SCIENTIFIC LETTER

Heart Transplantation in a Cardiac Patient after the Fontan Procedure

Trasplante cardíaco en paciente cardiópata en estadio Fontan

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As a result of advances in diagnosis and surgical techniques, even patients with complex congenital heart disease can survive into adulthood with a high risk of end-stage heart failure. Therefore, the number of patients with congenital heart disease requiring heart transplantation has increased in recent decades.

For patients with single ventricle physiology heart disease, staged palliative operations culminating in the Fontan procedure which involves the complete bypass of the right heart remains the gold standard. Unfortunately, even the ideal candidate with a "perfect" Fontan surgery and adequate follow-up undergoes slow functional deterioration with development of progressive heart failure and a survival of 86% at 5 years and 74% at 15 years.

Patients with Fontan surgery and advanced heart failure are a cohort of patients candidate for heart transplantation.

Before placing a patient with a complete right heart bypass on the transplantation list, the possibility of optimizing the Glenn-Fontan circulation by hemodynamic or surgical interventions should be considered. If this is not possible, the pre-transplantation assessment process should be initiated.

As these patients have undergone multiple operations, they usually have a history of blood product transfusions and exogenous material implants, which increases the titers of lymphocytotoxic antibodies, as well as a history of multiple vascular accesses, which predisposes to thrombosis of the vascular accesses.

Regarding the surgical technique, in addition to the difficulties inherent in reoperation (mostly third or fourth sternotomy), there are aspects of the univentricular pathophysiology that require technical variations compared to the biventricular physiology.

For these reasons, transplantation after Fontan surgery is a challenge not only from a surgical point of view, but also from the time of pre-transplantation assessment to the distant postoperative follow-up.

We present the case of an 18-year-old patient diagnosed with dextroisomerism, type II B tricuspid atresia, double-sided superior vena cava and partial anomaly of pulmonary venous return.

Regarding surgical history, a left systemic-to-pulmonary anastomosis and later a right one were performed in neonatal stage. At the age of 3, the patient underwent bilateral Glenn surgery, Fontan surgery with fenestration with a 20 mm polytetrafluoroethylene (PTFE) conduit, correction of the anomalous pulmonary venous return and plastic surgery of both pulmonary artery branches, in one-stage surgery. At the age of 5, the patient underwent catheterization and attempted fenestration closure with cribriform Amplatzer™ but had residual shunt; thus, at the age of 8, a stent was placed to close it.

He then developed severe mitral regurgitation which led to ventricular dysfunction and eventually heart failure. As a result, he was placed on the heart transplantation list.

The pre-transplantation echocardiogram showed heterotaxia, dextroisomerism, permeable extracardiac conduit (EC) with biphasic flow velocity of 0.39 m/sec, closed fenestration without residual shunt, collector with laminar flow velocity of 0.49 m/sec, wide atrial septal defect, right atrioventricular (AV) valve atresia, left AV valve with severe regurgitation caused by coaptation, single dilated and trabeculated ventricle with moderate to severe dysfunction, left ventricular diastolic diameter (LVDD) 8.5 cm, left ventricular systolic diameter (LVSD) 6.8 cm, shortening fraction 20%, mitral annulus plane systolic excursion (MAPSE) 9 mm, E/A inflow pattern 0.7, bilateral Glenn procedure

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with biphasic flow, the left one with a velocity of 0.34 m/sec and the right one of 0.5 m/sec.

Pre-transplantation catheterization was performed and showed pressures of 16 mmHg in bilateral Glenn and Fontan procedures and 95/56 (63) mmHg in aorta. A Qp/Qs ratio of 0.96 and a Rp/Rs ratio of 0.07 were obtained (Figure 1).

An HLA panel study was performed and showed sensitization of 18% for group I and of 37% for group II, for which the patient received gamma globulin (3 doses), and plasmapheresis in extracorporeal circulation was programmed at the time of heart transplantation.

Given the complexity of the setting due to the clinical and anatomical conditions and the requirements of highly complex medical technology, a transplantation simulation was performed in the operating room in the presence of cardiologists, cardiovascular surgeons, perfusionists, hematologists, anesthesiologists, surgical instrument technicians and intensivists. This allowed us to optimize time and reduce potential errors and adverse events, as these are not procedures we usually perform on other patients.

On the day of transplantation, femoral cannulation was performed, and the patient was placed in extracorporeal circulation via the femoral route due to adhesions between the single ventricle and the sternum. Following re-sternotomy, left Glenn cannulation was carried out. The patient was placed in deep hypothermia, the aorta was clamped and the heart was explanted. Subsequently, implantation was performed by anastomoses of the left atrium, the pulmonary artery and the aorta. The cardiac cavities were purged, and the aorta was declamped. Implantation was completed by anastomosis of the inferior vena cava and ligation of the superior vena cava, leaving both cavopulmonary anastomoses permeable, bilateral Glenn. (Figure 2)

In extracorporeal circulation, the plasmapheresis protocol was followed during cardiopulmonary bypass, which is indicated for patients with heparin-induced thrombocytopenia (HIT), patients with antiphospholipid syndrome (APS) and hypersensitized patients who remain on the heart transplantation list, as in this case.

The patient progressed favorably and was extubated

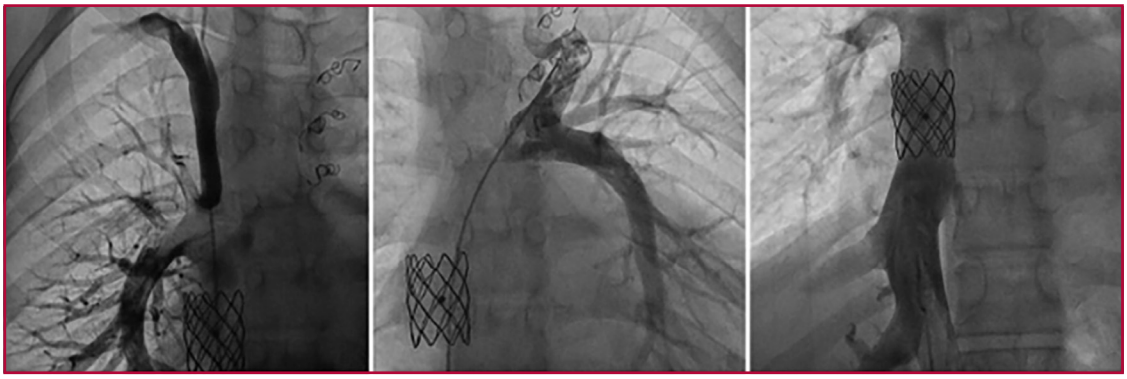

Fig. 1. Pre-transplantation catheterization. a) Right Glenn procedure. b) Left Glenn procedure. c) Extracardiac conduit with implanted stent to close fenestration.

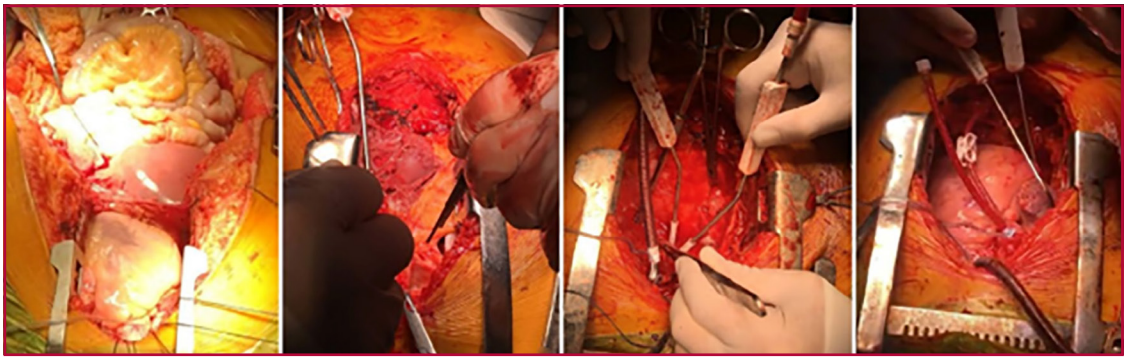

Fig. 2. Surgical images. a) Multi-organ ablation. b) Opening; patient in extracorporeal circulation via femoral approach; clamping; lesion in the anterior wall of the single ventricle. c) Empty mediastinal cavity; left Glenn cannulation. d) Implanted beating heart and declamped aorta.

48 hours after transplantation. He is currently receiving outpatient treatment with immunosuppressants, corticosteroids, diuretics, prophylactic antibiotics and insulin for diabetes secondary to glucocorticoids.

The control echocardiogram showed preserved ventricular function, LVDD 4.7 cm, LVSD 3 cm, shortening fraction 36%, left ventricular ejection fraction by Simpson method 65%, MAPSE 18 mm, tricuspid annulus plane systolic excursion (TAPSE) 14.4 mm, both outflow tracts free, competent aortic valve, trivial pulmonary valve regurgitation, bilateral Glenn with preserved and pulsatile flow.

Catheterization and biopsy were performed 2 months after surgery and showed no signs compatible with rejection.

Patients with univentricular circulation are usually candidates for heart transplantation in the long term. In our opinion, adequate patient selection and pre- and post-operative planning are necessary to improve the quality and success of transplantation in patients with congenital heart disease. Simulation with all the professionals involved in the transplantation made it easier to organize and optimize the operating time.

Conflicts of interest

None declared.

(See conflicts of interest forms on the website).

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