Coronary Flow Assessment in an Adult with Anomalous Left Coronary Arising from the Pulmonary Artery

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INTRODUCTION

Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) syndrome is a congenital anomaly of the left coronary artery that affects 1 in every 300,000 people.[1] Almost 90% of patients die in infancy if not treated with surgery.[1] Collateral blood flow from the right coronary artery (RCA) to the left coronary artery (LCA) may develop well enough that myocardial ischemia or cardiac dysfunction is almost absent, and patients can reach adulthood or, rarely, old age. Thus, ALCAPA syndrome may be detected incidentally in older individuals.[2] When the collateral flow from the RCA to the LCA becomes inadequate, heart failure or life-threatening arrhythmias may develop, leading to sudden death in adult patients.

Herein, we present the case of an older patient with ALCAPA syndrome. We were able to confirm the typical landmarks of ALCAPA syndrome and analyze the characteristics of the coronary circulation by color flow Doppler and pulsed-wave Doppler echocardiography before and after surgery.

CASE PRESENTATION

A 60-year-old woman was admitted to our hospital with symptoms of heart failure for the first time. The patient's comorbidities were obesity, hypertension, dyslipidemia, a uterine myoma, and a history of smoking. On admission, the patient was diagnosed with ALCAPA but refused surgical treatment and was subsequently discharged. However, exertional breathlessness continued, and 4 months later, the patient's dyspnea rapidly worsened during a visit to the outpatient gynecology clinic. The patient was immediately intubated and admitted to the intensive care unit.

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The patient provided informed consent for publication of this case and the associated images, and the study was approved by the appropriate ethics review board.

Examination and Course after Hospitalization

The patient had sinus tachycardia with a heart rate of 115 beats/min; blood pressure was 220/110 mm Hg, and arterial oxygen saturation fell to 60% in room air. Chest examination revealed a holosystolic murmur (Levine grade III/IV) at the apex, and coarse crackles were auscultated in both lung fields. Arterial blood gas analysis under 10 L/min oxygen inhalation by mask revealed hypercapnia and respiratory acidosis (pH = 7.1, pCO_2 = 61 torr, pO_2 = 100 torr). Blood tests showed mildly elevated liver enzyme levels (aspartate aminotransferase 58 IU/L [normal range, <30 IU/L], alanine aminotransferase 37 IU/L [normal range, <23 IU/L]) and markedly elevated cardiac biomarkers (N-terminal pro–brain natriuretic peptide 3,300 pg/mL [normal range, <124 pg/mL], troponin I 30 pg/mL [normal range, <26.2 pg/mL]).

Electrocardiography revealed ST-T-segment depression in leads II, aVF, and V_3 to V_6; negative T waves in leads aVL and V_4 to V_6; and no abnormal Q waves. Chest radiography showed signs of cardiomegaly, with a cardiothoracic ratio of 61% and bilateral pulmonary congestion.

On transthoracic echocardiography (TTE; EPIQ CVx; Philips), left ventricular global systolic function was normal, and the left ventricular ejection fraction using the method of disks was 59% (Table 1). However, ventricular dilatation and hypokinesis of the basal anterior to the lateral wall showing highly echogenic mitral valvular papillary muscle and chordae were visible (Figure 1, Video 1). Moderate mitral regurgitation was detected (effective regurgitant orifice area 0.32 cm^2, regurgitant volume 59 mL; Figure 2); although left atrial volume was enlarged (left atrial volume index 65 mL/m^2), tricuspid regurgitation velocity was 2.5 m/sec, with trivial tricuspid regurgitation, and E/e′ ratio was 5.3. Thus, left ventricular diastolic dysfunction was insignificant.

For precise assessment of the mitral valve apparatus, we used transesophageal echocardiography. This revealed a mildly enlarged mitral annulus (three-dimensional mitral valvular annular area 6.4 cm^2/m^2, which was analyzed using 3D MVQ QLAB mitral valve quantification software; Philips) and mild tethering of the valve (tenting height 4.7 mm). No organic change was detected in any leaflet.

Dual myocardial scintigraphy using ^{123}I β-methyl-p-iodophenyl-pentadecanoic acid (BMIPP) and ^{201}Tl were performed at rest. A large area of decreased uptake in the anterior to lateral wall of the left ventricle was observed on ^{123}I BMIPP scintigraphy compared with ^{201}Tl, and this mismatch suggested the presence of myocardial ischemia.[3]

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, apical four-chamber view (slightly rotated), systolic phase demonstrates an echo-bright papillary muscle (*arrows*). The wall motion on the lateral site is mildly decreased.
Video 2: Two-dimensional TTE, left parasternal window without (*left*) and with (*right*) color flow Doppler, diastolic phase, demonstrates the papillary muscle in the short-axis view. The collateral branch from the posterior descending artery (PDA) is seen as it passes through the intraventricular septum.
Video 3: Two-dimensional TTE, left parasternal window, right ventricular outflow tract, long-axis view, without (*left*) and with (*right*) color flow Doppler, diastolic phase, demonstrates the orifice of the left main trunk (LMT) at the PA with retrograde flow.

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Heart failure (Forrester subset II) and pneumonia were diagnosed; thus, diuretics and antibiotics were administrated intravenously for 5 and 9 days, respectively, until the conditions resolved. Thereafter, the patient was extubated, and the drugs were changed to oral medications. This time, the patient agreed to undergo surgical correction of ALCAPA syndrome.

Diagnosis and Coronary Circulation

Coronary computed tomography and two-dimensional TTE demonstrated that the LCA originated from the pulmonary artery (PA), and the dilated RCA originated normally from the aorta (Figure 3).

Perioperative TTE revealed an enlarged RCA orifice 10 mm in diameter. Abundant collateral flow was detected from the posterior descending artery to the left anterior descending coronary artery (LAD) via septal branches (Figure 4A, Video 2). The diastolic peak velocity of the posterior descending artery was approximately 75 cm/sec (Figure 4B). The flow in the septum was retrograde from the RCA to the LAD, and the diastolic peak velocity was elevated (140 cm/sec; normal range, 21 ± 8.0 to 28 ± 9.0 cm/sec; Figure 4C).[4] The coronary flow was dominant during the diastolic phase.

The origin of the LCA was located at the medial side of the main PA, with retrograde flow from the LCA to the PA (Figure 5A, Video 3). The left main trunk showed retrograde flow toward the ostium at the PA, and the diastolic peak velocity measured using pulsed-wave Doppler echocardiography was approximately 30 cm/sec (Figure 5B). The lower level of the Nyquist limits was set at the average coronary arterial flow velocity.

Operative Details and Postoperative Findings

On the 52nd day of hospital stay, the patient underwent patch closure of the LCA origin in the PA, mitral valve repair with a 28-mm Physio Flex annuloplasty ring (Edwards Lifesciences), and bypass grafting using the left internal thoracic artery (LITA) to the LAD. The surgery was completed successfully.

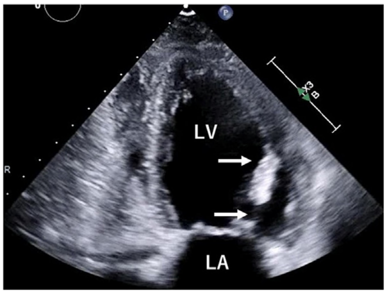

Figure 1 Two-dimensional TTE, apical four-chamber view (slightly rotated), systolic phase demonstrates an echo-bright papillary muscle (*arrows*). *LA*, Left atrium; *LV*, left ventricle.

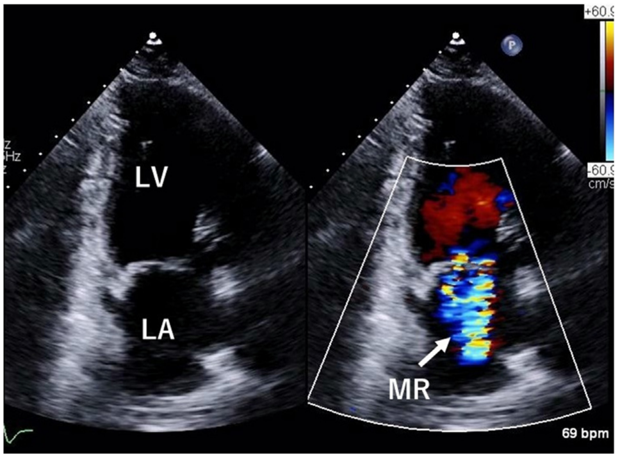

Figure 2 Two-dimensional TTE, apical two-chamber view, systolic phase without (*left*) and with (*right*) color flow Doppler demonstrates moderate mitral regurgitation (MR). *LA*, Left atrium; *LV*, left ventricle.

Postoperative TTE showed a reduction in mitral valve regurgitation and a reduction in the size of the left ventricle and left atrium (Table 1), but hypokinesis in the basal anterior to the lateral wall persisted.

LAD flow could be detected by TTE and color flow Doppler and was perfused from the LITA. The diastolic peak flow velocity of flow from the LITA to the LAD was approximately 95 cm/sec (Figure 6), while the diastolic peak flow velocity of collateral flow from the RCA to the LAD through the septum was approximately 190 cm/sec (Figure 7).

Although paroxysmal atrial fibrillation was noted, there was no recurrence of heart failure after surgery, and the patient was discharged on the 68th hospital day.

DISCUSSION

We have presented the case of an older patient with ALCAPA syndrome who developed severe heart failure and was treated successfully by surgical correction. Perioperative evaluation by echocardiography was useful to confirm the status of the coronary circulation and the results of surgical correction.

ALCAPA syndrome is a congenital heart disease in which the LCA originates from the PA.[1] When the LCA receives enough collateral flow from the RCA, left ventricular function is relatively well preserved, and patients can reach adulthood. However, myocardial

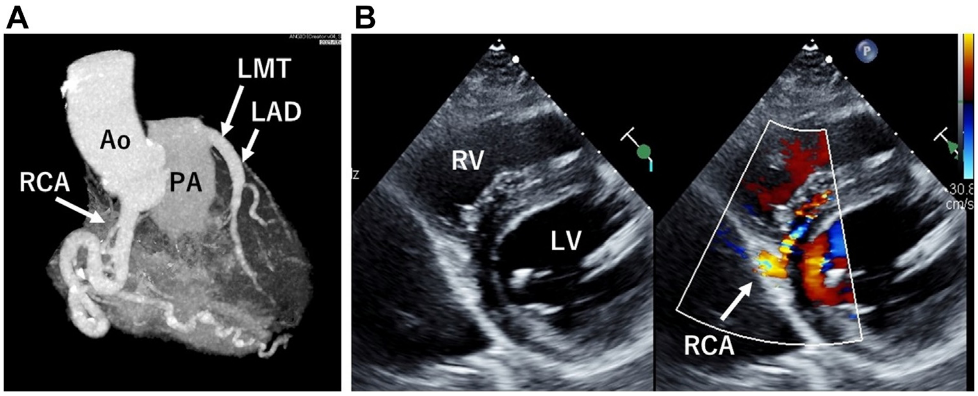

Figure 3 **(A)** Coronary computed tomography, three-dimensional whole-heart angiographic projection obtained on admission demonstrates ALCAPA. The RCA originates normally from the aorta and is enlarged and tortuous. **(B)** Two-dimensional TTE without (*left*) and with (*right*) color flow Doppler in the basal parasternal short-axis view. The RCA was dilated (*white arrows*). *Ao*, Aorta; *LMT*, left main trunk; *LV*, left ventricle; *RV*, right ventricle.

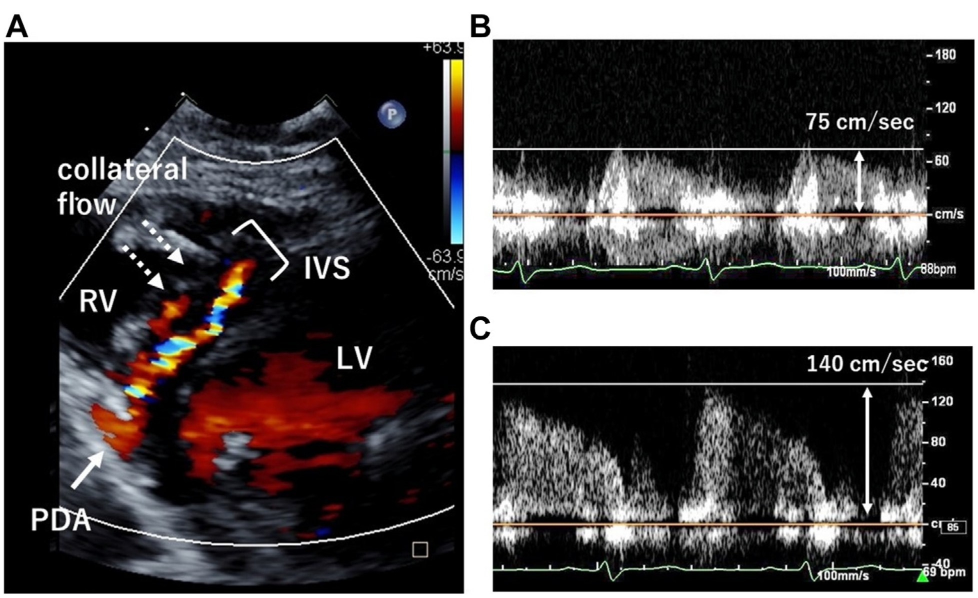

Figure 4 Color flow and pulsed-wave Doppler echocardiography performed at the level of the papillary muscle from the parasternal short-axis view in diastole. **(A)** The direction of the coronary blood flow is from the posterior descending artery (PDA; *white arrow*) to the collateral branch (*dotted arrows*). **(B)** The diastolic peak velocity of PDA flow (*white arrow*) is approximately 75 cm/sec. **(C)** The diastolic peak velocity of the collateral branch (*dotted arrows*) is approximately 140 cm/sec. *IVS*, Interventricular septum; *LV*, left ventricle; *RV*, right ventricle.

ischemia may repeatedly occur with and without symptoms and trigger the onset of heart failure, life-threatening arrhythmias, or even sudden cardiac arrest.[2] ALCAPA syndrome is rarely observed in older patients because they are commonly undiagnosed, and thus patients may be medically treated for signs of heart failure without undergoing thorough investigations.[5,6] Our patient had been living well and developed heart failure at 60 years of age for the first time and was admitted as an emergency case.

Because of the many imaging modalities available currently, diagnosing ALCAPA syndrome is not difficult. Visualizing the origin of the LCA from the PA by TTE may still be difficult, but there are typical echocardiographic findings that support ALCAPA syndrome: (1) enlarged RCA, (2) collateral vessels, (3) abnormal pulmonary arterial flow, (4) mitral regurgitation, (5) left ventricular dysfunction, and (6) highly echogenic papillary muscle and mitral valvular chordae.[1]

In adults, detection of the coronary artery may be more difficult than that in children because of attenuation of the ultrasound signals and the respiratory motion of the heart. However, in the present case, we were able to confirm all the landmarks of ALCAPA; further, we could assess the status of the coronary circulation before and after corrective surgery using color flow Doppler and pulsed-wave Doppler echocardiography.

Before surgery, the LCA received collateral blood from the RCA, and flow in the left main trunk was retrograde toward the ostium at the PA. After the closure of the ostium at the PA origin and grafting of the LITA to the LAD, the flow became normalized from the LITA to the LAD.

This is a unique case of an older patient with ALCAPA syndrome in whom remarkable abnormalities in the coronary circulation were demonstrated by the use of color flow Doppler and pulsed-wave Doppler echocardiography. Thus, analysis of coronary artery flow

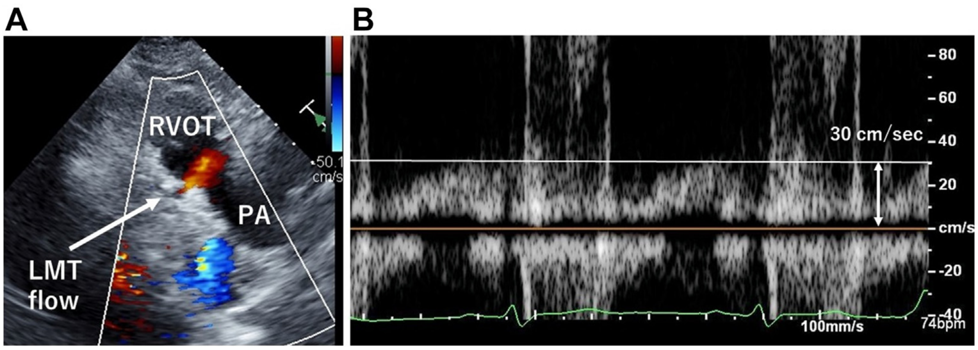

Figure 5 Color flow and pulsed-wave Doppler echocardiography performed from the right ventricular outflow tract (RVOT) view. **(A)** The orifice of the left main trunk (LMT; *white arrow*) opens at the PA. The flow direction is retrograde from the LMT to the PA. **(B)** The diastolic peak velocity of LMT flow (*white arrow*) is approximately 30 cm/sec.

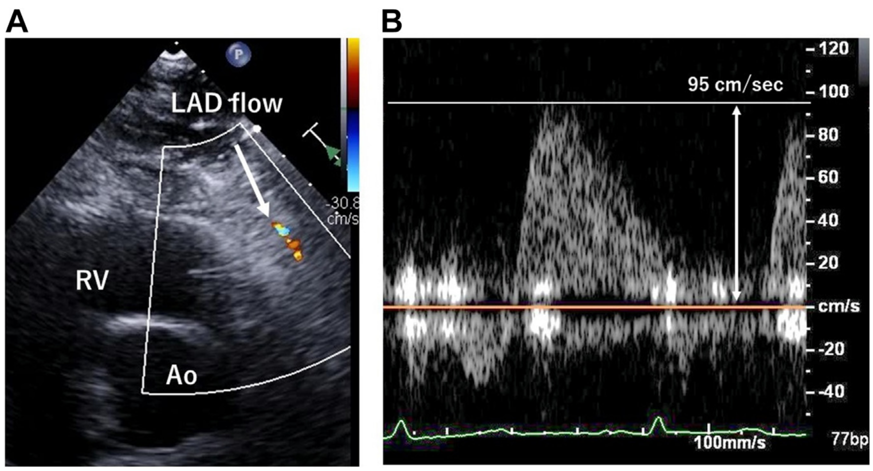

Figure 6 Postoperative color flow and pulsed-wave Doppler echocardiography performed at the level of the aorta from the right ventricular outflow view. **(A)** The proximal LAD is running in the interventricular septum, whose flow comes from a LITA graft. **(B)** The diastolic peak velocity of the proximal LAD is approximately 95 cm/sec. *Ao*, Aorta; *RV*, right ventricle.

Table 1 Comparison of echocardiographic measurements before and after bypass grafting

Variables	Before surgery	After surgery
LVEF, %	59	60
SVI, mL/m^2	42.3	38.3
LVEDd, mm	56	48 ↓
LVESd, mm	41	31 ↓
LVEDV, mL	102	70 ↓
LVESV, mL	42	28 ↓
LVMI, g/m^2	147	135
RWT	0.36	0.46
LAVI, mL/m^2	65	59
Mitral regurgitation	Moderate	Mild

LAVI, Left atrial volume index; *LVEDd*, left ventricular end-diastolic diameter; *LVEDV*, left ventricular end-diastolic volume; *LVEF*, left ventricular ejection fraction (measured using the biplane method of disks); *LVESd*, left ventricular end-systolic diameter; *LVESV*, left ventricular end-systolic volume; *LVMI*, left ventricular mass index; *RWT*, relative wall thickness; *SVI*, systolic volume index.

by echocardiography and pulsed-wave Doppler may make it possible to evaluate the coronary circulation and the presence of possible myocardial ischemia or microvascular dysfunction.[4,7]

Finally, the patient's heart showed reverse remodeling on the 62nd postoperative day in the form of a reduction in the diameter or volume of the left ventricle and the left atrium (Table 1). Collateral flow from the RCA is expected to decrease gradually during follow-up.[8]

CONCLUSION

Patients with ALCAPA syndrome may reach old age and develop acute heart failure. TTE can be used to confirm the typical findings of ALCAPA, and color flow Doppler and pulsed-wave Doppler echocardiography are essential in disclosing the flow pattern in the coronary arteries.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

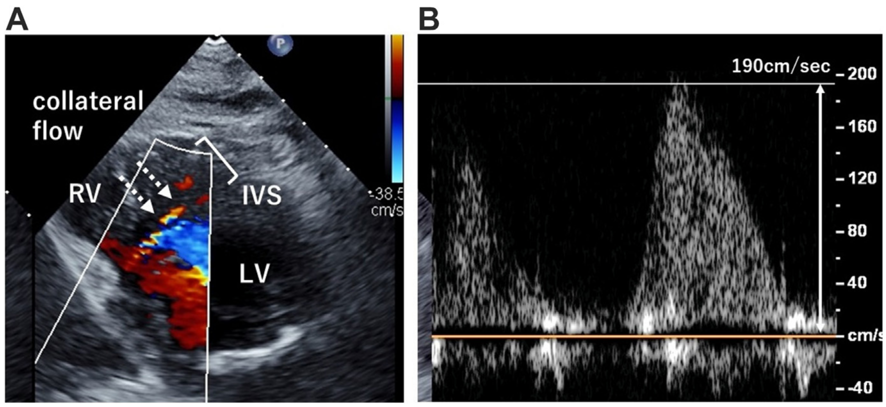

Figure 7 Postoperative color flow and pulsed-wave Doppler echocardiography performed at the level of the papillary muscle from the parasternal short-axis view. **(A)** The collateral flow (*dotted arrow*) remains after surgery. **(B)** The diastolic peak velocity of the collateral flow is approximately 190 cm/sec. *IVS*, Interventricular septum; *LV*, left ventricle; *RV*, right ventricle.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2022.10.002.

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