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CASE REPORT

Artificial Heart (Clinical)

Jarvik 2000 axial flow ventricular assist device in right single ventricle after Fontan operation

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Abstract
We present a case of successful ventricular assist device support in a 13-year-old female diagnosed with right single ventricle, asplenia, dextrocardia, who had undergone a Fontan operation at 4 years old in an associated children hospital. She underwent placement of Jarvik 2000 axial flow ventricular assist device to the morphologic right ventricle which worked as systemic ventricle. The postoperative course was not eventful. She was waiting for heart transplantation attending high school 3 years after implantation.

Keywords Ventricular assist device · Jarvik 2000 · Fontan

Introduction

Implantable ventricular assist device (VAD) operation and management in patients with complex congenital heart disease has not been well described. The implantable VAD in patients with single ventricle after a Fontan operation is one of the challenging treatments. Jarvik 2000 axial flow VAD is a unique intraventricular pump, which can be placed to not only the left ventricle but also the right ventricle. We present a case of successful implantable VAD support in a patient with right single ventricle, asplenia, who had undergone a Fontan operation.

Clinical summary

A 13-year-old female was diagnosed with right single ventricle, total anomalous pulmonary venous connection, asplenia, dextrocardia, who had undergone a bidirectional Glenn procedure (BDG) and pulmonary venous connection repair at 9 months old and a Fontan operation, total cavopulmonary connection (TCPC), at 4 years old in an associated children hospital. Thereafter, her cardiac function gradually declined. She was referred to adult congenital heart disease section of our hospital at 10 years old, and showed slow progression to New York Heart Association functional class IV symptoms despite the intensified heart failure therapy with beta-blocker, angiotensin-converting-enzyme inhibitor, and diuretics. She was taking phosphodiesterase 5 inhibitor, tadalafil to reduce the pulmonary vascular resistance. She was admitted for worsening cardiac failure, and then a continuous dobutamine infusion was started. Echocardiography revealed a dilated systemic morphologic right ventricle with severely reduced function in which ejection fraction was less than 0.30, and severe systemic atrioventricular valve regurgitation. Cardiac catheterization showed low cardiac output syndrome despite a smooth Fontan circulation (mixed venous oxygen saturation, 38.7%; cardiac index, 1.10 L/min/m^2; central venous pressure, 18 mmHg; pulmonary artery wedge pressure, 12 mmHg). Serum creatinine level was normal (0.43 mg/dL), serum total bilirubin level

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slightly elevated (2.2 mg/dL), and brain natriuretic peptide level elevated (389 pg/mL).

She underwent placement of Jarvik 2000 axial flow VAD to the morphologic right ventricle which worked as systemic ventricle. At the time of implant, she was 33.2 kg in weight, 139 cm in height, with a BSA of 1.14 m^2. A re-do median sternotomy was performed. Cardiopulmonary bypass was established through the graft sutured to the right femoral artery and two venous cannulae in left superior vena cava and inferior vena cava. The apical portion of the inflow was finally decided with epicardial echocardiography, and the resection of trabeculae was decided to be not necessary. After the ventriculotomy with the dedicated apical puncher (coring knife), the apical cuff was suture using 12 pledgeted 2–0 polyester sutures and the pump body was inserted into it. The outflow graft (16 mm Hemashield) was anastomosed to the ascending aorta with a running 4–0 polypropylene suture using the side-biting clamp. Tricuspid valvuloplasty was not performed to prevent the inflow obstruction. The VAD was initiated and cardiopulmonary bypass weaned off gradually under nitric oxide (NO) inhalation. The flow dial was set to 2 (9000 rpm). The patient was extubated 2 h after implantation. After starting oral intake, NO inhalation was switched to the oral administration of tadalafil. The postoperative course was not eventful. Mixed venous oxygen saturation and cardiac output remarkably increased and arterial pressure decreased (mixed venous oxygen saturation, 61.3%; cardiac index, 2.80 L/min/m^2; central venous pressure, 12 mmHg; pulmonary artery wedge pressure, 5 mmHg), and brain natriuretic peptide level decreased (11 pg/ml) after Jarvik 2000 implantation. 3D-reconstruction of CT images is shown in Fig. 1. Her condition was stable, and she was waiting for heart transplantation at home attending high school 3 years after implantation.

Fig. 1 Postoperative chest computed tomography showing the Jarvik 2000 axial flow ventricular assist device implanted in the systemic morphologic right ventricle

Comment

A few case reports demonstrated implantable continuous-flow VAD implantation in patient with single-ventricle palliation [1]. Jarvik 2000 axial flow VAD is an intraventricular blood pump and it is inherently usable as not only LVAD but also right VAD (RVAD) [2]. Case reports of RVAD with Jarvik 2000 axial flow VAD are also a few [3, 4]. We report Jarvik 2000 axial flow VAD placement in a small-body patient with right single ventricle and dextrocardia after a Fontan operation.

The clinical outcome of single-ventricle palliation and the strategy of Fontan-type operation have advanced remarkably. According to data regarding congenital heart surgery performed between January 2013 and December 2014 obtained from Japan Cardiovascular Surgery Database, the numbers of total cases of BDG and TCPC were 713 and 875, respectively, and the mortality rates of BDG and TCPC were less than 2% [5]. A single ventricle-supported circulation is an unnatural physiological state, and inherently has a limitation compared with the normal biventricular circulation. Case reports of successful mechanical circulatory support (MCS) including VAD in patient who reached a Fontan operation are a few, but it is expected that the number of cases will increase in the future. MCS for Fontan circulation have yet to be clearly established. It is necessary to consolidate and investigate as many case reports as possible, including not only successful cases but also unsuccessful cases.

Right heart failure is one of the serious problems after left VAD implantation. Single ventricle physiological state is the ultimate "right heart failure". The preparation of lung conditions to keep pulmonary vascular resistance low is playing an important role before, during, and after "systemic" VAD implantation. Appropriate NO inhalation during a perioperative period is essential, and should switch to the administration of pulmonary-hypertension-treatment drug after starting oral ingestion. Early extubation, NO inhalation, and tadalafil would be effective in this present case. The recent progress in the management for pulmonary hypertension is expected to contribute the treatment in MCS for Fontan circulation.

In summary, we report a case of Jarvik 2000 axial flow VAD implantation in a patient with right single ventricle after a Fontan operation. Single-ventricle cases requiring long-term management with implantable VAD are expected to increase in the future. Indication, timing, and management of VAD implantation need to be considered thoroughly.

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