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Case report

Implantation of a left ventricular assist device as a destination therapy in Duchenne muscular dystrophy patients with end stage cardiac failure: Management and lessons learned

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Abstract

Duchenne muscular dystrophy (DMD) is an X-linked recessive disorder, characterized by progressive skeletal muscle weakness, loss of ambulation, and death secondary to cardiac or respiratory failure. End-stage dilated cardiomyopathy (DCM) is a frequent finding in DMD patients, they are rarely candidates for cardiac transplantation. Recently, the use of ventricular assist devices as a destination therapy (DT) as an alternative to cardiac transplantation in DMD patients has been described. Preoperative planning and patient selection play a significant role in the successful postoperative course of these patients. We describe the preoperative, intraoperative and postoperative management of Jarvik 2000 implantation in 4 DMD pediatric (age range 12–17 years) patients. We also describe the complications that may occur. The most frequent were bleeding and difficulty in weaning from mechanical ventilation. Our standard protocol includes: 1) preoperative multidisciplinary evaluation and selection, 2) preoperative and postoperative non-invasive ventilation and cough machine cycles, 3) intraoperative use of near infrared spectroscopy (NIRS) and transesophageal echocardiography, 4) attention on surgical blood loss, use of tranexamic acid and prothrombin complexes, 5) early extubation and 6) avoiding the use of nasogastric feeding tubes and nasal temperature probes. Our case reports describe the use of Jarvik 2000 as a destination therapy in young patients emphasizing the use of ventricular assist devices as a new therapeutic option in DMD.

Keywords: Duchenne muscular dystrophy; Left ventricular assist device

1. Introduction

Duchenne muscular dystrophy (DMD) is an X-linked recessive disorder, characterized by progressive skeletal muscle weakness, loss of ambulation, and death secondary to cardiac or respiratory failure in the 2nd or 3rd decade [1]. Despite the development of end-stage dilated cardiomyopathy (DCM), patients are rarely candidates for cardiac transplantation due to the systemic nature of their disease, thus limiting treatment options [2].

A ventricular assist device (VAD) is a mechanical pump that is implanted to help the heart's weakened ventricle to pump blood throughout the body.

VADs are utilized as a bridge to transplantation [3], recovery [4] or as a destination therapy (DT) [5,6]. Recently, the use of VADs as destination therapy in DMD patients has been described [7,8].

The VAD can be used as a bridge-to-transplantation. This means it can help a patient survive until a donor heart becomes available for transplant. Otherwise, the VAD can be used as a destination therapy (DT), which is an alternative to heart transplantation. Destination therapy provides long-term support in patients who are not candidates for transplant. When used as a bridge-to-transplant or as DT, the VAD provides effective hemodynamic support, maintains or improves other organ function, improves exercise performance and enables participation in cardiac rehabilitation.

A VAD does not replace the heart. It assists the patient's own heart to pump blood throughout the body, decreasing the work of the ventricle.

A left ventricular assist device (LVAD) receives blood from the left ventricle and delivers it to the aorta. A right ventricular

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assist device (RVAD) receives blood from either the right atrium or right ventricle and delivers it to the pulmonary artery.

The survival of LVADs as a DT is around 50% at 36 months as reported in the "Sixth INTERMACS Annual Report: A 10,000-Patient Database" [9].

Preoperative planning and patient selection play a significant role in the successful postoperative course of these patients. We describe the preoperative, intraoperative and postoperative management of Jarvik 2000 implantation in 4 DMD pediatric patients.

2. Case reports

2.1. Patient MS

A 15 year old, 57 kg male with DMD, scoliosis, dilated cardiomyopathy, and NYHA IV was diagnosed with DMD at the age of 6 and by 10 was wheelchair dependent. Steroids were administered for 3 years as of the age of 7. He experienced during the last year, 3 episodes of acute cardiac failure despite maximal medical treatment and was admitted to our intensive care unit (ICU) with an ejection fraction of 15% unresponsive to inotropes. We began an empiric intravenous antibiotic therapy with ceftazidime due to an upper respiratory tract infection. A diagnostic cardiac catheterism showed a transpulmonary gradient of 12 mmHg. We decided to implant an LVAD. Preoperatively, non-invasive ventilation and cough machine cycles were started. On hospital day 40th Jarvik 2000 was placed without intraoperative complications. The following day the patient was successfully extubated. We also started elective non-invasive ventilation (NIV), respiratory physiotherapy and cycles of cough machine. The following day a pericardial effusion required drainage. Again the patient was successfully extubated after 24 hours. His ICU stay was uncomplicated and lasted a total of 34 days. Total hospital stay was 82 days. The patient has been home since 2010 and returns for his routine examinations.

2.2. Patient CM

We describe the case of a 12 year old male, 43 kg patient with DMD, scoliosis, and a previous history of cholelithiasis. He received steroid therapy since 2005. He was wheelchair dependent, at the age of 9 he was diagnosed with a dilated cardiomyopathy, treated with ace-inhibitors and carvedilol. He was transferred to our cardiology department with symptoms of fatigue, loss of appetite, nausea and abdominal pain. After maximizing oral cardiovascular medications, he was started on dopamine 8 mcg/kg/minute and a preconditioning cycle with levosimendan 0.1 mcg/kg/minute. Despite therapy his condition did not improve. He was transferred to the ICU with signs of low cardiac output. The patient had an adequate respiratory reserve and did not require preoperative NIV. Jarvik 2000 was placed without intraoperative complications. During the first postoperative night chest drain bleeding required red blood cell and plasma transfusions and extubation was delayed. Five days post-surgery chylothorax was diagnosed which was treated with a median chain triglyceride diet (MCT). The 13th postoperative day after nasogastric tube placement we observed a severe retropharyngeal bleeding and tongue edema which required intubation and surgical intervention. A few days later the presence of a bilateral pleural effusion required chest drain placement. This was complicated by bleeding from the thorax and a surgical revision was performed. The patient was extubated 16 days later and electively placed on non-invasive ventilation and cough machine cycles. Total intensive care stay was 72 days. Hospital stay was 180 days. Since discharge the patient has been at home for over a year.

2.3. Patient DLN

A 15 year old 45 kg male patient with DMD, scoliosis and acute dilated cardiomyopathy was transferred to our ICU after an episode of cardiac arrest which required cardiopulmonary resuscitation and institution of ECMO (extracorporeal membrane oxygenation). Preoperative inotropes included dopamine 8 mcg/kg/minute and adrenaline 0.3 mcg/kg/minute. The following day Jarvik 2000 was implanted. His postoperative course was extremely complicated. Due to his severe scoliosis while attempting to place a chest drain, his spleen ruptured. This led to abdominal bleeding which required several surgical interventions, unsuccessful embolization of the splenic artery and finally splenectomy. Due to his abdominal bleeding anticoagulation was suspended for 32 days. We attempted to utilize neutrally adjusted ventilator assist (NAVA) to facilitate ventilator weaning, however we were unable to obtain an adequate Edi signal due to interference with Jarvik 2000. After several attempts of weaning from mechanical ventilation due to paralysis of a vocal cord the patient after 30 days from the surgery underwent a tracheostomy. ICU stay was 110 days. Total hospital stay was 150 days. The patient approximately 1 year later died in a local hospital due to a bleeding complication from his tracheostomy tube.

2.4. Patient GA

A 17 year old male patient with DMD, dilated cardiomyopathy previous to scoliosis correction in 2012 was admitted to our ICU with respiratory insufficiency, low cardiac output, abdominal angina, and an ejection fraction below 20%. We began dopamine 6 mcg/kg/min and levosimendan 0.1 mcg/kg/min (for 72 hours). The patient started preoperative nocturnal non-invasive ventilation. After two weeks of therapy Jarvik 2000 was implanted. After 24 hours the patient was extubated and continued his cycles of NIV and cough machine. His clinical course was unremarkable. Total ICU stay was 30 days. Total hospital stay was 52 days. The patient is alive and doing well. Table 1 summarizes patient characteristics.

3. Discussion

Patients with DMD suffer from a progressive muscle weakness which results in a failure to walk by adolescence and eventual death from respiratory and cardiac failure before the end of the third decade [8]. Dilated cardiomyopathy occurs in over 50% of patients by 15 years of age [10].

Since 2010 at our institution Jarvik 2000 has been used as a device to sustain cardiac function in children with irreversible