

Case Report

A Rare Case of an Adult Pregnant Patient with the Left Coronary Artery Originating from the Pulmonary Artery: Successful Management and Healthy Maternal-Fetal Outcome

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Abstract

An anomalous left coronary artery originating from the pulmonary artery (ALCAPA) refers to the abnormal origin of the left coronary artery either from the main pulmonary artery, pulmonary artery sinus, or the left and right pulmonary arteries, with the main pulmonary artery or pulmonary artery sinus being the most common sites. If not diagnosed and treated promptly, this condition can result in death within the first year of life in 90% of patients. Asymptomatic children can survive into adulthood, but they are at a high risk of sudden death. In this article, we report a case of a 24-year-old pregnant woman who was diagnosed with ALCAPA during prenatal examination. The pregnancy was successfully maintained until 36 weeks, after which a cesarean section was performed. The patient was then admitted to the cardiac surgery department to improve cardiac function, and six weeks later, a successful left coronary artery transplantation was performed. The patient was discharged and followed up for three months, during which her condition remained stable.

Keywords

congenital heart disease; the left coronary artery originates from the pulmonary artery; pregnancy; surgical treatment

Introduction

Anomalous left coronary artery originating from the pulmonary artery (ALCAPA) is a rare congenital heart disease, with an incidence of 1 in 300,000 live births [1]. The management principles for adult ALCAPA complicating pregnancy have been rarely reported. In this report, we present a case of a rare occurrence in an adult pregnant patient, where the left coronary artery originated from the pulmonary artery. The case was successfully managed, and both the mother and the baby remained healthy. The patient provided informed consent for the publication of this

report. The CARE checklist was used when writing this case report **Supplementary Table 1** [2].

Case Report

A 24-year-old pregnant patient with gestational age of 36 weeks was found to have elevated liver transaminases during prenatal examination. The electrocardiogram (ECG) indicated sinus tachycardia and ST-T changes. The resting heart rate was 130 beats per minute. Additionally, echocardiography revealed left atrial and ventricular enlargement, mitral valve disease with severe mitral regurgitation, mild aortic valve regurgitation, moderate tricuspid valve regurgitation, right coronary artery dilation, and moderate pulmonary artery hypertension. The patient was advised to seek immediate medical attention at a tertiary hospital and was referred to our institution with a diagnosis of peripartum cardiomyopathy complicated by heart failure. The patient experienced slight dyspnea during exercise but had no chest pain or syncope. Physical examination revealed a grade 3/6 systolic murmur at the apex. The ECG demonstrated sinus rhythm, left ventricular hypertrophy with widened QRS, and nonspecific ST-T segment changes (Fig. 1). Laboratory tests, including cardiac troponin, N-terminal pro-B-type natriuretic peptide (NT-proBNP), and autoantibodies, were within normal ranges. The patient's family history was notable for the mother's sudden cardiac death, with the specific cause unknown. After undergoing cardiac ultrasound examination at our institution, it was revealed that the left coronary artery originated from the intramural left pulmonary artery (Fig. 2), approximately 7.4 mm wide, with abundant intramyocardial collaterals. A fine vessel connected the left coronary sinus to the aorta. The right coronary artery showed dilation being approximately 7.5 mm wide. Left atrial and ventricular enlargement, moderate to severe mitral valve regurgitation (Fig. 3A) with partial calcification of the chordae tendineae and papillary muscles, mild aortic valve regurgitation, generalized reduction in left ventricular wall motion, and reduced left ventricular systolic function (left ventricular ejection fraction 35%) were observed. After multidisciplinary

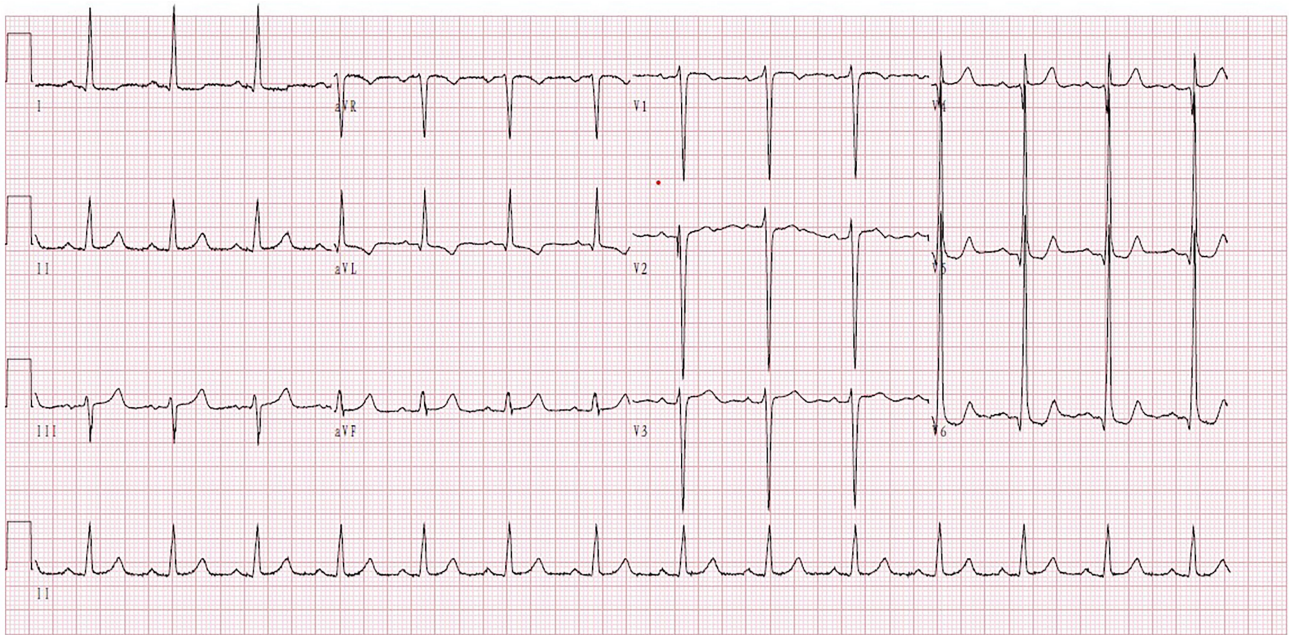


Fig. 1. ECG showing sinus rhythm, left ventricular hypertrophy with QRS widening, and nonspecific ST-T segment changes. ECG, electrocardiogram.

plinary consultations involving cardiovascular surgery, cardiology, obstetrics, anesthesia, neonatology, and the intensive care unit (ICU), it was decided to administer corticosteroids to promote fetal lung maturity. A cesarean section was performed three hours after the patient's visit. The newborn's vital signs are normal. Postoperatively, the patient was transferred to the cardiac surgery ward for a 6-week treatment course, including recombinant human brain natriuretic peptide and dopamine to improve cardiac function, furosemide for diuresis, and metoprolol to control ventricular rate. Weekly cardiac ultrasound examinations showed progressive improvement in ejection fraction, increasing from 35% to 45%. Coronary computed tomography angiography (CTA) revealed the left main coronary artery originating from the pulmonary artery trunk, while the right coronary artery had a normal origin. Both coronary arteries exhibited luminal dilatation, with the right coronary artery and its branches particularly prominent and tortuous (Fig. 4A,B). Coronary angiography demonstrated an abnormally enlarged right coronary artery and small left coronary artery, with extensive collateral circulation observed between them, supplying blood flow from the right coronary artery to the left coronary artery and then to the pulmonary artery (Supplementary Video 1). The right cardiac catheterization report demonstrated that the heart index was 6.475 L/min/m², the mean pulmonary artery pressure was 21 mmHg, the pulmonary vascular resistance was 0.007 wood, and the Qp/Qs was 1.86.

After a definitive diagnosis, a specialist in both congenital heart disease and adult heart disease performed the left coronary artery direct reimplantation surgery. The op-

eration proceeded with a median sternotomy, pericardial opening, systemic heparinization, and establishment of cardiopulmonary bypass through cannulation of the ascending aorta, superior and inferior vena cava. A drainage catheter was inserted into the root of the right upper pulmonary vein, the histidine-tryptophan-ketoglutarate solution (HTK solution) perfusion catheter was inserted into the root of the aorta, and another HTK solution perfusion catheter was inserted into the root of the pulmonary artery. Intraoperative exploration revealed significant tortuous dilation of the right coronary artery (Fig. 5A). Under cardiopulmonary bypass, the ascending aorta and pulmonary artery were occluded, and the aortic root was infused with cardioplegic solution while the pulmonary artery root was also infused with cardioplegic solution. Cardiac arrest occurred without a flutter, and ice slush was placed in the pericardial cavity. Intraoperative exploration revealed that the left coronary artery originated from the pulmonary artery sinus, and the left coronary artery was completely dissected and mobilized from its anomalous origin (Fig. 5B). Surrounding collateral vessels were sutured and ligated for hemostasis, and an end-to-side anastomosis was performed with the aortic wall (Fig. 5C). Concurrently, mitral annuloplasty was performed with LivaNova SB30M mitral valve plasty ring. Intraoperatively, abundant collateral vessels were observed on the posterior wall of the pulmonary artery, and the vessels exhibited increased fragility. The pulmonary artery was completely severed, the posterior wall of the pulmonary artery was exposed, the posterior wall of the pulmonary artery was closed with bovine pericardial patch, and the pulmonary artery was anastomosed end to end. But the result was not

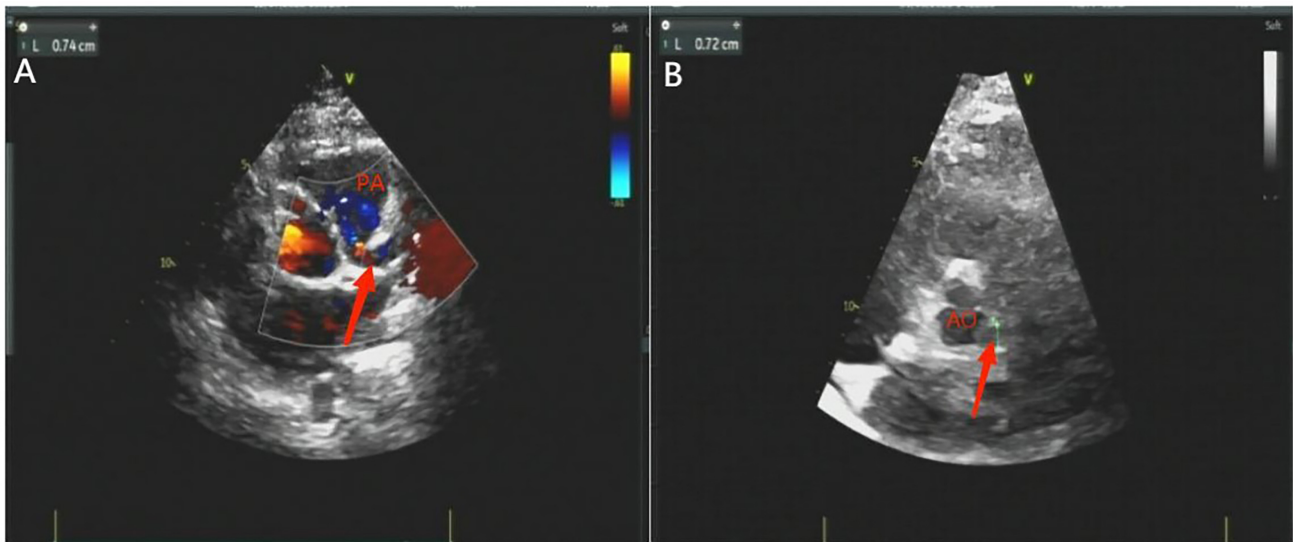


Fig. 2. Preoperative and postoperative cardiac ultrasound. (A) The left coronary artery originates from the red arrow near the medial wall of the common pulmonary trunk. (B) Left coronary artery transplantation to the aorta (red arrow). PA, Pulmonary Artery; AO, Aorta.

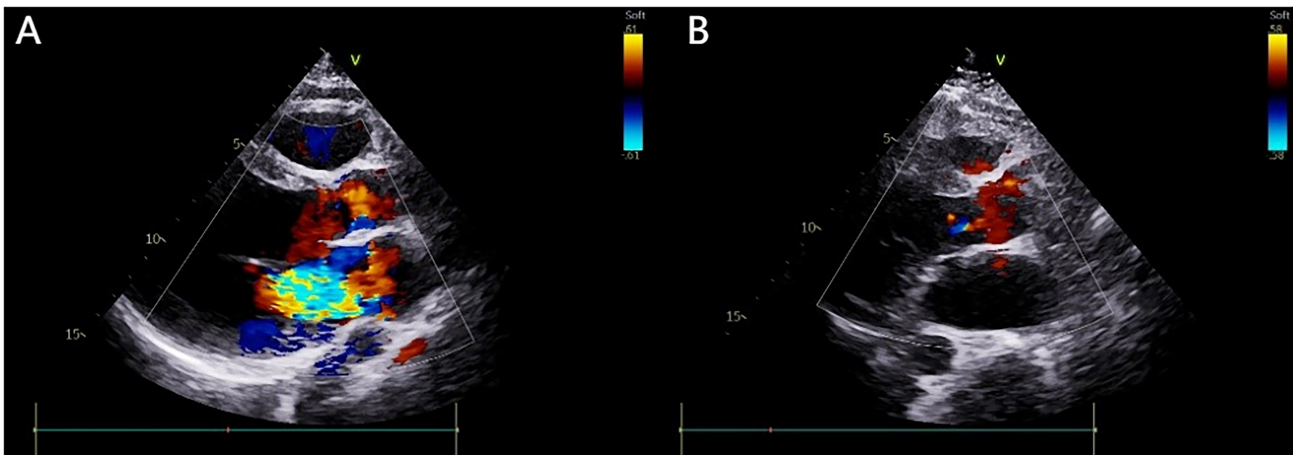


Fig. 3. Comparison of mitral regurgitation before and after operation. (A) Preoperative echocardiography showed moderate to severe mitral regurgitation. (B) Postoperative echocardiography Doppler showed that there was almost no mitral regurgitation.

satisfactory. Therefore, gauze packing was used and removed after 72 hours. In the first operation, the bypass time was 67 min and the aortic cross clamping time was 126 min. After the second operation, the patient had the tracheal tube removed the next day. 10 days after surgery, the patient recovered well and was discharged successfully. Follow-up cardiac ultrasound at 3 months showed that the left coronary artery was transplanted to the aorta (Fig. 2B), the left atrium had dimensions of 38 mm in both the left-right and anterior-posterior axes, and an ejection fraction of 57% was noted, Mitral regurgitation disappeared (Fig. 3B). Cardiac CTA imaging revealed unobstructed blood flow in the root of the left coronary artery and no significant stenosis was observed (Fig. 6).

Discussion

Among the different clinical types of anomalous origin of the coronary arteries, an anomalous left coronary artery originating from the pulmonary artery (ALCAPA) is one of the most common conditions. ALCAPA is a rare congenital coronary artery anomaly with an incidence of approximately 1 in 300,000, accounting for 0.25%–0.5% of all diagnosed cases of congenital heart disease [3]. This condition was first discovered in a postmortem study of a 3-month-old infant by Bland, Garland, and White in 1933 and was named Bland-White-Garland syndrome [4]. During embryonic development, ALCAPA may be associated with abnormal separation of the truncus arteriosus or persistence of the pulmonary artery bud, as well as inadequate de-

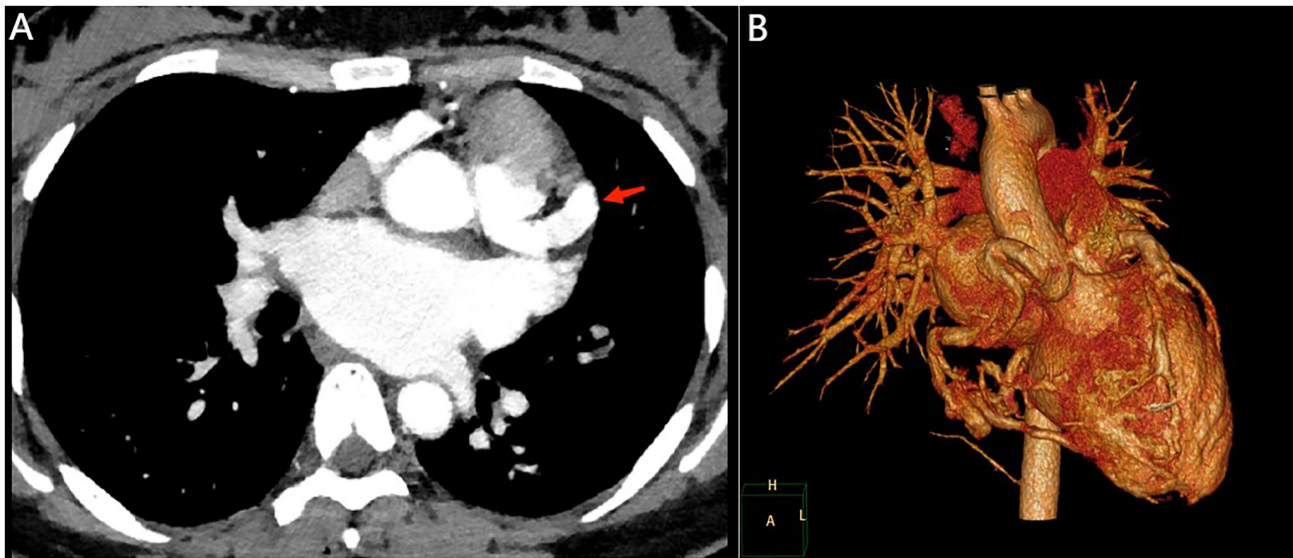


Fig. 4. Preoperative CT and 3D images. (A,B) The left coronary artery originates from the root of the pulmonary artery (red arrow). CT, computed tomography; 3D, three dimensional.

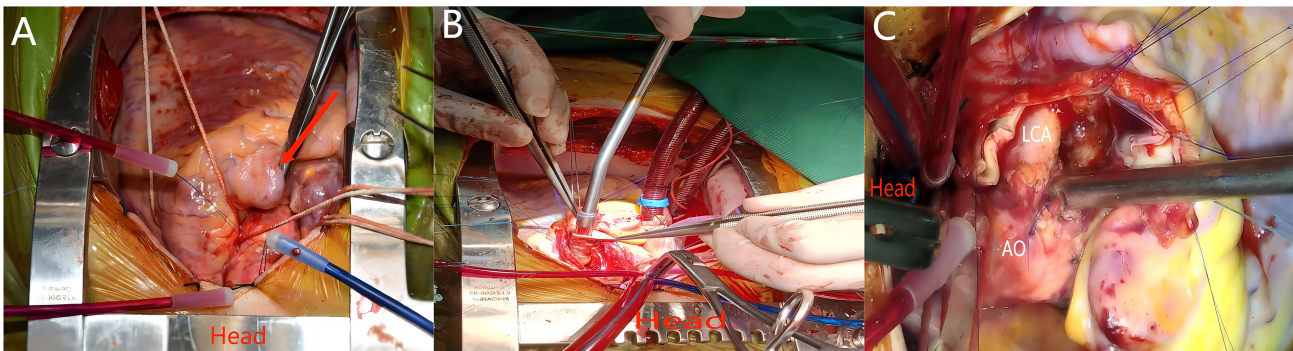


Fig. 5. Intraoperative image. (A) Clearly tortuous and thickened right coronary artery (red arrow). (B) The forceps were applied to the left coronary artery opening. (C) The left coronary artery was successfully transplanted into the aorta. AO, Aorta; LCA, left coronary artery.

velopment of the aortic bud of the coronary arteries [5]. AL-CAPA can be divided into two clinical types: infantile type and adult type, distinguished by the presence or absence of collateral circulation between the right and left coronary artery systems. The majority of infants with the infantile type experience inadequate left coronary artery blood supply leading to left ventricular dysfunction within a few months after birth, with a mortality rate of over 90% in children under 1 year of age. Approximately 10% to 15% of patients have sufficient collateral circulation and can survive asymptotically into adulthood or present with symptoms such as angina, dyspnea, or palpitations [6]. Furthermore, due to retrograde coronary artery flow into the low-pressure pulmonary artery system, coronary perfusion pressure decreases, resulting in myocardial ischemia of the left ventricle, a condition known as “coronary steal”. In adulthood, approximately 80%–90% of patients will experience sudden death due to malignant ventricular arrhythmias, with an

average age of survival of 35 years [7]. In this case, the patient’s mother died suddenly when the patient was young, and the specific cause was unknown. Similar cases have been reported by Prandi *et al.* [8], where the patient’s immediate family members had a mother who died suddenly due to heart disease (specific disease not specified), although the genetic probability of this disease still requires further multicenter large-sample studies. According to the American Heart Association’s guidelines for the treatment of adult congenital heart disease [9], once diagnosed with adult AL-CAPA, regardless of myocardial survival rate, immediate surgical intervention should be performed. This recommendation emphasizes the importance of early intervention.

This article presents a case study of a pregnant patient who did not exhibit any apparent specific symptoms during pregnancy but was diagnosed with a rare cardiac condition of an anomalous left coronary artery originating from the pulmonary artery (ALCAPA) through prenatal ultrasound

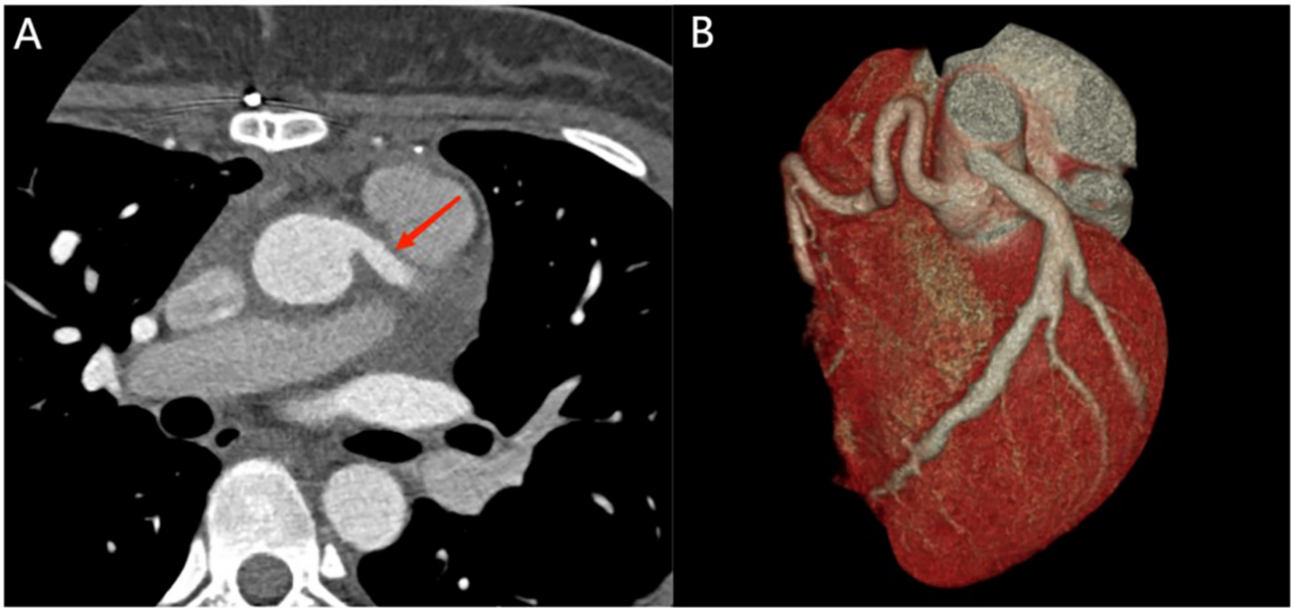


Fig. 6. Postoperative CT and 3D images. (A,B) After surgery, the left coronary artery root flow was smooth, and no obvious stenosis was observed.



Video 1. Aortic root angiography video presentation. Aortic root angiography shows a large right crown and absence of the left crown. The right corona-left corona-pulmonary artery is visible (<https://doi.org/10.59958/hsf.6407>).

examination. Previously, a misdiagnose case of peripartum cardiomyopathy was reported by Frigault *et al.* [10], where the patient presented with symptoms such as dys-

pnea, cough, severe edema, reduced left ventricular ejection fraction, and diffuse hypokinesis of the left ventricular inferior wall. This misdiagnosis highlights the importance of accurate diagnosis for such patients, particularly in ruling out the possibility of ALCAPA. Currently, transthoracic echocardiography is a commonly used method for diagnosing ALCAPA. Echocardiography can reveal significant cardiac enlargement, impaired left ventricular systolic function, and the inability to visualize the origin of the left coronary artery from the aortic sinus. Studies have shown that color flow Doppler imaging detecting abundant flow signals within the interventricular septum and left ventricular wall is also an important marker of anomalous coronary artery origin from the pulmonary artery [11]. The most important diagnostic sign is the identification of anomalous origin of the left coronary artery from the pulmonary artery along the long axis of the pulmonary artery, accompanied by the observation of retrograde blood flow. If the diagnosis cannot be confirmed by cardiac ultrasound, coronary computed tomography angiography can aid in determining the diagnosis by accurately displaying the origin and distribution of coronary arteries and evaluating their anatomical relationships with surrounding structures, providing crucial information for surgical planning. Additionally, cardiac magnetic resonance imaging is a valuable technique that can accurately assess the extent of myocardial scar tissue and coronary artery blood flow patterns. It is also of significance in assessing the risk of ventricular arrhythmias and can be used to monitor the recovery of cardiac function after ALCAPA repair [12]. In summary, timely exclusion of ALCAPA is crucial for patients with suspected peripartum cardiomyopathy. Transthoracic echocardiography is a com-

monly used diagnostic and follow-up tool, while coronary computed tomography angiography and cardiac magnetic resonance imaging can serve as adjunctive measures to help physicians confirm the diagnosis and evaluate the patient's condition. For pregnant patients, accurate diagnosis and assessment of the disease are essential for ensuring maternal and infant health.

Although the incidence of cardiac disease during pregnancy is relatively low, it remains a major cause of maternal mortality [13]. Physiological changes during pregnancy, such as increased total blood volume, cardiac output, heart rate, and a prothrombotic state, can lead to increased myocardial oxygen consumption, making patients susceptible to serious complications during the peripartum period, such as heart failure. According to existing guidelines for adult congenital heart disease (ACHD) and pregnancy, patients with complex congenital heart disease should be managed and delivered in a tertiary center with a multidisciplinary team experienced in ACHD. This medical team should include cardiac specialists, high-risk obstetricians, anesthesiologists, and neonatal experts. Early collaboration and communication among members of the multidisciplinary team are crucial for improving maternal outcomes [14]. Based on this case of the ALCAPA patient, our team recommended opting for a cesarean section delivery to reduce cardiac burden. This is because vaginal delivery, with prolonged uterine contractions and increased myocardial oxygen consumption due to hemodynamic changes and pain, can increase the risk of myocardial ischemia [15]. The patient successfully delivered under the multidisciplinary care of cardiac surgery, cardiology, obstetrics, anesthesiology, neonatology, and intensive care unit personnel. In general, postpartum healthy women require 4–8 weeks for hemodynamic recovery to reach the pre-pregnancy normal state, while patients with congenital heart disease may require a longer period of 6–12 weeks [16]. Therefore, we planned to perform the surgery after 42 days postpartum to ensure the safety of the procedure and closely monitor the vital signs of the mother.

The surgical treatment principle of ALCAPA aims to restore myocardial perfusion in the dual coronary artery system and ensure long-term patency of the corrected coronary artery [17]. The surgical corrective methods for ALCAPA include the following options: (1) Ligation of the anomalous coronary artery: This procedure has a relatively high short-term and long-term mortality rate and is currently seldom employed. (2) In addition to ligation, combined with coronary artery bypass grafting of the left coronary artery system: Primarily applied in adult patients with small coronary artery openings or intramural course. (3) Intrapulmonary tunnel (Takeuchi procedure): This surgery carries a higher risk of postoperative complications, potentially leading to pulmonary artery tunnel fistula or stenosis. (4) Coronary artery reimplantation: This procedure fully restores the left coronary system both physiologi-

cally and anatomically, resulting in favorable short-term and long-term outcomes, making it the preferred choice. Compared to infantile patients, the surgical procedure of directly implanting the left coronary artery into the aorta may be more challenging in adult patients. This is because adult patients with ALCAPA often have abundant collateral vessels around the abnormally originated left coronary artery, which are fragile and prone to bleeding. This characteristic contrasts sharply with the infantile form, where the coronary artery is easily dissected, making direct anastomosis of the left coronary artery to the aorta feasible [18]. In the present case, abundant collateral vessels were found in the posterior wall of the pulmonary artery during surgery. Although the surgeon completely severed the pulmonary artery, exposed the posterior wall of the pulmonary artery, closed the posterior wall of the pulmonary artery with bovine pericardial patch, and anastomosed the pulmonary artery end to end, the hemostatic effect was still unsatisfactory. Therefore, gauze packing was used and closure was delayed until the patient's coagulation function recovered. There has been ongoing debate regarding whether concomitant mitral valve repair should be performed during surgery for patients with concurrent mitral valve insufficiency. Based on our clinical experience, we support concomitant mitral valve repair for patients with moderate to severe mitral regurgitation, regardless of structural changes. Our team [19] conducted a single-center clinical study spanning 20 years, including 96 patients with coronary arteries originating from the pulmonary artery. Among this cohort, 40 patients underwent mitral valve repair. There were no significant differences in the cardiopulmonary bypass time (154 ± 97 vs 138 ± 81 minutes, $p = 0.384$) and aortic cross-clamp time (91 ± 28 vs 85 ± 43 , $p = 0.419$) between the mitral valve repair group and the non-repair group, indicating that concomitant mitral valve repair did not significantly increase the operation time. Simultaneous mitral valve repair effectively reduces postoperative mitral regurgitation, promotes early recovery of cardiac function, and lowers the probability of future reoperation due to mitral valve insufficiency.

Conclusions

In summary, the incidence of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is relatively low. Currently, direct implantation of the left coronary artery to the aortic root has become the main surgical approach for treating ALCAPA, and is associated with fewer complications. For patients with concomitant moderate to severe mitral regurgitation, we advocate simultaneous mitral valve repair. Lastly, for pregnant patients with ALCAPA, early preoperative diagnosis and a comprehensive multidisciplinary management plan are crucial. This plan should include assessing the risks to both the mother

and the fetus, adjusting cardiac medications if necessary, continuous follow-up during pregnancy, devising an appropriate delivery plan, selecting an optimal timing for postpartum surgery, and preventing and treating potential intraoperative complications. Furthermore, ALCAPA surgery is technically challenging and requires high-level postoperative care, necessitating the expertise and coordination of cardiac surgeons, anesthesiologists, cardiopulmonary bypass teams, and postoperative care teams.

Availability of Data and Materials

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions

HW and ZL designed this study and made a major contribution to the manuscript. HW and ZL contributed equally to this work. GZ and HF contributed to the data analysis and discussion. DL was involved in acquisition, analysis, and interpretation of data for the work. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work to take public responsibility for appropriate portions of the content and agreed to be accountable for all aspects of the work in ensuring that questions related to its accuracy or integrity.

Ethics Approval and Consent to Participate

Institutional review board approval was obtained before publication of this manuscript. This study was conducted according to the “Helsinki Declaration” and was approved by Suzhou Municipal Hospital (IRB: KL901393). Before deciding to participate in this trial, the patient was given written informed consent.

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Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at <https://doi.org/10.59958/hsf.6407>.

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