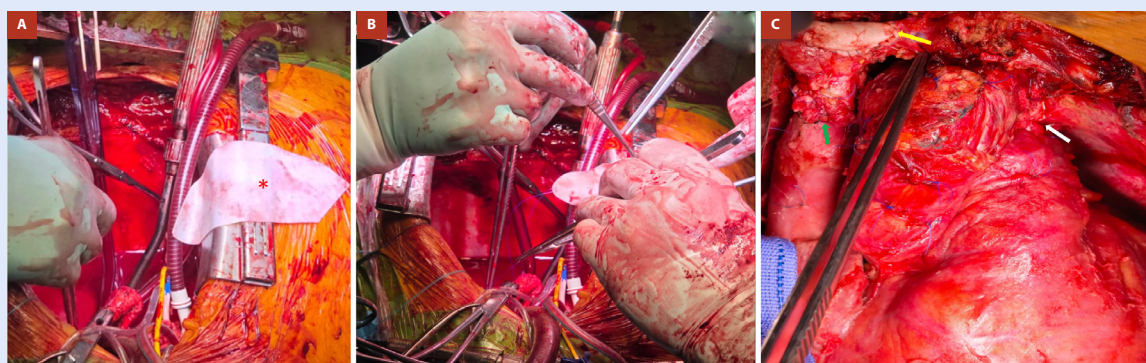


Figure 3. **A)** Preparation of a hexagonal bovine pericardial patch (red asterisk) to reconstruct the anterior wall of both pulmonary artery branches before anastomosis of the donor pulmonary artery. **B)** Anastomosis of the patch to the pulmonary artery branches using 5-0 polypropylene sutures. **C)** Anastomosis of the donor pulmonary artery to the reconstructed pulmonary artery branches using 5-0 polypropylene sutures (white arrow), anastomosis of the donor and recipient superior venae cavae using 5-0 polypropylene sutures (green arrow), and pericardial patch reconstruction of the junction between the jugular and innominate veins (yellow arrow), which had been used for superior venous cannulation.

day 13 showed improved right ventricular function, with an FAC of 32%, an LVEF of 68%, and a tricuspid annular plane systolic excursion of 9 mm. Because of prolonged mechanical ventilation, tracheostomy was performed on day 21, together with bilateral pleural drainage for pleural effusions.

Initial immunosuppression comprised intravenous methylprednisolone followed by prednisone, which was gradually tapered to 10 mg/day. Tacrolimus initiation was delayed until day 5 because of chronic kidney disease requiring haemodialysis. Mycophenolic acid was added on day 11 after resolution of lymphopenia, completing the triple immunosuppressive regimen.

By day 22, the patient was awake, breathing spontaneously through the tracheostomy, and receiving enteral nutrition. He developed recurrent bilateral pleural effusions, which were managed with repeated drainage and medical treatment. His recovery was complicated by recurrent respiratory infections and limited mobilisation due to critical illness neuropathy. Biventricular function remained preserved, with adequate pulmonary blood flow and no evidence of stenosis or rejection.

The patient died on postoperative day 87 following cardiorespiratory arrest secondary to respiratory failure, extreme hypercapnia, and refractory respiratory acidosis, in the setting of suspected tracheostomy tube obstruction caused by tracheobronchial haemorrhage.

Discussion

Fontan circulatory failure is characterised by haemodynamic deterioration, multiorgan involvement, and lymphatic dysfunction. In this setting, heart transplantation is the definitive treatment, although it is technically demanding and carries substantial perioperative mortality, particularly in patients with advanced disease. Contemporary evidence shows that post-transplant survival has progressively improved and that outcomes depend on appropriate patient selection, preoperative optimisation, and centre experience^(3,6).

Post-transplant survival is approximately 80% at 1 year, 70-75% at 5 years, and 60% at 10 years, despite technical and clinical challenges, including reconstruction of the great vessels, Fontan-associated liver disease, and protein-losing enteropathy⁽⁷⁾. Pretransplant mechanical ventilation, prolonged cardiopulmonary bypass times, and the need for postoperative circulatory support have been associated with early mortality^(8,9).

Assessment with CT and angiography is essential to delineate complex vascular anatomy, identify collateral vessels, and detect abnormalities that influence the surgical

strategy^(3,7). In this case, these investigations enabled vascular reconstruction to be planned before the donor organ arrived. Meshulami et al. report that surgical planning reduces ischaemic time, although up to 25% of patients require reoperation for bleeding⁽⁷⁾.

Vascular reconstruction is one of the main technical challenges of transplantation in patients with Fontan circulation. Previous interventions can result in distortion, hypoplasia, or adhesions involving the pulmonary arteries and great vessels. Prosthetic or pericardial material must therefore be available, and reconstruction must be planned in advance^(2,3,10). In our case, preoperative planning enabled patch reconstruction of both pulmonary artery branches and reconstruction of the innominate vein.

Postoperative management requires close monitoring because of the high incidence of bleeding, vasoplegia, and primary graft dysfunction⁽²⁾. Inhaled nitric oxide can facilitate adaptation of the pulmonary circulation after transplantation, even when pulmonary vascular resistance is normal^(3,10). Our patient developed severe bleeding and primary right ventricular graft dysfunction, requiring vasoactive support and inhaled nitric oxide. Although graft function recovered and remained stable, respiratory and infectious complications ultimately determined the outcome. Assessment of immunological status and treatment adherence is also essential, given the high prevalence of immunological sensitisation in this population^(4,10).

In conclusion, heart transplantation in adults with a failing Fontan circulation is a highly complex procedure because of altered vascular anatomy, multiorgan deterioration, and the risk of perioperative complications. Haemodynamic and imaging assessment, individualised planning of vascular reconstruction, and multidisciplinary coordination are essential to optimise outcomes. In this case, despite recovery of graft function, respiratory and infectious complications determined the outcome, underscoring the complexity of managing this patient population.

Ethical aspects

Written informed consent was obtained from the patient or their legal representative for publication of this case report and the accompanying clinical images. The patient's anonymity was maintained throughout.

Author contributions

RCM: Conceptualisation, investigation, writing-original draft, visualisation. **EMC and SZC:** Investigation, writing-original draft. **VRV, RZS, and FSP:** Supervision, writing-review and editing.

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