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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) diagnosed in children and adolescents

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Abstract

Background: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare but potentially fatal congenital coronary anomaly associated with early infant mortality and sudden adult death. By the development or lack of coronary collateral, it can be classified as infantile or adult type. However, even with the compensatory mechanism in adult patients, there is an estimated 80 to 90% incidence of sudden death at the mean age of 35 years.

Methods: We enrolled 9 patients with ALCAPA within the age group 5 to 16 years.

Results: Only one patient developed symptoms (apnoea), whereas other patients were asymptomatic, and there was no evident left ventricular dysfunction found in any of the cases.

Conclusion: With the development of imaging techniques, asymptomatic adult-type ALCAPA patients could be identified and diagnosed in childhood or adolescence. As a potential cause of sudden death, ALCAPA should be surgically repaired soon after the diagnosis.

Keywords: Congenital heart disease, Anomalous origin of the left coronary artery from the pulmonary artery

Background

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare but potentially fatal congenital coronary anomaly associated with early infant mortality and sudden adult death. Krause and Brooks reported the first incidences of ALCAPA syndrome in 1865 and 1885, respectively [1, 2]. In 1933, Garland, Bland, and White made the first clinical pathologic correlation, and they also proposed a review of congenital variations in the coronary vessels and a discussion of their embryological background [3, 4]. Garland and associates estimated that ALCAPA is present in 1 out of 300,000 live births or 0.5% of the children

with congenital heart disease, and these data were widely cited during past years. With the development of diagnostic techniques, more asymptomatic patients have been diagnosed and treated surgically.

Conventionally, ALCAPA syndrome is classified into two types. There is little or no coronary collateral development in the infantile type of circulation; and after the closure of patent ductus arteriosus, it could lead to severe myocardial ischemia, left ventricle (LV) dysfunction, dilatation, and mitral regurgitation (MR) when the pressure of pulmonary artery (PA) falls [5–7]. The infantile type patients die within weeks to months after birth without surgical correction. About 10 to 15% of the ALCAPA patients are adult-type [7], and their survival is due to a large dominant right coronary artery (RCA) with extensive inter-coronary collaterals, as well as a restrictive opening between the ALCAPA and the PA [3, 8–10]. With

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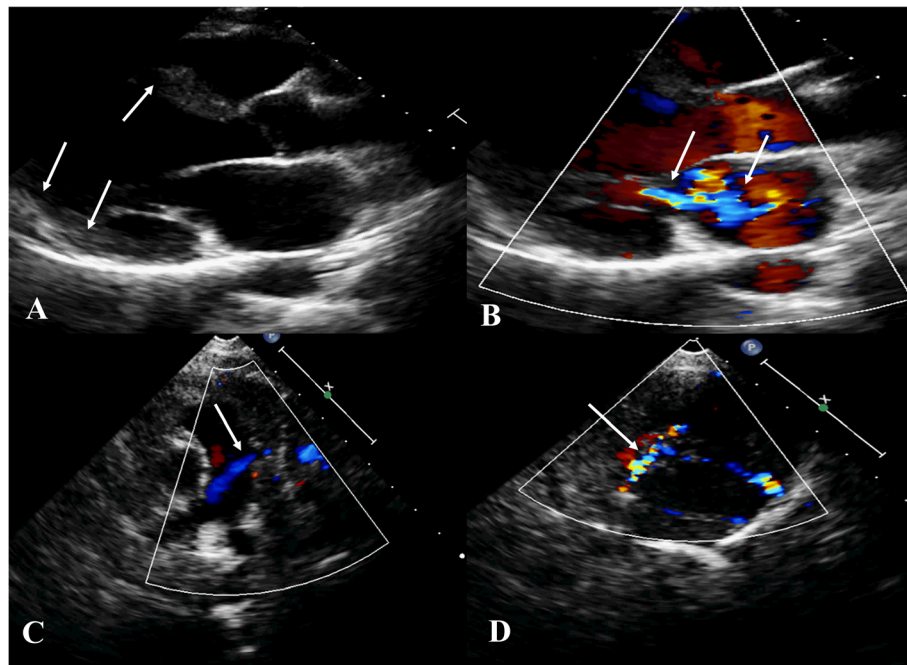


Fig. 1 **a** 2D on parasternal long axis view. White arrows show the dilated left ventricle (LV). **b** Color 2D on parasternal long axis view. White arrows show Mitral regurgitation (MR). **c** Color 2D on parasternal short axis view. White arrow shows the retrograde flow from the left coronary artery (LCA) into the pulmonary artery. **d** Color 2D on parasternal short axis view. White arrow shows the increased flow in the intraventricular collateral vessels from the RCA to the LCA

them needed surgery. CT, CMR, or coronary angiography confirmed the diagnosis in all the patients. These methods identified a single large RCA derived from the aorta with extensive LCA-derived collaterals draining into the PA (Fig. 2).

Surgical correction was attempted in 8 patients (88.9%). Re-implantation ($n = 6$) (Fig. 3) and Takeuchi ($n = 2$)

procedure were performed in these patients. One of the patients (11.1%) underwent VSD closure; two of the patients (22.2%) underwent surgical mitral valve repair at the same time, and one of them (11.1%) was implanted with a temporary pacemaker. During the surgery, the mean times of cardiopulmonary bypass (CPB) and aortic occlusion were 141 ± 31.5 and 105 ± 26.7 min, respectively. Blood

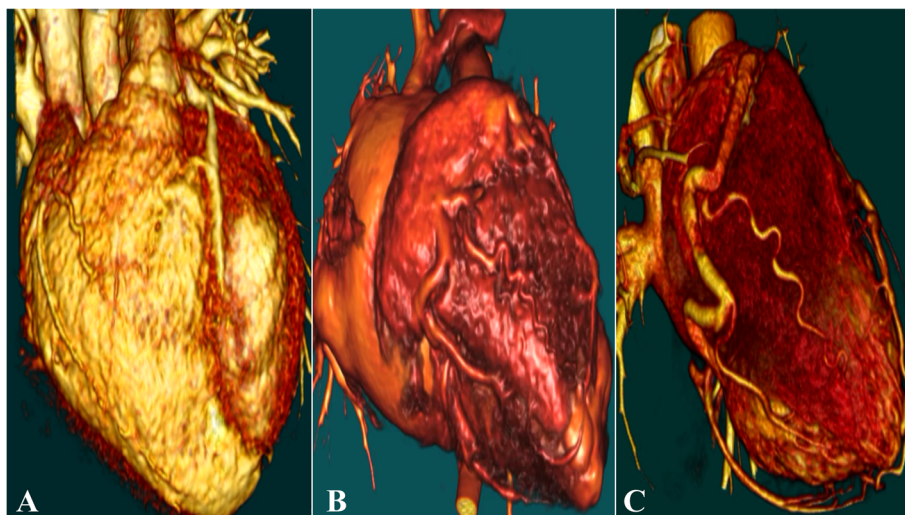
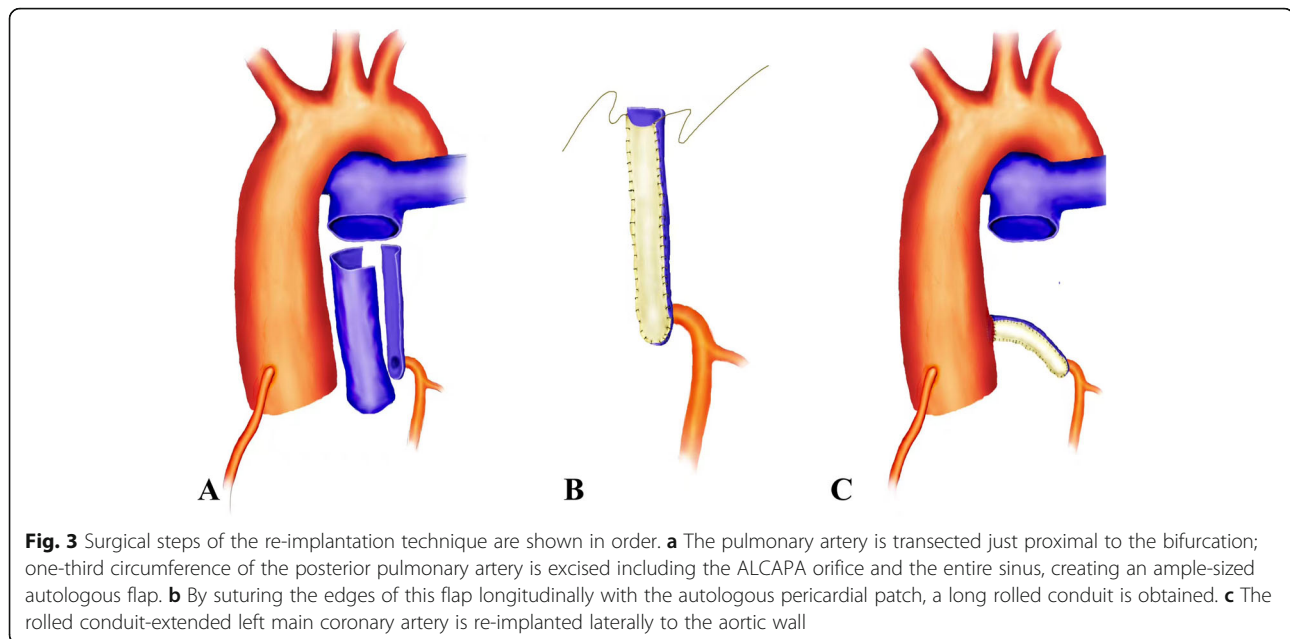


Fig. 2 CT coronary angiogram - 3D reconstruction shows different levels of collateral vessels from the right coronary artery (RCA) to the left coronary artery (LCA) in 5-year-old boy (a), 7-year-old girl (b), 16-year-old boy(c)



transfusion was necessary for the patients who underwent surgical repair. One patient (11.1%) refused to undergo an operation.

At a mean follow up of 2 ± 0.8 years, all the patients were alive. One patient underwent another operation to deal with the dissociative steel wire after surgical repair, and the other 7 patients (77.8%) reported no complications. The patient who declined surgery was not followed up after discharge.

Discussion

We present data on 9 patients diagnosed in childhood or adolescence with ALCAPA syndrome whose ages were between 5 and 16 years. Only one patient developed symptoms (apsychia), whereas other patients were asymptomatic, and there was no evident left ventricular dysfunction found in all the cases. ALCAPA syndrome mostly presents at the very beginning of life. Asymptomatic ALCAPA patients diagnosed in childhood or adolescence are even rarer and there are only a few cases reported [13].

ALCAPA also can be accompanied by other congenital heart diseases, but it is mostly an isolated malformation [5, 6, 14]. In these cases, one patient had a VSD and a dextropositioned aorta. Different imaging modalities were applied for the diagnosis of ALCAPA and they revealed the presence of collateral arteries. The main protective mechanism of ALCAPA patients is the development of collaterals which supply oxygenated blood from RCA to LCA [9, 10, 15]. When coronary steal syndrome arose or the collateral vessels were stenosed, those who survived the infantile period may present with symptoms [5, 6, 15]. ST changes and Q waves in the anterior and lateral leads

are common ECG changes in ALCAPA patients [4–6]. In our series, ECG changes were observed in 7 (77.8%) patients. Echocardiography made the early diagnosis of ALCAPA in most of our patients and would play a more important role in the early diagnosis of asymptomatic adult-type ALCAPA in childhood or adolescence. The poor image quality in patients with difficult echo windows is the main limitation of echocardiography. It is also difficult to visualize the anomalous origin of LCA in the echocardiography. CT and CMR can directly provide visualization of the coronary artery anatomy with 3D reconstruction. Moreover, CMR could assess the size and function of biventricular and estimate abnormalities and shunts of the valve [16]. Coronary angiography could provide the detailed visualization of collaterals and quantify the left to right shunt.

In ALCAPA patients, MR is a common symptom that is observed early than even ventricular dysfunction [15, 17]. Our cases suggest that the accompaniment of dilated cardiomyopathy and MR is observed in asymptomatic children or adolescents with ALCAPA too. The timing of mitral valve repair at the time of ALCAPA remains controversial because mitral regurgitation degree may gradually decrease or remain stable in patients with preoperative mild and moderate mitral regurgitation without concomitant mitral valve repair, mitral valve repair or replacement is generally not necessary in this situation particularly in infants [18]. However, some authors advocate routine mitral valve repair at the of coronary artery revascularization because early postoperative cardiac output is improved and operative mortality is reduced [19]. We did not recommend concomitant mitral valve repair because mitral regurgitation in

ALCAPA patients is related both to ischemic left ventricular dilatation with mitral annulus enlargement and to ischemic dysfunction of the papillary muscles and the mitral regurgitation may recover after the surgery [18]. Moreover performing complex mitral valve repair is time-consuming, thus increasing the ischemic time, which may be deleterious in patients with compromised myocardial function [20]. Due to the lifelong risk of ischaemia, ventricular arrhythmias, and sudden death due to cardiac arrest, surgery is the only definitive treatment of ALCAPA suggested even in asymptomatic adult patients [5, 6, 15]. However, the treatment for asymptomatic children or adolescents with ALCAPA was not mentioned. Considering the abnormal coronary system, dilated cardiomyopathy, and MR, early surgery is also necessary to recover normal circulation and prevent long-term myocardial ischaemia and fibrosis.

Conclusion

With the development of imaging techniques, more asymptomatic adult-type ALCAPA patients could be diagnosed early in childhood or adolescence. Suspicion of ALCAPA should be raised when the echocardiograph shows a large RCA or a retrograde flow in the LCA. Early surgery is also necessary for asymptomatic children or adolescents with ALCAPA.

Abbreviations

ALCAPA: Anomalous origin of the left coronary artery from the pulmonary artery; PA: Pulmonary artery; LCA: Left coronary artery; LAD: Left anterior descending artery; RCA: Right coronary artery; PDA: Patent ductus arteriosus; LV: Left ventricle; VSD: Ventricular septal defect; CXR: Chest X-ray; CT: Cardiac Computed Tomography; MRI: Magnetic Resonance Imaging; CMR: Cardiovascular Magnetic Resonance; AHA: American Heart Association

Acknowledgements

Not applicable.

Consent for publish

Written informed consent to publish personal or clinical details were obtained from all the study participants.

Authors' contributions

Qian Yongjun and Ling Yunfei conceived and designed the study. Zhang jinmei and Wang yue wrote the paper. Qian Yongjun reviewed and edited the manuscript. All authors read and approved the manuscript.

Funding

The study was supported by the Major Science and Technology Project of Sichuan Province, China (No. 2017SZ0056) and the Scientific Research Project of Sichuan Health and Family Planning Commission, China (No. 17PJ177).

Availability of data and materials

All the data and material were restored in the database of West China Hospital.

Ethics approval and consent to participate

The study protocol was approved by the local ethics committee (Sichuan University West China Hospital) and the study was performed according to the Declaration of Helsinki.

Consent for publication

All co-authors permit editorial amendments and the paper's publication.

Competing interests

The authors declare that they have no conflict of interest.

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Received: 26 December 2019 Accepted: 27 April 2020

Published online: 12 May 2020

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