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Cardiac Resynchronization Therapy in Infant with Dilated Cardiomyopathy during Extracorporeal Membrane Oxygenator

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Although heart transplantation is a final therapeutic option in pediatric patients with dilated cardiomyopathy (DCMP), the shortage of pediatric heart donors is a major obstacle. In adults with DCMP characterized by cardiac dyssynchrony, cardiac resynchronization therapy (CRT) is known to be an effective treatment option. However, there is a lack of evidence on the effectiveness of CRT in infants with DCMP. Several studies have reported improvement in hemodynamics and cardiac performance following CRT in infants with DCMP. Here, we report CRT in an infant with DCMP during extracorporeal membrane oxygenation with 5 months of follow-up.

Key words: 1. Cardiomyopathy
2. Cardiac resynchronization therapy
3. Extracorporeal membrane oxyenation

CASE REPORT

A 5-month-old girl who was diagnosed with dilated cardiomyopathy (DCMP) at 3 months of age was admitted to our hospital with excessive sweating, poor oral intake, and oliguria. On admission, the echocardiogram showed a markedly enlarged left ventricular (LV) end diastolic dimension of 47 mm, decreased LV ejection fraction of 15%, and more than moderate mitral regurgitation (MR) (Table 1). An electrocardiogram revealed a prolonged QRS duration of 130 ms (Table 1). Despite maximum medical treatment including infusion of a high dosage of inotropes and vasodilators in the intensive care unit, decreased urine output and severe lactic acidosis persisted, and the blood level of brain natriuretic peptide (BNP) remained high at 9,669 pg/mL (Table 1). Emergency extracorporeal membrane oxygenation (ECMO) was initiated through an open cervical approach, and the patient was listed as a candidate for heart transplantation. Considering the extreme shortage of heart donors in young children, we attempted to wean the patient off ECMO and succeeded after 8 days of support. While awaiting heart transplantation, the patient's condition rapidly deteriorated and she required repeated ECMO support 20 days after being weaned off ECMO. Unfortunately, ischemic insult to the brain, kidney, and bowel was evident, and significant sequelae were expected to remain. Therefore, the heart transplantation was aborted, and a decision was made to offer cardiac resynchronization therapy (CRT) as another option. To evaluate whether CRT could improve the patient's ventricular performance, we performed echocardiography with tissue synchronization imaging, again. Transthoracic echocardiography still showed LV global hypokinesia and asynchronous ventricular

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Table 1. Parameters before and after cardiac resynchronization therapy implantation

Variable	Preoperative	Postop 7 wk	Postop 5 mo
QRS duration (ms)	130	120	92
Left ventricular end diastolic dimension (mm)	47	36.3	33.4
Left ventricular ejection fraction[a] (%)	15	41.7	59.7
Mitral regurgitation (grade)	3−4	2−3	1−2
Septal-to-posterior wall motion delay (ms)	236	-	3
Brain natriuretic peptide (pg/mL)	9,669	146	29

Postop, postoperative.
[a]Measured by the M-mode.

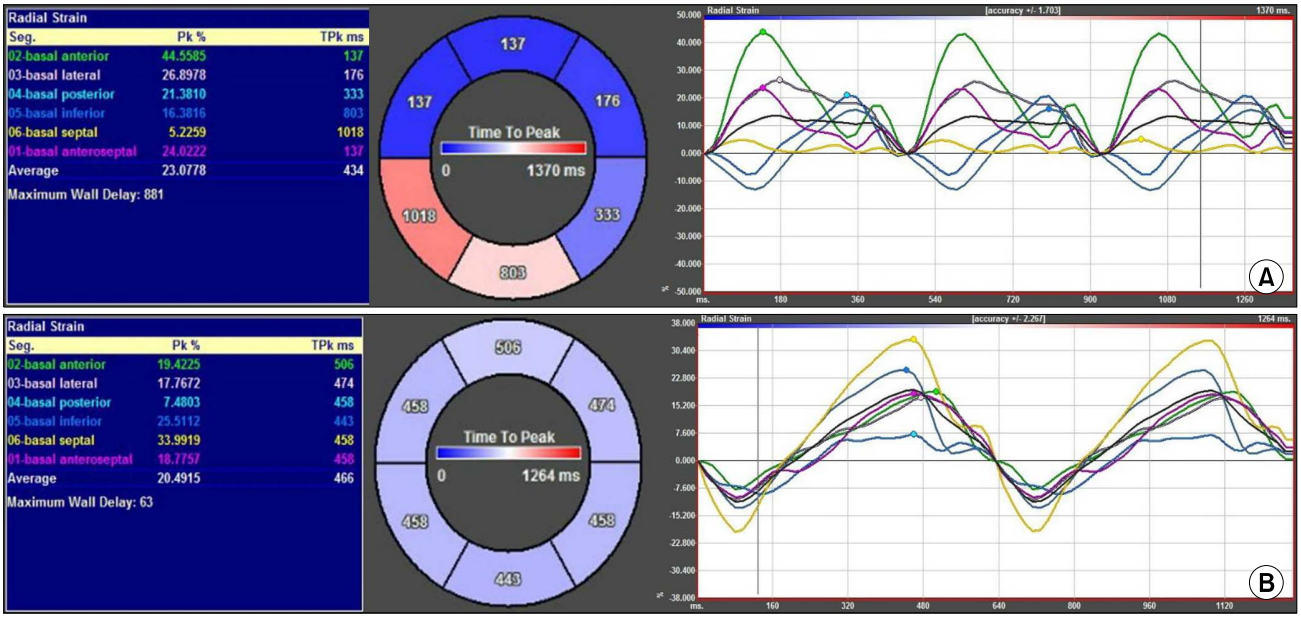

Fig. 1. TSI showed improvement of intraventricular synchrony after CRT. (A) Before CRT, TSI showed intraventricular dyssynchrony with a maximum wall delay of 881 ms in basal segments. (B) At 5 months after CRT, TSI demonstrated considerably improved intraventricular synchrony with a maximum wall delay of 63 ms in the same segments. TSI, tissue synchronization imaging; CRT, cardiac resynchronization therapy.

contraction. Septal-to-posterior wall motion delay was 236 ms by the M-mode (Table 1). Tissue synchronization imaging showed intraventricular dyssynchrony (Fig. 1). Thus, we decided to perform CRT during ECMO support.

The patient's weight and height were 4.7 kg and 65 cm, respectively. The operation was performed by left anterolateral thoracotomy and subxiphoid incision. The patient was positioned in the right semilateral decubitus position, and bipolar steroid eluting leads (CapSure Epi bipolar; Medtronic, Minneapolis, MN, USA) were selected for CRT. Through left anterolateral thoracotomy, the pericardium was opened along with the phrenic nerve anteriorly, and an LV lead was implanted on the lateral wall near the obtuse marginal branch of the left circumflex coronary artery. Through a subxiphoid incision, the right ventricular lead was implanted near the apex, and the right atrial lead was implanted near the SA node. A generator (Consulta CRT-P, Medtronic) was placed over the posterior sheath of the rectus abdominis. No significant bleeding was observed after CRT, and it was possible to wean the patient off ECMO in 2 days. At 7 weeks of follow-up, echocardiography showed an improved LV ejection fraction of 41.7%, decreased LV end diastolic dimension of 36.3 mm,

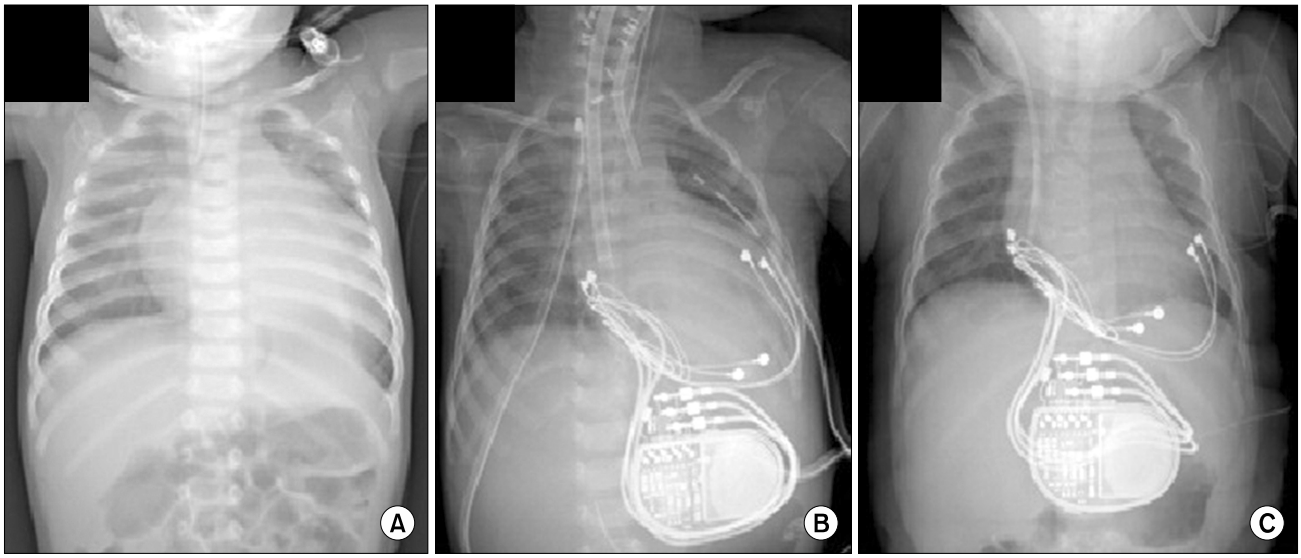

Fig. 2. Serial chest radiography showed the improvement of cardiomegaly. (A) At admission. (B) One day after cardiac resynchronization therapy during extracorporeal membrane oxygenation. (C) At 5 months of follow-up.

and moderate MR (Table 1). The blood level of BNP decreased to 146 pg/mL (Table 1). All cardiac medication could be discontinued by 7 weeks after CRT. At 5 months of follow-up, echocardiography with tissue synchronization imaging revealed a markedly improved LV ejection fraction of 59.7%, decreased LV end diastolic dimension of 33.4 mm, considerably improved LV synchrony, and mild MR (Table 1). The QRS duration decreased from 130 ms to 92 ms, and the blood level of BNP was normalized to 29 pg/mL (Table 1). Tissue synchronization imaging showed intraventricular synchrony (Fig. 1). Chest radiography demonstrated a reduced cardiac shadow (Fig. 2). Although her cardiac function has been nearly normalized, the patient still requires intensive care because of multiple organ dysfunction related to ischemic insult during the 2nd ECMO.

DISCUSSION

It is generally believed that CRT is the optimal treatment for adults with severe LV dysfunction and cardiac dyssynchrony [1,2]. The indication of CRT in adults with heart failure is expanding with the accumulation of knowledge obtained from well-designed studies [3]. CRT implantation is recommended in cases that meet the following criteria: (1) patients who have a LV ejection fraction less than or equal to 35%; (2) sinus rhythm, left bundle branch block with QRS duration equal to or longer than 150 ms; and (3) New York Heart Association class II, III, or IV symptoms. In contrast, there have been no definite guidelines for CRT in pediatric patients because of the lack of experience and the heterogeneous etiology of heart failure. Although several large cohort studies regarding CRT in pediatric patients have been reported [4,5], most reports are case series that focus on heart failure after surgery for congenital heart diseases, right ventricular pacing-induced cardiomyopathy, and heart failure in patients with a functional single ventricle. In our case, DCMP is a rare disease entity for CRT in the pediatric population, and it is extremely rare that CRT is performed during ECMO in infants despite several reports for adults [6,7]. It is challenging to perform CRT in a small infant during ECMO. Since the lead must be epicardially implanted in the operating theater in small infants, it could be difficult to safely implant the LV lead in an appropriate site and to avoid major bleeding complications during anticoagulation. To achieve these objectives, we decided to use two separate incisions. The LV lead was implanted through the left anterolateral thoracotomy, which allowed better exposure and less bleeding compared with the full sternotomy, and a subxiphoid incision was made

to implant the right atrial lead, the right ventricular lead, and the generator. We tried to implant the LV lead on the segment showing the maximum delay in preoperative echocardiography. However, there is not enough space to select the site for the implantation of the LV lead in small infants. Further, tissue synchronization imaging is not available in our operating room. Therefore, we implanted pacing leads at the site where the maximum increase of aortic pressure and the maximum decrease of QRS duration could be achieved during biventricular pacing. Following CRT, the echocardiographic parameters improved and cardiac medication could be discontinued 7 weeks later. Although the patient's cardiac function has been nearly normalized at the latest follow-up, the patient still requires intensive care because of multiple organ dysfunction related to ischemic insult during the 2nd ECMO. Had we considered CRT to be the primary option instead of heart transplantation, this baby would have been happier. Even though there is currently no definite guideline for CRT in pediatric patients, CRT must be strongly considered a treatment option in infants having heart failure with ventricular dyssynchrony.

According to our experience, CRT could be safely performed in small infants during ECMO, and the outcome of CRT might be promising in selected infants with end-stage heart failure.

CONFLICT OF INTEREST

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

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