Ventricular Assist Device Implantation Late After Double Switch Operation for L-Transposition of the Great Arteries

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We provided a left ventricular assist device (LVAD) for a 22-year-old man with congenital L-transposition of the great arteries after anatomic repair at the age of 7 years. He was hospitalized for progressive low-output syndrome caused by intractable biventricular failure. He received LVAD in his morphologic left ventricle with a concomitant pulmonary valve replacement. After the surgery, critical multiorgan failure with severe right heart failure occurred. It took three postoperative months to normalize all organ function following improvement of morphologic right ventricular function. He has remained stable with LVAD support for 1.5 years.


In congenitally corrected L-transposition of the great arteries (CCTGA), dysfunction of the systemic ventricle, which is morphologically the right ventricle, is observed with advancing age. At our institution, anatomic repair—so-called double-switch operation for CCTGA—has been conducted as a first-line treatment since 1983 when ideal cardiac conditions are confirmed [1]. However, some patients in the long term after anatomic repair have demonstrated unfavorable systemic ventricular dysfunction even though the ventricle is morphologically the left ventricle. We describe our experience of using a continuous-flow left ventricular assist device (LVAD) to achieve recovery from severe multiorgan failure in a patient with intractable biventricular failure long after anatomic repair for CCTGA.

A 22-year-old university student with CCTGA was admitted to the hospital for drastic progression of symptomatic biventricular failure. He received a diagnosis at 6 months of age and underwent anatomic repair consisting of a Senning-Rastelli procedure at 7 years of age. Laboratory test results indicated liver and renal dysfunction. Inotropic therapy was administered in addition to respirator support, intraaortic balloon pump circulatory support, and continuous hemodialysis. However, his condition deteriorated to INTERMACS (Interagency Registry for Mechanically Assisted Circulatory Support) Profile 1. Preoperative echocardiography revealed a left ventricular end-diastolic dimension/end-systolic dimension of 6.0/5.5 cm, with a left ventricular fractional shortening of 0.08, a right ventricular fractional shortening of 0.20, severe pulmonary regurgitation, moderate tricuspid regurgitation, and severe mitral regurgitation.

An implantable, continuous-flow EVAHEART LVAD (Sun Medical Technology Research Corp, Nagano, Japan) was implanted [2]. The patient’s extremely slender physique (Fig 1) meant that the LVAD pump had to be implanted into his abdominal cavity (Fig 2). Replacement of the pulmonary valve (21 mm; Carpentier-Edwards Perimount Bioprosthesis; Edwards Lifesciences Corp, Irvine, CA) was also performed concomitantly. Immediate postoperative laboratory test results suggested severe liver and renal dysfunction caused by severe right heart failure. Recovery was achieved with LVAD circulatory support and dedicated intensive management to handle severe right heart failure, including administration of inotropes, phosphodiesterase III and V inhibitors, and nitric oxide while the sternum remained open with skin closure. Renal and liver function had normalized by 3 months postoperatively. He was discharged from the hospital at 281 days after LVAD implantation and has remained stable with LVAD support for 1.5 years as of December 2013, being regular outpatient management and awaiting heart transplantation.

Comment

We have described a case of successful recovery from severe biventricular failure with multi-organ failure in CCTGA long after anatomic repair. In terms of the operative method, we considered implantation of the LVAD pump as well as inflow and outflow cannulas possible if the pump was placed in the abdominal cavity, but right ventricular support was not realistic because of the anatomic features of the patient and the fact that paracorporeal right ventricular assist device (RVAD) has been the only option available in Japan. Management of postoperative right heart failure was controlled by continuous-flow LVAD support together with long-term sternal remaining open with skin closure, and administration of inotropes, phosphodiesterase III and V inhibitors, and nitric oxide.

The EVAHEART LVAD was used because of its high flow characteristic as well as its pump size and flexible inflow and outflow conduits, enabling it to be implanted in patients with special anatomic features. Anatomically,
the left ventricle was located at the right anterior of the right ventricle, making it difficult to place the pump in the normal position above the peritoneum. However, after incising the peritoneum, we were able to create a pouch by suturing a Gore-Tex sheet to the peritoneum. The body of the pump was wrapped by the pouch and placed intraperitoneally. Flexible inflow and outflow conduits enabled this anatomically difficult implantation.

In this patient, implantation of a RVAD between the right atrium or ventricle and pulmonary artery was avoided for several reasons. First, the patient's status was post-anatomic repair with a double-switch operation at the right atrium. Accordingly, cannulation to the right atrium was avoided. Second, given the patient's extreme thinness and the limited space of the left chest cavity where his right ventricle was located, it was not considered favorable to implant an RVAD. Third, the pulmonary artery was located posterior to the aorta, which made cannulation to the pulmonary artery extremely difficult. Because of these factors, implantation of an RVAD was considered infeasible, although CircuLite Synergy Micro-Pump (CircuLite, Inc, Saddle Brook, NJ) could have been an option if it were available in Japan. Regarding pulmonary valve replacement, a 21-mm Carpentier-Edwards Perimount bioprosthesis was implanted after considering the prevention of postoperative thrombosis. Regarding the tricuspid regurgitation, we avoided performing tricuspid annuloplasty for the following reasons. First, the tricuspid regurgitation was expected to be decreased because of right ventricular size reduction owing to the suppression of pulmonary valve regurgitation. Second, it is not preferable to try tricuspid annular plasty, which requires cardiac arrest because of the trans-atrial-septal approach through the right side of the left atrium where a Senning procedure was performed for possible postoperative cardiac functional deterioration.

Left heart failure owing to systemic (left) ventricle dysfunction many years after anatomic repair for CCTGA has been reported previously [3, 4]. As anatomic repair has been used as the first-line treatment for CCTGA at our institution since 1983 [1], over several decades we have occasionally encountered patients who experienced biventricular failure, mainly from left ventricular dysfunction. Left ventricular dysfunction is speculated to be due to, in some cases at least, a specific anatomic configuration of the left ventricle leading to physical pressure effects from the sternum and postoperative adhesions. In the present case, however, pathologic findings from a specimen taken from the left ventricle showed myocardial fibrosis resembling dilated cardiomyopathy.

In the treatment of CCTGA without anatomic repair, there have been some reports of successful VAD.
implantation to the right (systemic) ventricular dysfunction [5, 6]. On the other hand, the present case is the first successful implantation of an LVAD to the left ventricle for systemic ventricular dysfunction after anatomic repair in a double-switch operation. Anatomic repair has been performed in a large number of cases over the past 30 years. Accordingly, there is potential for more such cases in the future. The anatomic characteristics of each case, the position of the implantable pump, and the placement of the graft must be carefully considered. In summary, this case report describes one successful strategy to rescue a patient with CCTGA who had biventricular failure after anatomic repair with a double-switch operation.

References