Paracorporeal Ventricular Assist Device Implantation in a Patient with Transposition of the Great Arteries After Senning Operation

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Summary
We experienced a 33-year-old patient with D-looped transposition of the great arteries (D-TGA) and a history of Senning operation who was referred to our institute with cardiogenic shock and subsequently underwent urgent paracorporeal ventricular assist device (VAD) implantation, which was a first in Japan, that was eventually converted to a durable VAD. Central venous pressure was maintained relatively high to obtain VAD filling and recover end-organ dysfunction, given the migration of the inflow cannula due to rich trabeculae carneae of the anatomical right ventricle (systemic ventricle in this case).

Key words: Hemodynamics, Adult congenital heart disease, Mechanical circulatory support

The atrial switch operation for D-looped transposition of the great arteries (D-TGA), pioneered by Senning and Mustard, has improved the survival of patients with D-TGA, allowing them to survive until adulthood. However, various long-term complications including right ventricular dysfunction, tachyarrhythmias, baffle obstructions and shunts, and sudden death have been recognized recently.

The use of durable mechanical circulatory support (MCS) to salvage the failing systemic ventricle (right ventricle in this case) in patients with D-TGA who received atrial switch surgery has been introduced in Europe and the United States. However, there have been few such reports in Japanese patients, who tend to have a smaller body size. We experienced a patient with D-TGA after the Senning procedure who presented with hemodynamic deterioration accompanied by end-organ dysfunction, and was rescued by urgent implantation of a paracorporeal ventricular assist device (VAD) to the failing right ventricle.

Case Report

The patient was a 33-year-old woman who was hospitalized in a tertiary care hospital for worsening dyspnea after influenza infection. Despite intensive care, her end-organ dysfunction worsened and she was referred to our hospital for possible MCS therapy.

She was born in 1983 and was diagnosed as D-TGA type 1. She underwent balloon atrio-septostomy at birth and a left Blalock-Taussig shunt operation later on. Since her mean pulmonary artery pressure/mean aortic pressure was 0.20, the Senning operation (Figure 1A), or atrial switch operation, was conducted at the age of 1 year and 3 months instead of the Jatene operation.

Although she was followed as an outpatient, her right ventricular function gradually worsened and tricuspid regurgitation and development of ventricular tachyarrhythmia progressed due to the high afterload on the systemic ventricle (i.e., right ventricle in this case).

At age 32, she was hospitalized for further assessment. Right heart catheterization showed she had a mean right atrial pressure (RAP) of 12 mmHg, end-diastolic right ventricular pressure of 13 mmHg, and cardiac index of 2.0 L/minute/m². Transthoracic echocardiography revealed a right ventricular end-diastolic volume index of 177 mL/m², right ventricular ejection fraction of 25%, and moderate tricuspid regurgitation.

At age 33, she became severely ill due to influenza infection, resulting in hospitalization for hemodynamic deterioration and end-organ dysfunction, followed by mechanical ventilation, continuous infusion of inotropes, and continuous hemodiafiltration (CHDF). Her systolic blood pressure was 82 mmHg on admission to our hospital. Chest X-rays showed pulmonary congestion and cardiomegaly (Figure 2A). Electrocardiogram showed right axis deviation and complete right bundle branch block (Figure 2B). Transthoracic echocardiography revealed enlargement of the right ventricle (systemic ventricle) and moderate tricuspid regurgitation (Figure 1B-D). Her...
plasma level of N-Terminal pro B-type natriuretic peptide was 22,499 pg/mL, serum creatinine was 3.47 mg/dL, total bilirubin was 5.3 mg/dL, and lactate was 6.0 mmol/L.

Following the initial assessment and management, we at first considered the initiation of extracorporeal membrane oxygenation, which was soon abandoned due to bilateral femoral vein obstruction because of previous multiple surgeries. She then underwent urgent implantation of a paracorporeal VAD (Nipro VAD [Nipro, Osaka, Japan]). We managed to insert the inflow cannula at the apex of the right ventricle with rich trabeculae carneae toward the tricuspid valve under the support of transesophageal echocardiographic monitoring to determine the appropriate device positioning.

After the surgery, high central venous pressure (CVP) (>20 mmHg) was required to maintain the VAD pump filling and her blood pressure, probably due to cardiac tamponade which was improved by the exclusion of pericardial hematoma. In addition, computed tomography images showed a massive left thoracic hematoma and inflow

**Figure 1.** A: Schema of blood flow of D-looped transposition of the great arteries after Senning operation. B-D: Blood circulates as follows; IVC and SVC → LA → LV → PA, PV → RA → RV → Ao. Transesophageal echocardiography images on admission, showing enlarged anatomical right ventricle and right atrium and moderate tricuspid regurgitation. IVC indicates inferior vena cava; SVC, superior vena cava; LA, left atrium; LV, left ventricle; PA, pulmonary artery; PV, pulmonary vein; RA, right atrium; RV, right ventricle; Ao, aorta; and TR, tricuspid regurgitation.

**Figure 2.** A: Chest radiograph on admission showed pulmonary congestion and cardiomegaly. B: Electrocardiogram on admission showed atrial fibrillation and complete right bundle branch block.
cannula positioned toward the opposite direction of the tricuspid valve. Removal of the left thoracic hematoma by thoracoscopy was performed (Figures 3, 4). Due to the sucking that occurred, a still relatively high CVP (> 10 mmHg) was required to maintain preload and VAD flow and intravenous inotropes were concomitantly required for approximately 30 days after VAD implantation to improve end-organ dysfunction (Figure 4). There was no critical hemolysis during LVAD support.

After VAD implantation, her tricuspid regurgitation improved slightly and a decrease in RV size and improved left ventricular systolic function were achieved. She was extubated on the 38th day and withdrawn from CHDF on the 46th day. After rehabilitation, she was listed for heart transplantation on the 58th day. On the 141st day, the paracorporeal VAD was successfully converted to a durable VAD (Jarvik 2000® [Jarvik Heart, Inc., New York, NY]).

Discussion

In this report, we present a patient with a history of the Senning operation for D-TGA, who was rescued by urgent paracorporeal VAD implantation without extracorporeal membrane oxygenation due to multiple vessel obstruction. Relatively higher CVP was required to maintain appropriate VAD flow and recover end-organ dysfunction due to the inflow cannula migration as well as the rich trabeculae carneae of the right ventricle.

Operations for TGA: TGA is the most common cyanotic
heart disease in infancy.\textsuperscript{11,12} Surgical atrial septectomy was originally introduced in 1950s,\textsuperscript{33} followed by the atrial switch method such as the Senning operation\textsuperscript{11} and the Mustard operation.\textsuperscript{21} In the 1970s, Jatene introduced the arterial switch method, which allows the left ventricle to be a systemic ventricle and avoids multiple atrial incisions that predispose the patient to arrhythmia, showing relatively favorable midterm outcomes.\textsuperscript{14-16}

In this case, her left ventricular end-diastolic pressure was already high compared with the right ventricular end-diastolic pressure. The attending surgeons probably preferred the Senning method to the Jatene method, considering that her left ventricle could not tolerate her systemic afterload.

VAD implantation for TGA: Patients with congenital heart disease often have abnormal vascular connections (such as interrupted systemic veins, abnormal pulmonary arteries, or unfavorable vascular positioning), and case-specific optimal perioperative management with sufficient understanding of their previous surgical histories might be required.\textsuperscript{34} In our case, we decided not to use extracorporeal membrane oxygenation considering the obstruction of multiple peripheral vessels.

Furthermore, optimal positioning of the inflow cannula was challenging. There were rich trabeculae carneae among the remodeled right ventricle, which might have caused migration and suction of the inflow cannula. In cases of VAD implantation to the anatomical right ventricle, such as in patients with D-TGA or corrected-TGA, exclusion of trabeculae as widely as possible might be required during the operation to obtain sufficient VAD flow and prevent sucking.

Management to maintain optimal volume status was also required even after the removal of the intra-thoracic hematoma due to the sucked inflow cannula migration as well as the rich trabeculae carneae of the right ventricle. The required CVP level gradually decreased, most likely due to the improvement of anatomical left ventricular function via increased coronary flow, inotrope administration, and withdrawal of the mechanical respiration.

Due to her relatively small physique and rich trabeculae carneae of the right ventricle, we chose Jarvik 2000, which has a small device profile. Nevertheless, it required great effort to exclude the trabeculae as widely as possible to prevent device sucking. Thanks to such efforts, she did not experience any sucking events following implantation of Jarvik 2000.

Disclosure

Conflicts of interest: There is no conflict of interest to report related to this manuscript.

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