

Central-Approach Surgical Repair of Coarctation of the Aorta with a Back-up Left Ventricular Assist Device for an Infant Presenting with Severe Left Ventricular Dysfunction

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A two-month-old infant presented with coarctation of the aorta, severe left ventricular dysfunction, and moderate to severe mitral regurgitation. Through median sternotomy, the aortic arch was repaired under cardiopulmonary bypass and regional cerebral perfusion. The patient was postoperatively supported with a left ventricular assist device for five days. Left ventricular function gradually improved, eventually recovering with the concomitant regression of mitral regurgitation. Prompt surgical repair of coarctation of the aorta is indicated for patients with severe left ventricular dysfunction. A central approach for surgical repair with a back-up left ventricular assist device is a safe and effective treatment strategy for these patients.

Key words: 1. Coarctation
2. Infant
3. Heart failure
4. Ventricular assist device

CASE REPORT

A two-month-old male infant presented with tachypnea, diaphoresis, and poor feeding. He was born at full term with no perinatal medical problems at a primary care hospital. His primary care physician noted a cardiac murmur, but recommended reevaluation at the age of two months because his physical examination did not reveal any other abnormalities. The patient did well for the first month, but experienced progressive worsening of the aforementioned symptoms. The patient was referred to our hospital for further evaluation. On physical examination, the patient's skin was cold and wet. He

was tachypneic (60 respirations/min) and tachycardic (155 beats/min). A systolic murmur was heard at the cardiac apex and the femoral pulses were weak. The upper extremity blood pressure was approximately 20 mmHg higher than the lower extremity blood pressure. No evidence of infection or inflammation was found based on blood chemistry panels, a complete blood count, and serologic markers. Creatine kinase and creatine kinase-MB levels were within normal limits. However, troponin-T was elevated (0.075 ng/mL; reference range, 0.0–0.014 ng/mL). The serum N-terminal of the pro-hormone was 9,601 pg/mL (reference range, 0–154 pg/mL). A chest X-ray showed cardiomegaly and mild pulmonary

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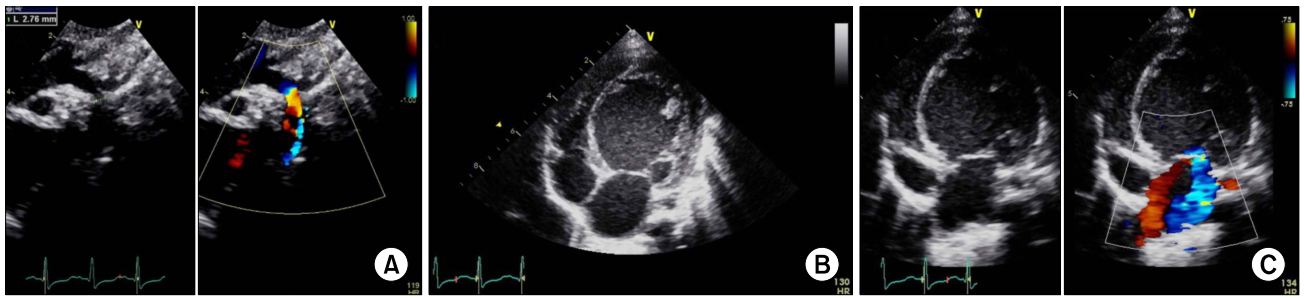


Fig. 1. Preoperative echocardiography demonstrating (A) coarctation of the aorta, (B) a markedly dilated left ventricle, and (C) grade III/IV mitral regurgitation.



Fig. 2. Postoperative echocardiography demonstrating (A) the repaired non-obstructive aortic arch, (B) improved left ventricular enlargement, and (C) mitral regurgitation.

edema. Sinus tachycardia and left ventricular hypertrophy with strain was observed on an electrocardiogram. Echocardiography revealed coarctation of the aorta (diameter, 3 mm; peak pressure gradient, 30 mmHg; mean pressure gradient, 14 mmHg), a patent ductus arteriosus (PDA) 1.5 mm in size, a patent foramen ovale (PFO), and grade III/IV mitral regurgitation. The left ventricle was globally hypokinetic (ejection fraction, 13%; shortening fraction, 8%–10%) and markedly dilated. The left ventricular end-diastolic and end-systolic dimensions were 36 mm (138% of normal range) and 31 mm (188% of normal range), respectively (Fig. 1).

Intravenous infusion of prostaglandin E1 was initiated, but the patient's peripheral perfusion and left ventricular function did not improve. Therefore, an urgent operation was scheduled. A median sternotomy approach was chosen for possible intraoperative or postoperative mechanical circulatory support. Cardiopulmonary bypass (CPB) was commenced through bicaval venous and innominate arterial cannulation. The aortic arch was repaired using resection of the aortic coarctation and an extended end-to-side anastomosis technique under regional

antegrade cerebral and myocardial perfusion. Under cardioplegic myocardial protection, the PFO was closed. Aortic cross-clamp time was five minutes. The patient was weaned from CPB with barely tolerable vital signs. No pressure difference was found between the upper and lower extremities. Post-repair echocardiography demonstrated severe left ventricular dysfunction with a left ventricular ejection fraction of 15%. Epicardial echocardiography was carried out with extreme caution, but during echocardiographic evaluation, the patient's vital signs slowly deteriorated. CPB was resumed and was changed over to left ventricular assist device (LVAD) support. The LVAD system was constructed with an 18-French left atrial drainage cannula placed through the right superior pulmonary vein, a 10-French ascending aortic cannula, and a centrifugal pump. Left ventricular function improved gradually. On postoperative day 5, the left ventricular ejection fraction had recovered to 40% with minimal LVAD support (200 mL/min) and the LVAD was decannulated. The sternum was closed on postoperative day 6 and the patient was weaned from ventilator on postoperative day 9. A follow-up echo-

cardiogram revealed recovered left ventricular function (ejection fraction, 50%) with regression of mitral regurgitation (grade I/IV) (Fig. 2). He was discharged home on postoperative day 33 without any complications.

DISCUSSION

Patients with coarctation of the aorta usually present in infancy with symptoms of congestive heart failure or with hypertension later in childhood. Neonates and infants are symptomatic when blood flow to the lower body is restricted due to PDA closure or increased afterload on the left ventricle, resulting in acute congestive heart failure [1]. A clinical review of patients who underwent neonatal repair of coarctation revealed that neonates with an intact ventricular septum had more significant global left ventricular dysfunction than those with ventricular septal defects [2], suggesting that the pressure load on the left ventricle can be the cause of left ventricular dysfunction. When the heart is exposed to pressure load, concentric hypertrophy develops. As adverse remodeling develops under a long-standing pressure load, eccentric hypertrophy with heart failure occurs. Some reports have suggested that coarctation in adult patients can be an occult cause of dilated cardiomyopathy [3]. However, aortic coarctation complicated by severely depressed left ventricular function in very early infancy, as occurred in our patient, has rarely been reported [4]. Our patient's left ventricular dysfunction and mitral regurgitation recovered after relief of the pressure load caused by coarctation of the aorta. Left ventricular dysfunction can be completely restored, even in adult patients diagnosed with dilated cardiomyopathy [3]. For patients with severe left ventricular dysfunction, coarctation of the aorta should be considered as a possible cause that is reversible.

Repair of aortic coarctation through left thoracotomy is the standard procedure for neonates and infants who have no aortic arch hypoplasia or intracardiac anomalies. However, when the patient's left ventricular function is severely depressed, the patient cannot tolerate the sudden increased afterload on the left ventricle during the cross-clamping of the aortic arch. If the patient deteriorates hemodynamically during the clamp-and-sew procedure, it is extremely difficult to perform cannulation for mechanical circulatory support, espe-

cially in neonates and small infants. In order to avoid this risky situation, we used a central approach and repaired the aortic coarctation under CPB and regional cerebral perfusion. For patients with cardiogenic shock or hemodynamic instability due to severely depressed left ventricular function, the safety of the central approach outweighs the risks related to CPB and regional cerebral perfusion. If necessary, CPB can be easily converted to a LVAD. In order to minimize operation-related myocardial damage, we attempted to reduce the aortic cross-clamp time. Coronary perfusion and non-working beating of the heart were maintained during the aortic arch repair. The ascending aorta was only clamped to close the PFO. The PFO might be beneficial to vent the left side of the heart for patients with extracorporeal membrane oxygenation (ECMO) support. However, for patients with LVAD support, if left atrial blood is drained vigorously by a LVAD, right atrial blood can be shunted through the PFO, resulting in systemic hypoxemia.

Perioperative ECMO support with repair of the aortic coarctation under ECMO could have been another option for our patient. ECMO has the advantages of peripheral cannulation, biventricular support, and respiratory support. Currently, ventricular assist devices are preferred for patients with preserved pulmonary function because they are associated with less inflammation, less need for anticoagulation, and better clinical outcomes [5].

In conclusion, coarctation of the aorta can be complicated by severe left ventricular dysfunction, even in early infants. Myocardial dysfunction can be reversed with surgical repair of the aortic coarctation, and a central approach with a back-up LVAD is a safe and effective treatment strategy for these patients.

CONFLICT OF INTEREST

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

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