KJTCVS
The Korean Journal of Thoracic and Cardiovascular Surgery
https://doi.org/10.5090/kjtcs.2020.53.2.79
pISSN: 2233-601X eISSN: 2093-6516
Korean J Thorac Cardiovasc Surg. 2020;53(2):79-81

Case Report

The First Pediatric Heart Transplantation Bridged by a Durable Left Ventricular Assist Device in Korea

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ARTICLE INFO
Received May 8, 2019
Revised November 1, 2019
Accepted November 4, 2019

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Treatment options for children with end-stage heart failure are limited. We report the first case of a successful pediatric heart transplantation bridged with a durable left ventricular assist device in Korea. A 10-month-old female infant with dilated cardiomyopathy and left ventricular non-compaction was listed for heart transplantation. During the waiting period, the patient's status deteriorated. Therefore, we decided to provide support with a durable left ventricular assist device as a bridge to transplantation. The patient was successfully bridged to heart transplantation with effective support and without any major adverse events.

Keywords: Transplantation, Pediatric, Cardiomyopathies, Ventricular assist device

Case report

Treatment options for children with end-stage heart failure are limited. Heart transplantation is the only viable option for these patients, and durable mechanical circulatory support is widely accepted as a bridge to transplantation despite the high rate of complications [1-3]. We report the first case of successful pediatric heart transplantation bridged with a durable pulsatile left ventricular assist device in Korea.

A 10-month-old female infant was referred to our hospital for severe cardiomegaly. She had presented to her primary care physician with symptoms of poor oral intake and lethargy lasting for a month. She became very lethargic during the examination by the primary care physician and was promptly transferred to our cardiac intensive care unit for evaluation of the cardiogenic cause. The patient's initial vital signs were as follows: blood pressure, 110/63 mm Hg; heart rate, 159 beats/min; and respiratory rate, 54 breaths/min. A chest X-ray examination revealed marked cardiomegaly (Fig. 1). On an echocardiographic examination, the left ventricular ejection fraction was estimated as 13%, and severe dilatation of the left ventricle and the left atrium was noted, which was consistent with dilated cardiomyopathy. A tricuspid annular plane systolic excursion of 11 mm, a tissue Doppler imaging–derived tricuspid lateral annular systolic velocity of 8 cm/sec, and trivial tricuspid regurgitation suggested a mild degree of right ventricular dysfunction. The sponge-like features of the left ventricular myocardium suggested left ventricular non-compaction. Medications including an angiotensin-converting enzyme inhibitor, an aldosterone antagonist, and a beta-blocker were administered along with intravenous milrinone. The patient partly recovered from her lethargic condition, but tachypnea and activity limitation persisted. The patient's left ventricular ejection fraction slightly improved to 20%. Because her heart failure symptoms were refractory despite optimal medication, she was listed for heart transplantation. Because her blood type was O-positive, she was expected to have a long waiting period. After 50 days of medical treatment, the patient's condition deteriorated rapidly. She was intubated and mechanically ventilated. Nevertheless, she was hypotensive and developed oliguria. Veno-arterial extracorporeal membrane oxygenation (ECMO) was initiated via neck cannulation. After 3 days of ECMO support, a left ventricular assist device was implanted with an extracorporeal pulsatile device (Berlin Heart EXCOR; Berlin Heart GmbH, Berlin, Germany). Her

https://doi.org/10.5090/kjtcs.2020.53.2.79

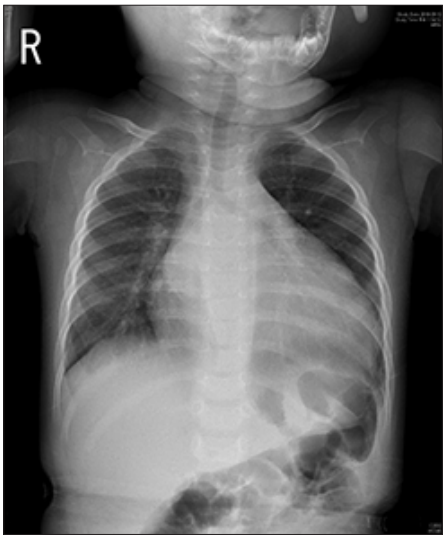

Fig. 1. Initial chest radiography demonstrated marked cardiomegaly and pulmonary congestion.

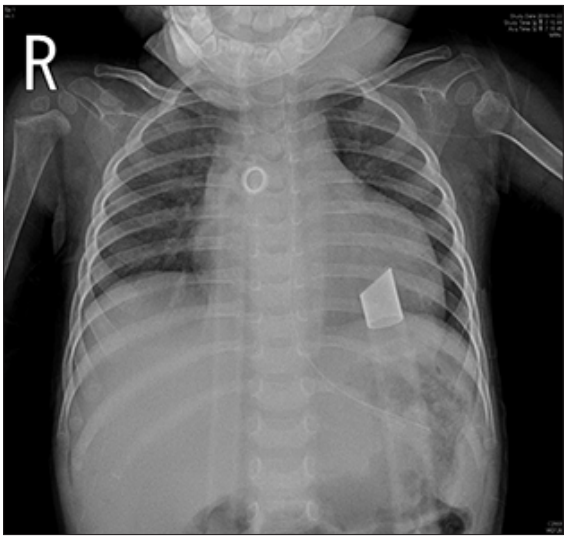

Fig. 2. Chest radiography after Berlin Heart EXCOR (Berlin Heart GmbH, Berlin, Germany) implantation.

body surface area was 0.4 m^2, and she was supported with a 15-mL pump. When performing apical cannulation of the left ventricle, a thorough and extensive resection of the trabeculation was performed to address left ventricular non-compaction. A 9-mm apical cannula was fixed to the apical hole with multiple interrupted stitches with an autologous pericardial strip. For outflow graft connection, a short, 10-mm expanded polytetrafluoroethylene tube graft was anastomosed to the ascending aorta with partial clamping on the beating heart, and the outflow cannula was connected to the graft. Two silk ties were used to fix the cannula to the graft. Anticoagulation therapy with heparin was started 24 hours after the operation, and acetylsalicylic acid was initiated on postoperative day 3. On post-implantation day 6, delayed mediastinal bleeding was noted, and the hematoma compressing the right atrium was surgically removed. Bleeding from a needle hole was observed at the aortic graft anastomosis site. The patient recovered uneventfully and was managed in the general ward from postoperative day 8 onwards (Fig. 2). Anticoagulation therapy was maintained with warfarin, acetylsalicylic acid, and clopidogrel at a target international normalized ratio range of 3.0–4.0. The pump was checked for abnormal membrane motion and thrombi by the nursing team every 2 hours. On post-implantation day 24, the patient underwent heart transplantation from a 5-year-old donor with a body weight of 29 kg. The total ischemic time was 192 minutes. The transplanted heart was oversized for the patient, but she recovered without any evidence of hyperperfusion syndrome. She was able to be weaned from mechanical ventilation on postoperative day 4 and was discharged home on postoperative day 25. She is doing well, and has not shown any rejection episodes over the course of 6 months.

The parent of this patient provided written informed consent for the publication of clinical details and images.

Discussion

In small children and infants with end-stage heart failure, heart transplantation is the only long-term treatment option. However, waitlist mortality is high because of the scarcity of heart donors. According to the data collected from the United States registry, during 6 months after being listed for heart transplantation, 23% of patients died on the waiting list [4]. Although the total number of transplantations has not increased, the number of pediatric heart transplantations that are bridged with ventricular assist devices is increasing worldwide [5,6]. Post-transplant outcomes in patients after ventricular assist device support are similar to those of medically supported patients and are superior to that of patients treated with ECMO [7]. In Korea, the number of pediatric heart transplantations remains low, with less than 20 transplants a year, and the frequency of transplantations in young children and infants is especially low [8]. Until recently, mechanical circulatory support for children in Korea was most often provided by temporary support via ECMO. However, time limitations due to possible thromboembolic and hemorrhagic complications are a major drawback of ECMO. The

Berlin Heart EXCOR (Berlin Heart GmbH) is a paracorporeal, pneumatically-driven pulsatile pump device that is available with chambers of various stroke volumes. It is the most frequently used device in infants with end-stage heart failure as a bridge to transplantation [6]. The success rate of bridging to transplantation ranges from 37% to 72.5% [9].

The present case is the first successful pediatric heart transplantation bridged with the Berlin Heart EXCOR (Berlin Heart GmbH) in Korea. The severely decompensated infant was successfully switched from ECMO to a durable ventricular assist device and underwent transplantation safely. Frequent cerebrovascular complications have been reported with the Berlin Heart EXCOR (Berlin Heart GmbH) [10]. In this case, the patient received warfarin and dual antiplatelet therapy, and was free of any neurologic events. However, this case does not provide sufficient information on the effect of the Berlin Heart EXCOR (Berlin Heart GmbH) on growth and development due to the short duration of support. Considering that coverage by the National Health Insurance Service for the Berlin Heart EXCOR (Berlin Heart GmbH) was approved in 2018, we expect an increase in the number of pediatric heart transplantations bridged by durable ventricular assist devices in the future.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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