

—Reports on Experiments and Clinical Cases—

Successful treatment of anomalous origin of the left  
coronary artery from the pulmonary artery in  
a 5-week-old male infant

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**Abstract**

A 5-week-old male infant who was referred to our hospital because of tachypnea and poor feeding. An electrocardiogram showed a deep Q wave in lead  $aV_L$ , negative T waves in leads I, II, III,  $aV_F$  and  $V_6$  and a positive T wave in  $V_1$ . Echocardiography revealed severely impaired left ventricular function. Aortography confirmed with a diagnosis of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). Takeuchi's procedure was performed and the patient maintained postoperatively on assisted circulation for 7 hours even though sternal closure delayed until 7 days post operatively. His left ventricular function showed and marked improvement gradually. (J Nippon Med Sch 1998 ; 65 : 312—315)

**Key words:** anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). The brand-white-garland syndrome (BWG syndrome), two-coronary system

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), or the Brand-White-Garland syndrome is a rare congenital heart disease<sup>1,2</sup> that can lead to acute myocardial infarction or cardiac failure in infants, or a heart murmur or other abnormality detected by electrocardiography in adults. Signs of chronic myocardial ischemia or cardiac failure generally appear 3 weeks after birth. However, symptoms such as irritability, failure to thrive and tachypnea were often overlooked. The diagnosis and immediate treatment of ALCAPA in newborns are very rare. We diagnosed and treated a 5-week-old male infant with severely impaired left ventricular function and ALCAPA.

**Case Report**

A 5-week-old male Japanese infant had shown weight gain until 1 day before admission. He was admitted because of tachypnea and poor feeding. There was no family history concerning heart disease. His weight on admission was 3924 g. His features were normal, and his skin appeared slightly icteric. He was experiencing moderate respiratory distress, with slight intercostal retractions. There was no significant murmur, but heart sounds were distant. Breath sounds were rough, but no wheezing or crackles were audible. He had hepatomegaly, because the liver was palpable 3 cm below the right costal margin. A chest X-ray showed marked cardiomegaly (cardiothoracic ratio: 0.69) with an increase in pulmonary vascularity (**Fig. 1**). An elec-

trocardiogram showed a deep Q wave in lead  $aV_L$  and decrease of magnitude of the R wave from  $V_4$  to  $V_6$ . T wave were negative in leads I, II, III,  $aV_F$  and  $V_6$  and a positive T wave in  $V_1$ . (**Fig. 2**). These findings indicated the presence of an anterolateral myocardial infarction. Echocardiography showed poor contractility of the left ventricle (ejection fraction: 0.39) associated with severe hypokinesis in the anterolateral left ventricular wall (**Fig. 3**). The left atrium was enlarged due to moderate mitral regurgitation. One coronary originating from the aorta was identified, another coronary artery branched from the left posterior side of the pulmonary artery. The results of cardiac catheterization on day 3 suggested the presence of ALCAPA. Aortography showed a dilated right coronary artery originating from the aorta, and the left coronary artery was opacified from the right coronary artery via collaterals (i.e., from the conus artery to the LAD, from the RV branch to the LAD, and from the terminal branch to the LCX). These collaterals drained into the left posterior side of the pulmonary artery (**Fig. 4**). The patient required treatment for cardiac failure. Dopamine was infused just before surgery. Takeuchi's procedure, in which an aortopulmonary windows is created together with an intrapulmonary tunnel<sup>13</sup> was performed. After the aorta was unclamped, the patient was supported with a left ventricular assist pump for 7 hours. LV function was monitored using real-time transesophageal echocardiography. The chest was left open for 7 days because of hemodynamic instability. The

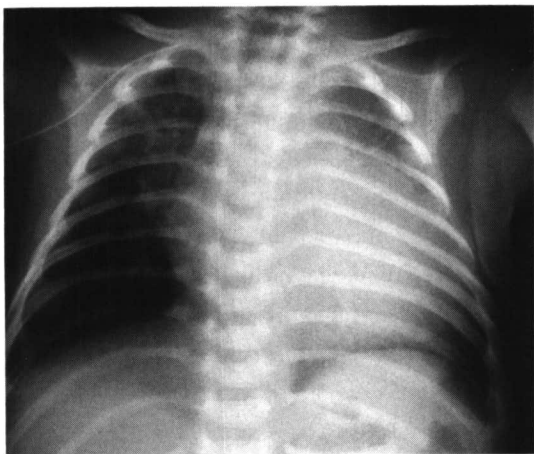


Fig. 1 chest X-ray. It showed cardiomegaly (cardiothoracic ratio was 0.69) with pulmonary congestion.

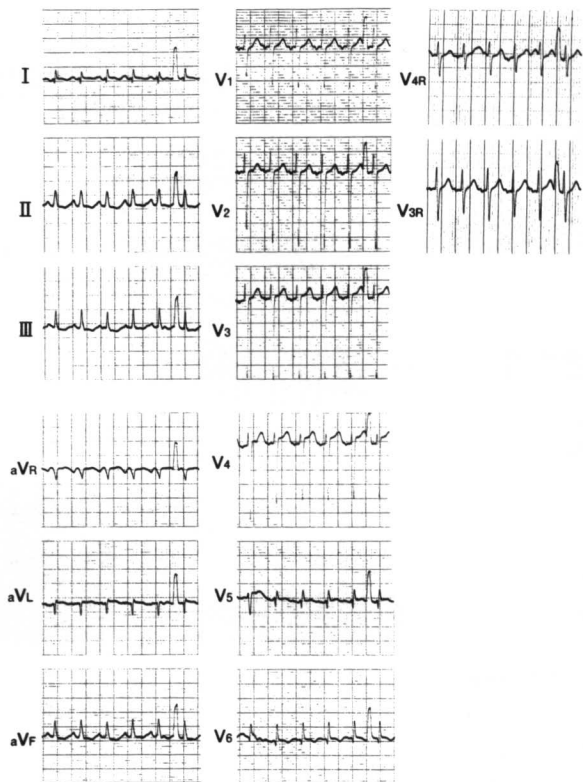


Fig. 2 An electrocardiogram showed a deep Q wave in lead  $aV_L$ , decreased magnitude of the R wave from leads  $V_4$  to  $V_6$ . Negative T waves in leads I, II, III,  $aV_F$  and  $V_6$  and a positive T wave in  $V_1$ .

left ventricular function showed gradual and marked improvement.

## Discussion

Edwards, Baue and others classified of ALCAPA by pathologic physiology and clinical course<sup>4</sup>. In our patient, the disease process was in the ischemic phase at the time of diagnosis. ALCAPA is rare, and because early signs, such as sweating, irritability, failure to thrive or tachypnea, are often overlooked, diagnosis is often delayed. The present case was diagnosed when the patient was 5 weeks old.

In the last decade, surgical procedures have improved considerably, allowing radical correction of the disease. Historically, palliative operations such as pericardial poudrage<sup>5</sup>, banding of pulmonary artery<sup>6</sup> and ligation of anomalous left coronary artery<sup>7</sup> were performed. The left coronary artery should be reimplanted into the aorta<sup>8,9</sup> if technically possible. If this is not feasible, an alternative approach which are tunneling operation<sup>3,10</sup> and coro-

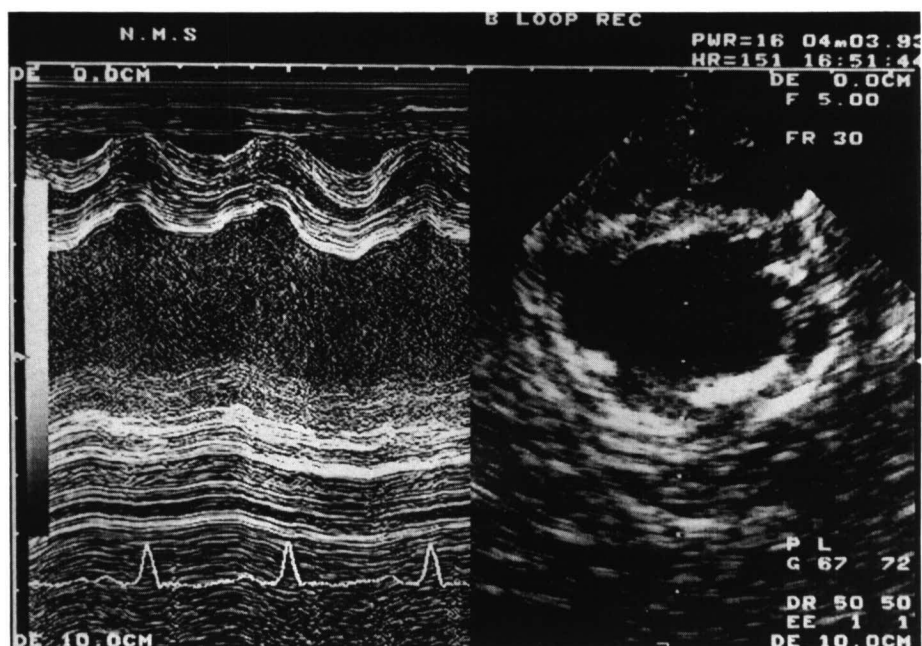


Fig. 3 M-mode and B-mode echocardiography showed poor contractility of the left ventricle (ejection fraction: 0.39) and severe hypokinesia of the left ventricular wall.

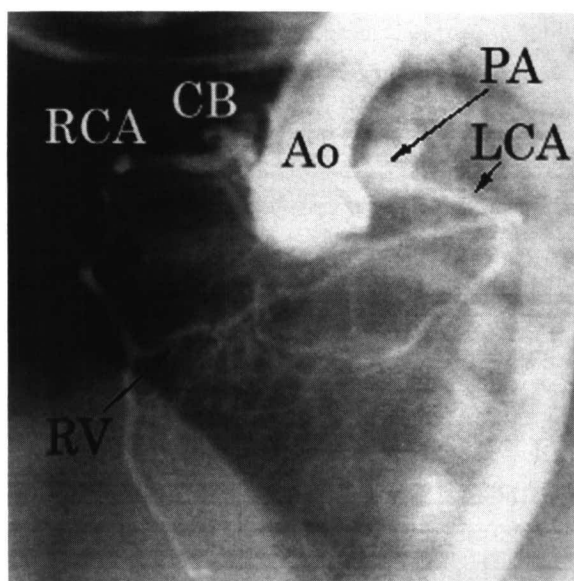


Fig. 4 Aortography (RAO 30°, cranial 20°) showed that the right coronary artery (RCA) arose normally from the aorta (Ao) and communicated via abundant intercoronary anastomoses with the left coronary artery (LCA), which originated from the pulmonary artery (PA). CB: conus branch, RV: right ventricular artery

nary artery bypass grafting<sup>11,12</sup> should be considered. In these procedures, occlusion of bypass graft of tunnel is the most serious complication. El-Said used saphenous vein grafts to restore perfusion into the left coronary artery; long-term results were

very poor with two occluded grafts<sup>13</sup>. Donaldson demonstrated that 4 out of 5 grafts were patent after 14 years<sup>14</sup>. Combining the reports of Moodie and Chiarello, 11 of 15 grafts were patent after 12 years<sup>15,16</sup>. Use of the internal mammary artery should improve the repair of ALCAPA. Patency of tunneling repair has not been studied, however, Migdley reported a patent tunnel conduit 14 months after a repair in an infant<sup>17</sup>. In this case, we choose the tunnel operation with Takeuchi's method, because graft veins and arteries were very small for performing graft operation and did not expect of graft patency for long time.

The ideal time for surgery in patients with ALCAPA has not been determined. A recent study has suggested that the preoperative severity of left ventricular dysfunction, not age, predicts operative mortality<sup>18</sup>. Evidence suggests that surgery should be performed immediately in patients with severe left ventricular impairment when medical treatment is ineffective. We believe the present case falls into this category. Left ventricular wall motion was markedly decreased and serum levels of myocardial enzymes were elevated preoperatively in the patient, suggesting the possibility of subendocardial infarction. Therefore, we wanted to establish adequate coronary perfusion as early as possible to halt the ischemia-induced progression of LV impairment

and to rescue jeopardized tissue.

ALCAPA is suffered from a gradual progression of ischemia resulting from decreased pulmonary artery pressure after perineonative period; the left coronary flow is supplied from the right coronary artery via collaterals. The heart is in the condition defined as deteriorated myocardial contractility, resulting from decreased oxygen consumption by myocytes, in association with morphologic alterations<sup>19</sup>. Shivalkar et al<sup>19</sup> have suggested that prolonged hypoperfusion may cause a subcellular adaptive response. In the presence of such a structural adaptation, LV function may not recover immediately after surgery. Therefore, our patient was maintained for a prolonged period on an assist circulation. Previous studies have suggested that this device is useful after re-establishment of coronary perfusion in patient with ALCAPA<sup>20,21</sup>.

Takeuchi's procedure was successful in our patient with ALCAPA. In the patient presented, severely impaired LV function exhibited improvement gradually. Prolonged treatment with an assist circulation and delayed sternal closure were useful. The present findings indicate that ALCAPA should be suspected in an infant whose LV function is severely impaired and that surgery should be performed immediately after diagnosis in such a patient.

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