CASE REPORT

Journal of Indonesian Thoracic Cardiac and Vascular Surgery *(JINATCVS)* 2024,
Volume 1, Number 1: 10-14

Published By :
The Indonesian Association of Thoracic and Vascular Surgeons

# ALCAPA repair using Takeuchi procedure in infants with cardiomyopathy and low ejection fraction, without mechanical circulatory support: case report

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Received: 2024-01-03
Accepted: 2024-02-10
Published:2024-03-12

## ABSTRACT

**Introduction:** Bland-White-Garland syndrome, often referred to as the abnormal origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA), is a rare congenital cardiac disease that causes left ventricular (LV) failure and myocardial ischemia. Reimplantation of the ALCAPA into the aorta and the Takeuchi operation, which creates an intrapulmonary baffle, are two recent surgical procedures that have drastically decreased death rates to less than 20%. This study aimed to report a case of ALCAPA repair using the Takeuchi procedure in infants with cardiomyopathy and low ejection fraction without mechanical circulatory support.

**Case description:** A male infant 4 months old presents with choking when bottle-feeding, leading to cyanosis and shortness of breath. The patient was treated in the intensive care unit for several days, and the imaging showed dilated cardiomyopathy leading to ALCAPA. MSCT findings showed an ALCAPA with a dilated RCA diameter of 2.2 mm. Ventricular Fibrillation and cardiopulmonary resuscitation were reported during the Pediatric Intensive Care Unit (PICU) stay, then the patient decided to go for the surgical procedure. The Takeuchi procedure was done with a pericardial baffle from the left coronary going through the ascending aorta. The patient was transferred to the ICU for open chest management for four days. The patient was still hemodynamically stable afterward without mechanical circulatory support, and the LV dysfunction could still be tolerated by optimizing inotropes and after-load reduction medicines.

**Conclusion:** ALCAPA leads to myocardial ischemia and left ventricular dysfunction, posing significant risks, especially in infancy. Timely surgical intervention is essential to improve outcomes, with recent advancements in surgical techniques showing promising results in reducing mortality rates in case of severe LV dysfunction without mechanical circulatory support (ECMO or LVAD).

**Keywords:** ALCAPA, low ejection fraction, mechanical circulatory support, Takeuchi procedure.
**Cite This Article:** Susanti, D.S., Wardoyo, S., Makdinata, W., Dwiprakoso, A.N., Muzakky, A., Lintangella, P., Namretta, L. 2024. ALCAPA repair using Takeuchi procedure in infants with cardiomyopathy and low ejection fraction, without mechanical circulatory support: case report. *Journal of Indonesian Thoracic Cardiac and Vascular Surgery* 1(1): 10-14.

## INTRODUCTION

Bland-White-Garland syndrome, often referred to as the aberrant origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA), is a rare congenital cardiac abnormality that was initially reported by Edward Bland, Paul Dudley White, and Joseph Garland in 1933.[1] The development of collateral circulation from the right coronary artery (RCA) to the ALCAPA as a result of this abnormality causes left ventricular (LV) dysfunction and myocardial ischemia. With an estimated incidence rate of 1 in 300,000 live births, ALCAPA is a rare congenital disability that accounts for 0.25–25% of all congenital cardiac disorders.[2,3] It is divided into two forms: infantile and adult. The infantile form carries a grim prognosis if untreated surgically due to the absence of coronary collaterals. However, the adult variant typically involves the presence or development of collaterals, ensuring sufficient blood flow to the myocardium.

As a result, depending on the degree of the defect and the rate at which LV dysfunction progresses, patients may have varying symptoms. Some kids could not exhibit any symptoms for a long time and only develop LV dysfunction in their later years. ALCAPA syndrome in infants can be fatal, with patients displaying signs including silent myocardial ischemia, mitral regurgitation (MR), myocardial infarction, or left ventricular dysfunction that can lead to sudden cardiac death (SCD).[1,4] Prompt diagnosis and timely surgical intervention are crucial, given that the mortality rate in infants with ALCAPA, delayed diagnosis and treatment, can surpass 90%, especially within the initial year of life.[5]

The construction of an intrapulmonary baffle or the reimplantation of the

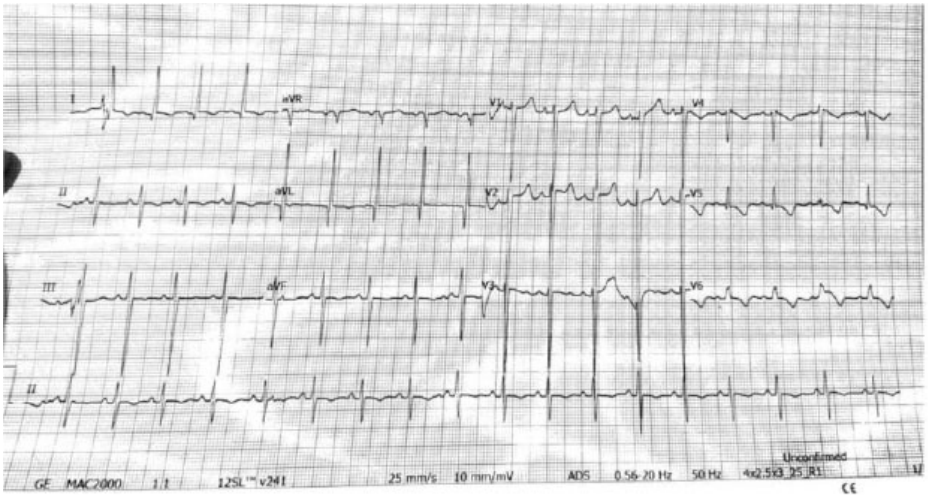

**Figure 1.** Preoperative electrocardiogram.

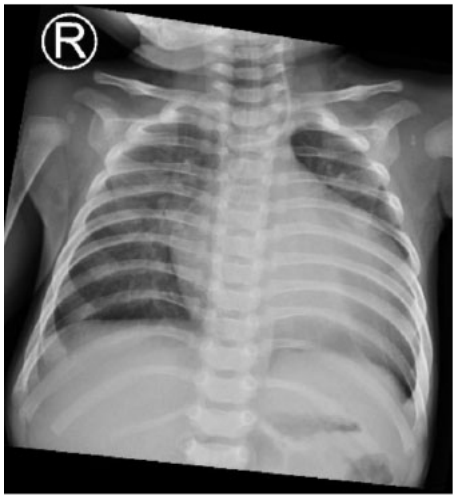

**Figure 2.** Preoperative chest X-ray.

ALCAPA into the aorta are two recent surgical procedures that have dramatically decreased death rates to less than 20%.[1] The Takeuchi method, first reported in 1979, entails making an intrapulmonary tube and an aortopulmonary window.[4] This causes the physiological repair of ALCAPA by rerouting blood flow from the aorta to the opening of the native left coronary artery. Usually, pericardial tissue, synthetic material, or flap tissue from the pulmonary artery wall was used to establish the intrapulmonary partition.[6] This study aimed to report a case of ALCAPA repair using the Takeuchi procedure in infants with cardiomyopathy and low ejection fraction without mechanical circulatory support.

## CASE DESCRIPTION

We describe a case of a 4-month-old male infant who choked on a bottle, resulting in cyanosis and dyspnea. The patient was diagnosed with aspiration pneumonia and spent several days in the intensive care unit. Imaging revealed dilated cardiomyopathy leading to ALCAPA, pneumonia caused by a viral infection, Bland White Garland Syndrome, and human immunodeficiency virus. Ventricular fibrillation was noted during the patient's stay in the PICU, and the patient received cardiopulmonary resuscitation before spontaneous circulation returned. The patient was treated with intravenous nitroglycerine, which can not be withdrawn. The patient has been diagnosed with Acquired Immunodeficiency Syndrome (AIDS) from his mother, who was diagnosed with

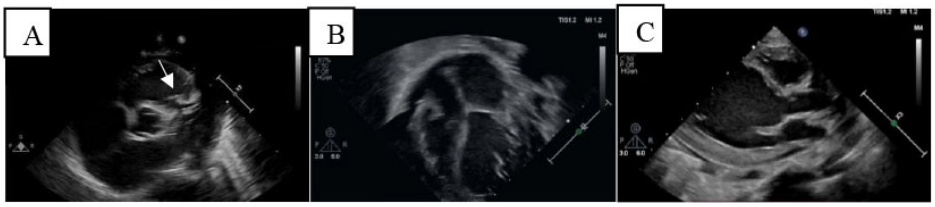


**Figure 3.** Preoperative echocardiography. A. Left main coronary artery from PA, B. Four chamber view and C. PLAX view showing dilated LV and small aorta.

AIDS and received antiviral therapy for two years; CD4 evaluation was 50+. The patient is first born through Cesarean delivery, with no active resuscitation. There is no history of alcohol consumption and smoking cessation during pregnancy. The family did not have any hereditary or congenital illnesses. Digoxin 0.02 mg twice daily, furosemide 2 mg twice daily, spironolactone 6.25 mg once daily, and captopril 1.5 mg twice daily are the therapies the patient is given in the PICU. The existence of the coronary abnormality was verified by further testing, such as coronary angiography, computed tomography scanning, and echocardiography.

Echocardiography presented atrial situs solitus, normal pulmonary and systemic venous drainage, AV VA concordance, dilated LA LV, intact IAS, mild to moderate TR, PG 25 mmHg, no MR, Intact IVS, No AR, no PR, LMCA from PA, No PDA seen, Left aortic arch, no coarctation of the aorta, Well contracting ventricles, no paradoxical movements, LV systolic function EF 29% FS 13% (Teicholz), EF 35 % (Simpson), LV diastolic function E/A 0.7, RV systolic function TAPSE 10 mm, No vegetation, Minimal pericardial effusion at posterior 3.5-4.9 mm, anterior

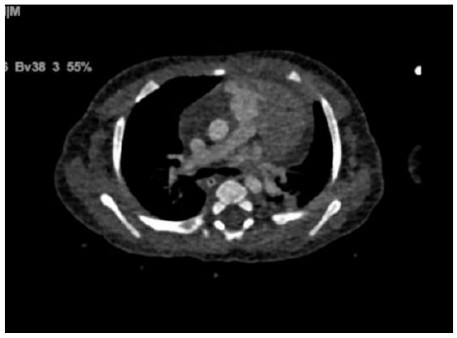

**Figure 4.** Perioperative cardiac CT

6.9 mm, apex 1.5 mm, Mild pleural effusion 4.8 mm (right), pleura 5.5 mm (left). Preoperative electrocardiography, chest x-ray, echocardiography, and cardiac CT is shown in Figure 1, Figure 2, Figure 3, and Figure 4.

From Multislice Computerized Tomography (MSCT) findings, there was an ALCAPA with a dilated RCA diameter of 2.2 mm. A possible cause of left ventricular dilatation is dilated cardiomyopathy—Stenosis of the left inferior pulmonary vein. Along with consolidation and atelectasis in segments 2 and 6 of the left lung, there is stenosis in the left main bronchus, which runs between the anterior wall of the left pulmonary vein and the posterior wall of the descending aorta.

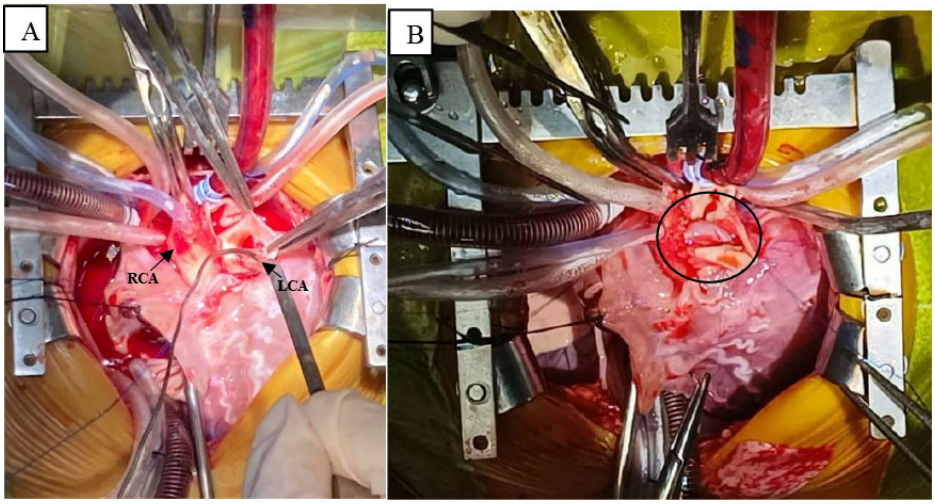


**Figure 5.** Intraoperative findings. A. Coronary arteries, B. Takeuchi baffle.

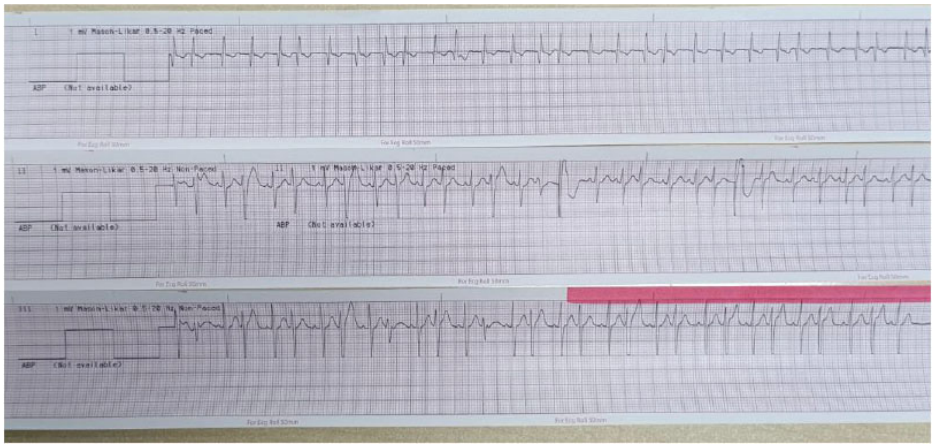

**Figure 6.** Postoperative electrocardiogram.

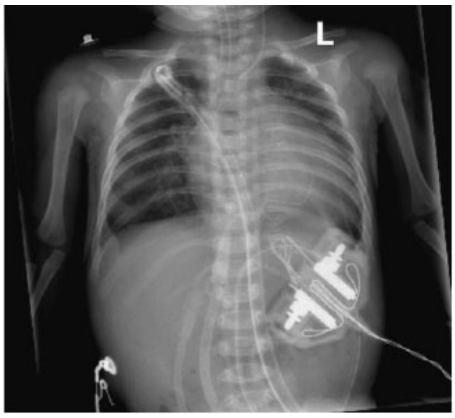


**Figure 7.** Postoperative chest x-ray.

Intraoperative findings (Figure 5) were cardiomegaly, global hypokinetic, innominate vein (+), aorta and main pulmonary artery is side by side, RCA rooting from ascending aorta, RCA aneurysm and dominant. LCA non-facing sinus on the left posterior far from the aorta, patent ductus arteriosus, and patent foramen ovale. We decided to preserve the pericardium with glutaraldehyde and do an intrapulmonary tunnel operation (Takeuchi procedure) with a pericardial baffle from the left coronary through the ascending aorta. A circular incision was made in the aorta using an aortic punch and a circular incision in the pulmonary artery. Aortopulmonary creation and intrapulmonary tunnel were carried out from the aorta to the anomalous LCA ostium. A horizontal incision creates a flap on the pulmonary artery's anterior wall. A tunnel was formed from the aortopulmonary window to the anomalous left coronary artery origin by suturing the flap to the pulmonary artery's posterior wall. The anterior wall of the PA is closed with pericardium. The patent ductus arteriosus was ligated, and 3 mm PFO was left open. The patient was transferred to the ICU in open chest management without requiring Extracorporeal Membrane Oxygenation (ECMO).

During treatment in the cardiovascular intensive care unit (CICU), the patient had several instabilities of hemodynamics, but there were no significant ST-T changes. The patient had functional LV dysfunction with LVEF 18% and RV dysfunction with TAPSE 6 mm at postoperative day one and several episodes of SVT until postoperative day two. At postoperative day four, we decided to undergo chest closure because the patient had stable hemodynamics with minimal inotropic support (adrenaline 0.048 mcg/kg/min and milrinone 0.7 mcg/kg/min), even though he still had functional LV dysfunction with LVEF 19%. The patient was extubated on the fourth day after the chest closure procedure and given an NIV supplement for three days, then stepped down to the general wards. The patient was still given minimal inotropic support and after-load reduction until seven days after the chest closure procedure to cope with LV dysfunction. LVEF gradually increased and reached 60% on the twelfth day post-chest closure. After nine days treated in the general ward for improved nutrition, clinical status, and oxygen dependency with a nasal cannula. With a nasal cannula for at-home oxygen supplementation, the patient was released from the hospital in good physical condition. Postoperative electrocardiography, chest x-ray, and echocardiography are shown in Figure 6, Figure 7, and Figure 8.

A follow-up is scheduled for three months from now. Stress echocardiography, electrocardiography, transthoracic echocardiography, and magnetic resonance imaging (MRI) are vital follow-up tests critical for assessing cardiovascular state, functional class, and physical activity.

## DISCUSSION

The goal of surgical repair methods for instances of ALCAPA is to create a dual-coronary artery system. Regardless of the patient's age or heart condition, the time for surgical intervention is advised to begin as soon as a clinical diagnosis is made. Due to severe myocardial ischemia, particularly in the early stages of infancy, there is a lack of coronary collateral improvement following a decrease in pulmonary vascular resistance and a

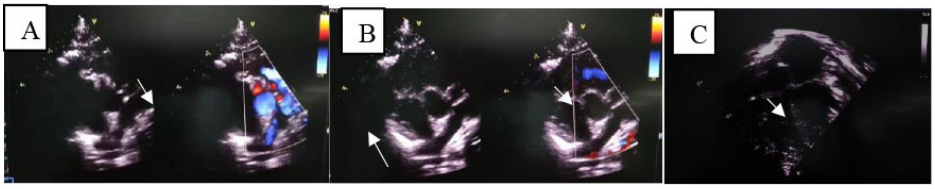


**Figure 8.** Postoperative echocardiography before discharge. A and B short axis view showing Takeuchi tunnel, C. 4-chamber view showing improved LV volume and contractility (reduced dilatation of LV).

deficit in coronary perfusion.[1,7,8]

Aortic implantation, Takeuchi tunneling, coronary artery bypass grafting, anomalous coronary ligation, and anastomosis to the subclavian artery are among the surgical techniques suggested for treating ALCAPA. We decided to do the Takeuchi procedure because the LCA was on the non-facing sinus, left posterior side, and far from the aorta. We also preserved the pericardial patch used for tunneling with glutaraldehyde to prevent tunnel obstruction that frequently occurs in this procedure.[9]

Evidence shows that individuals with ventricular failure before surgery require artificial circulatory assistance. However, there is no correlation between preoperative LV fractional shortening (LVFS) and mortality with temporary left ventricular assist device (LVAD) in 100% of the population in the study.[7–10] Severe left ventricular dysfunction postoperative is said to be the main mortality factor. Therefore, mechanical circulatory support is required to allow the LV to recover. Extracorporeal membrane oxygenation (ECMO) support causes high costs and morbidity afterward. Hence, many centers choose LVAD as these patients' initial option until biventricular dysfunction is evident.[10] The survival percentage of patients with postoperative low cardiac output syndrome can be improved with aggressive ECMO therapy.[10,11]

Research revealed that LVEF either does not improve much on the day of surgery or worsens because of surgical injury, especially in the early infancy population. According to El-Louali F. et al., these patients' worse clinical circumstances and less favorable outcomes were linked to the absence of coronary supply via collaterals. It concerns the capacity to form collaterals and the time needed for normalizing pulmonary resistance. The data also supported by Kwiatkowski DM et al. showed that 16% of patients with late presentation (>1 year old) required ECMO perioperatively. In contrast, no patients required ECMO perioperative in age <1 year old.[12]

In our case, mechanical circulatory support was unavailable since the patient was of low socio-economic status, and the government's insurance coverage did not support both devices. The patient had severe LV dysfunction and several episodes of supraventricular tachycardia (SVT) on postoperative days one and two. We suspected those were caused by reperfusion injury. Hence, in our case, the patient is still hemodynamically stable afterward and the LV dysfunction could still be tolerated by optimization of inotropes and after-load reduction medicines. Despite studies that encourage the usage of mechanical circulatory support, according to some research, individuals who need mechanical assistance following repair seem to be more likely to need a transplant or repeat surgery, usually to address mitral regurgitation. They should be aware of difficulties in the central system even though the appropriate placement of ECMO can significantly lower the mortality of severely sick infants following the procedure.[13,14] Post-therapy assessment should be assessed long-term after therapy, not just post-surgery. It is still planned and we have not included the results in this study. Thus, it is a limitation of this study.

## CONCLUSION

In summary, ALCAPA leads to myocardial ischemia and left ventricular dysfunction, posing significant risks, especially in infancy. Timely surgical intervention is essential to improve outcomes, with recent advancements in surgical techniques showing promising results in reducing mortality rates in case of severe LV dysfunction without the need for mechanical circulatory support (ECMO or LVAD). Experienced surgical teams with adequate perioperative care management are required to achieve this outcome in a low-middle-income country, even though we still encourage the availability of mechanical circulatory support for ALCAPA surgical correction.

## DISCLOSURES

### Funding
None.

### Conflict of Interest
There is no conflict of interest, according to the authors.

### Author Contribution
DSS, SW, and WM were involved in concepting, designing, and supervising the manuscript. AND and AM were involved in data analyzing and editing. PL and LM contributed to conducting and writing the manuscript.

### Ethical Consideration
The patient consented to the publication of this case for research purposes or in a journal.

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