

Case Report

Successful Intra-postoperative Extracorporeal Circulatory Support with Atrial Communication for Treatment of Anomalous Left Coronary Artery from the Pulmonary Artery: A Case Report

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We describe a successful case of surgical treatment for anomalous left coronary artery from the pulmonary artery (ALCAPA) syndrome with severe left ventricular dysfunction. Because of the severe left ventricular dysfunction, we planned to use an extracorporeal membrane oxygenation for heart support until a satisfactory recovery had been established. The left ventricular function significantly recovered in a few days, and the patient could be discharged without any complications.

Key words: coronary circulation, anomalous left coronary artery from the pulmonary artery, extracorporeal circulation, extracorporeal membrane oxygenation

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a relatively rare congenital heart disease, representing a mere 0.5% of all of congenital heart diseases [1]. Because the myocardium in a case of ALCAPA does not have adequate oxygen supply and may suffer from ischemic injury, the treatment for ALCAPA is to surgically establish a 2-coronary system. Risk factors for perioperative death of ALCAPA patients are low-output syndrome (LOS) and acute cardiac infarction (AMI) before the operation. But successful repair of ALCAPA usually results in a marked recovery of the contractile function in late follow-up, with the left ventricular function returning to normal or nearly normal levels within 1 to 2 years after repair

[2-5]. Mechanical support for the left ventricle may be required in some children who are unable to be weaned off of cardiopulmonary bypass (CPB) or who require very high-dose inotropic support to avoid other organ injuries. ALCAPA patients with LOS or AMI need a strategy to recover their cardiac function in a safer way after their operations.

Case Report

A 3-month-old girl, delivered at a gestational age of 41 weeks with a birth weight of 2,680 g, suffered from tachypnea and wheeze. She visited a neighboring hospital and was diagnosed by echocardiography as having severe heart failure. A chest X-ray revealed cardiomegaly (CTR 70%) and atelectasis of the right upper pulmonary lobe (Fig. 1). Her aortography showed no left coronary artery (Fig. 2). The left coronary artery was suspected of branching off

from the pulmonary artery, but pulmonary arteriography could not be performed because of her bad general condition.

Her respiratory distress and cardiac failure worsened despite medical treatment. At the age of 4 months, she was referred to our institute for semi-urgent surgical intervention. After being transferred to our hospital, she suffered suddenly from bradycardia and cardiopulmonary arrest. Though a few minutes of cardiopulmonary resuscitation (CPR) was initially successful, her circulatory condition was unstable, and repetitive CPR for one and a half hours was required. After CPR, her cardiochogra-

phy revealed severe left ventricular dysfunction: FS was less than 10%, and there was both remarkable dilation of the left ventricle (LVDd = 33.1 mm) (Fig. 3) and severe mitral valvular regurgitation. A blood examination showed an increase in creatinine kinase and lactate dehydrogenase to 378 IU/l and 390 IU/l, respectively. Although her light reflex was severely impaired immediately after the recovery of her heart-beat, it gradually recovered to a normal reaction time. In addition, there was no neurological finding



Fig. 1 Preoperative chest X-ray revealed severe cardiomegaly (CTR 70%) and atelectasis of the right upper pulmonary lobe.

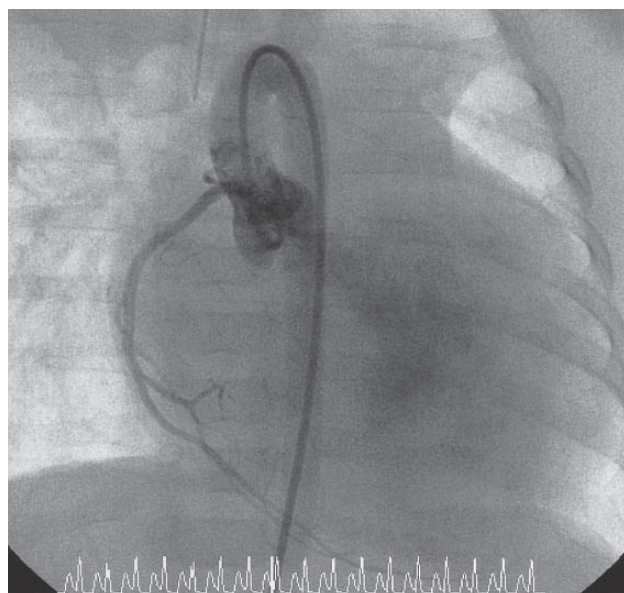


Fig. 2 Ascending aortography showed no left coronary artery.

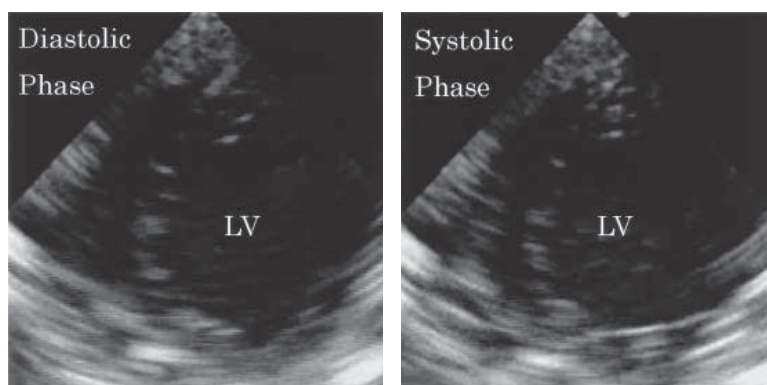


Fig. 3 Cross-sectional view of the left ventricular preoperative cardiochography revealed severe left ventricular dysfunction (FS = 10%, LVDd = 33.1 mm). LV, left ventricle.

to suspect lasting neurological damage. We decided to operate immediately.

The operation was performed via median sternotomy. It was found that the coronary artery branched off from the posterior wall of the pulmonary artery, which had shifted slightly to the left side. Therefore, the Takeuchi procedure, a reasonable method for someone with this anatomy, was performed. After the procedure, atrial communication was left open because this was important for a biventricular assist in this case. The adjustable atrial communication was created as follows. A mattress suture was placed at the center of the atrial septal defect and ligated, and a pledgeted purse-string suture was placed around the atrial septal defect. The string, which could be used by ligation to close the atrial septal defect later without atriotomy, was passed through the right atrial wall to the outside of the heart (Fig. 4). A mitral valvular plasty was ruled out because a longer aortic cross-clamp needed to be avoided due to the patient's low left ventricular function and the episode of cardiopulmonary arrest. We converted her to extracorporeal membrane oxygenation (ECMO) following the CPB and did not try to wean her from it.

Her left ventricular function recovered gradually but markedly day by day. On the 7th post-operative-day (POD), she could be weaned from the ECMO, and the atrial septal defect was closed by ligating the purse-string suture without atriotomy. Postoperative echocardiography just after weaning from the ECMO revealed marked recovery of the left ventricular

function: FS increased to 23% and LVDD decreased to 23.3 mm (Fig. 5). Chest X-ray on the 14th POD showed a CTR of 54%, and the atelectasis of the right upper pulmonary lobe had disappeared (Fig. 6). She was discharged on the 22nd POD without any

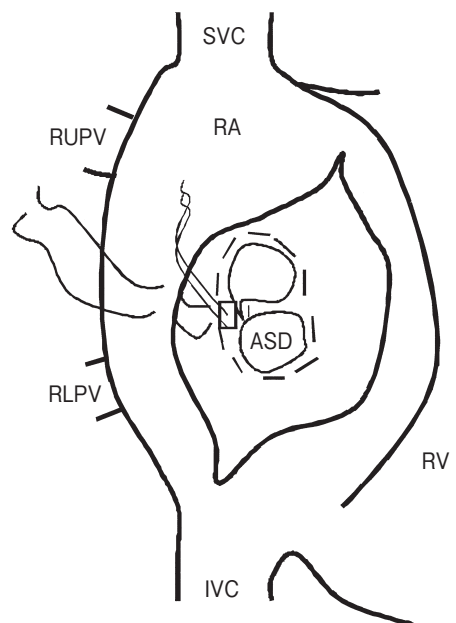


Fig. 4 A mattress suture was placed at a center of the atrial septal defect; a pledgeted purse-string suture was placed around the atrial septal defect; and the string was passed through the right atrial wall to the outside of the heart. ASD, Atrial septal defect; IVC, Inferior vena cava; RA, Right atrium; RLPV, Right lower pulmonary vein; RUPV, Right upper pulmonary vein; RV, Right ventricle; SVC, Superior vena cava.

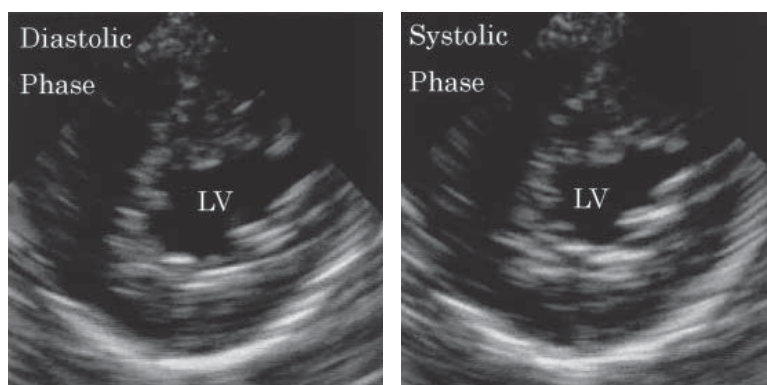


Fig. 5 Cross-sectional view of the left ventricular postoperative cardioechocardiography just after weaning from the ECMO revealed marked recovery of left ventricular function (FS = 23%, LVDD = 23.3 mm). LV, left ventricle.

complications (Fig. 7). At 12 months after the operation, the patient is clinically doing much better.

Discussion

As mentioned above, generally, if a 2-coronary-system is successfully established, postoperative cardiac functional recovery is excellent in infants with



Fig. 6 Postoperative chest X-ray revealed a significant decrease of CTR (54%) and disappearance of atelectasis of the right upper pulmonary lobe.

ALCAPA syndrome. Most post-operative deaths are either intraoperative deaths or early post-operative deaths due to cardiac failure. Sauer *et al.* reported 6 operative deaths among 31 ALCAPA cases on whom a coronary transfer from the pulmonary artery to the aorta or a coronary bypass with the left subclavian artery was performed; these included 5 intraoperative deaths and 1 death 12 h after the operation. They also noted that preoperative factors associated with a statistically significant higher perioperative mortality were young age at operation, left and balanced types of coronary circulation, and electrocardiographic signs of extensive acute myocardial infarction, namely, marked ST elevation (≥ 0.2 mV in at least 2 leads) and left-axis deviation on electrocardiograms of an extreme right-dominant type of coronary circulation [6].

Although extra-corporeal support after operation for ALCAPA has been occasionally reported [7-8], there is only 1 article to date that investigates perioperative extracorporeal support for ALCAPA infants. Its outcome seems to be relatively good. Del Nido *et al.* reported 7 cases of ALCAPA infants who needed left-ventricular assist device support after the reestablishment of a 2-coronary-system. They said that 1 child died shortly after the start of the left-ventricular assist, and another died of arrhythmia within 24 h of a successful decanulation; the

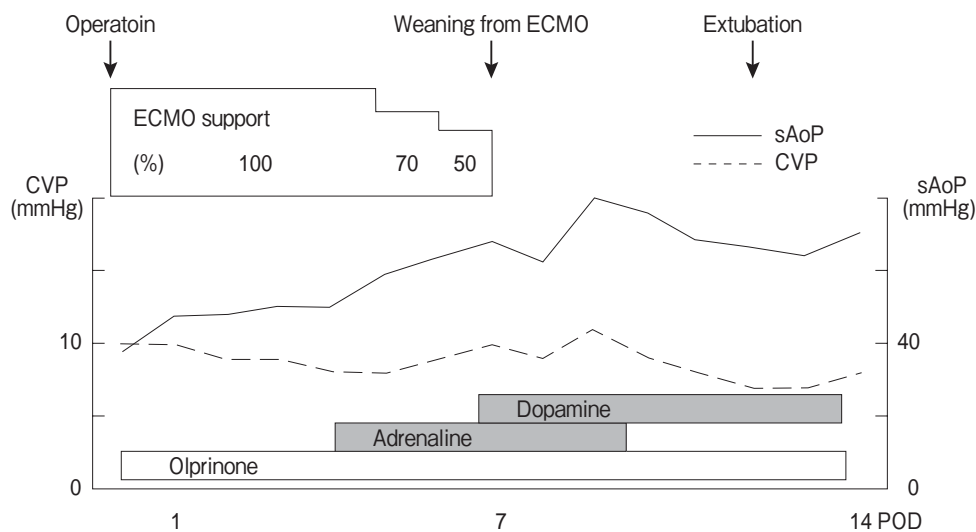


Fig. 7 Clinical course. CVP, Central venous pressure; ECMO, Extracorporeal membrane oxygenation; POD, Post-operative day; sAoP, Systolic aortic blood pressure.

remaining 5 children survived with significant improvement in left ventricular function [9]. The rate of surviving discharge for primary cardiac indications of LVAD or ECMO in children was reported to be 40.6% to 45% [10–11]. Because significant recovery from cardiac dysfunction can be achieved for almost all ALCAPA infants, these rates may become much higher.

We performed the operation with intention of intra-postoperative ECMO support because the patient was expected to have suffered from severe ischemic damage preoperatively. Because ECMO cannot provide left ventricular support, atrial communication is useful in order to decrease the preload of the left ventricle by venting the blood from the left atrium. We could minimize inotropic support using ECMO, and the maximum dose of inotropes was adrenalin 0.05 μ g, dopamine hydrochloride 5 μ g and olprinone hydrochloride 0.4 μ g. Smooth recovery of cardiac function was achieved. Aggressive extracorporeal support (LVAD or ECMO) decreases the perioperative mortality of ALCAPA infants who suffer from preoperative severe myocardial injury, with conditions such as cardiopulmonary arrest, acute myocardial infarction, and severe low output syndrome. We recommend that unreasonable weaning from CPB should not be attempted because a rest cure for the myocardium will lead to better mortality and cardiac function after the operation. For ALCAPA infants with severe ischemic myocardial damage, questions concerning which patients should be given intra-postoperative extracorporeal support and which devices should be used (LVAD or ECMO) are still controversial, and further clinical examination and discussion is needed.

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