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ORIGINAL ARTICLE

Diagnosis of anomalous origin of the right coronary artery from the pulmonary artery by echocardiography

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Abstract

Purpose To review the imaging characteristics and evaluate the diagnostic value of echocardiography for diagnosing anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA).

Methods We retrospectively reviewed the echocardiographic records and compared these images with operative findings in six children with ARCAPA.

Results ARCAPA was characterized by dilation of the left coronary artery, inappropriate origin of the right coronary artery from the pulmonary artery, and collaterals within the interventricular septum. The associated malformations included atrial septal defect in four cases. Four of six cases were diagnosed correctly, while the remaining two cases were misdiagnosed: one of fistula between the pulmonary artery and the left coronary artery and another of ARCAPA diagnosed intraoperatively in one case of atrial septal defect.

Conclusion ARCAPA can be diagnosed by echocardiography but is prone to misdiagnosis. This malformation should be distinguished from coronary artery fistula.

Keywords Coronary vessel anomalies · Pulmonary artery · Echocardiography · Doppler

Introduction

Anomalous origin of the right coronary artery (RCA) from the pulmonary artery (PA) (ARCAPA) is a very rare malformation affecting 0.002% of the population. The first case series was described by Brooks in 1885; since then, only about 200 cases have been reported, most of which remained asymptomatic because they usually had adequate intracoronary artery collaterals and good ventricular function [1, 2]. ARCAPA cases are usually diagnosed by autopsy or during surgery or at angiography of other cardiac malformations, while only a few pediatric cases have recognized during transthoracic echocardiography (TTE) [3–6]. As adverse outcomes including an increased risk of myocardial infarction and sudden cardiac death have been described in ARCAPA patients regardless of symptoms, surgical correction is recommended whenever this anomaly is diagnosed [7, 8].

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Between January 1999 and December 2017, six patients with this malformation underwent surgery at our hospital. The echocardiographic findings were retrospectively analyzed to explore the diagnostic value of TTE and transesophageal echocardiography (TEE).

Patients and methods

We retrospectively reviewed the echocardiographic records and compared these images with the findings of cardiac CT, cardiac angiographic examination, and operation in six children with ARCAPA.

Patients

Six children who were diagnosed with ARCAPA by cardiac computed tomography (CT), cardiac angiography, and surgical findings between January 1999 and December 2017 were enrolled in this retrospective study. All parents provided written informed consent. The study was approved by the hospital's institutional review board.

Echocardiography

Echocardiography was performed on all patients using HP Sonos 5500 and iE33 ultrasound systems (Hewlett-Packard, Andover, MA, USA) with an S4, S8, S5-1, or S8-3 transducer. Two-dimensional, M-mode, and Doppler echocardiography was performed using standard imaging techniques in accordance with American Society of Echocardiography recommendations [9, 10]. The aortic valve annulus was observed at the parasternal long-axis view, the coronary artery was seen on the parasternal short-axis view, and the ratio of the coronary artery to the aortic valve annulus was calculated. Doppler recordings of the tricuspid inflow were taken of the sample volume between the tips of the tricuspid leaflets. Doppler recordings of the right ventricular (RV) outflow were taken with the sample volume above the pulmonic valve; myocardial performance index (MPI) was calculated by dividing the isovolumic portion by the ejection time. The studies were recorded on magneto-optical disk and Super VHS tapes for later review. The anatomy diagnosis was confirmed by TEE before operation, and the residual defect and heart function were assessed intraoperatively. Routine TTE was performed 6 months postoperatively, and global RV heart function was assessed using MPI.

Electrocardiography

We reviewed the electrocardiographic findings in six cases with ARCAPA using a MEMRS-ECG network system.

Cardiac CT and cardiac angiographic examination

We reviewed the findings of cardiac CT (GE Prospeed AX Advantx) in four cases and cardiac angiographic examination (GE Advantx LC-LP) in two cases with ARCAPA.

Results

Demographic and clinical findings

Six cases of ARCAPA were identified among 186,937 cases of congenital heart disease in our heart center, representing a 0.0032% incidence among patients with congenital heart disease. The six cases were comprised of two boys and four girls ranging in age from 4 months to 18 years. Cardiac CT was performed in four cases preoperatively and showed the origin and flow of the coronary artery (Fig. 1). Cardiac angiographic examination was performed in two cases and showed the origin and collaterals (Fig. 2).

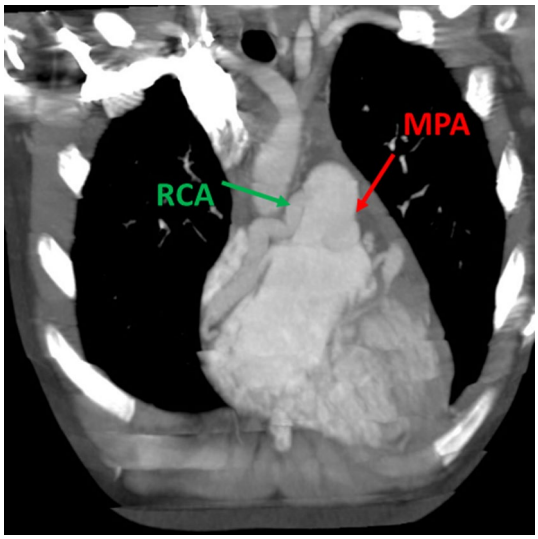

Fig. 1 In case 6, multislice spiral computed tomography showing the anomalous right coronary artery (green arrow) arising from the MPA root (red arrow). *MPA* main pulmonary artery, *RCA* right coronary artery

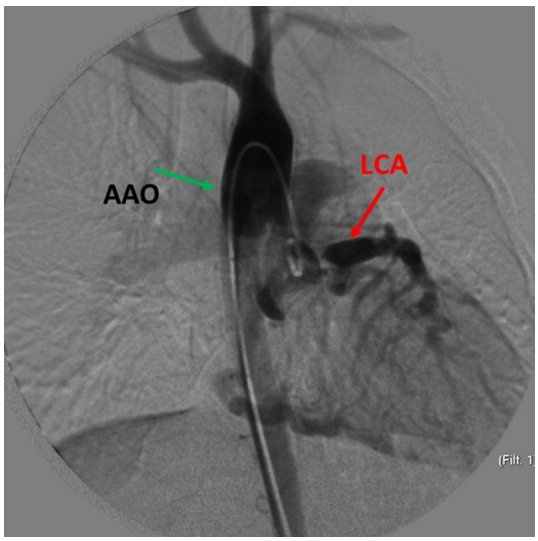

Fig. 2 In case 4, aortic angiography showing the LCA (red arrow) arising from the aorta (green arrow), and the right coronary artery receiving contrast media through several collaterals originating from the LCA and then draining into the main pulmonary artery. *AAO* ascending aorta, *LCA* left coronary artery

All six cases were subjected to surgical correction; four underwent reimplantation of the RCA to the aorta, and two underwent simple ligation of the RCA. Based on the operative findings, four of the six cases were diagnosed correctly, while the remaining two cases were misdiagnosed (one as a fistula between the PA and the left coronary artery (LCA) and the other as an atrial septal defect). Two cases were isolated ARCAPA, while the other four

were ARCAPA complicated by atrial septal defects. There were four cases of sinus tachycardia; three of ST-segment and T-wave changes, deep Q waves at V1, and III leader; two of an incomplete right bundle branch block; and one case had normal ECG.

Echocardiographic Findings

Echocardiography findings included the following: (1) the parasternal short-axis view showed dilation of the LCA with normal origin (Fig. 3), the right coronary orifice was not seen in the right coronary sinus, and the ratio of the diameter of the coronary artery originating from the normal sinus to the diameter of the aortic annulus (range 0.21–0.47) in five cases was increased (the normal value of this ratio was 0.10–0.28.[11]); (2) the parasternal PA long-axis view showed an anomalous origin of the RCA from the main PA (Fig. 4), while the color Doppler flow was antegrade (from the main PA toward the distal coronary bed) in four cases (Fig. 5); (3) the collaterals between the LCA and RCA on color Doppler showed prominent intracoronary collaterals within the interventricular septum in six cases of ARCAPA (Fig. 6); (4) the LV and RV were normal in size and right ventricular global function was normal in six cases (RV MPI: range 0.27–0.34, the normal value of RV MPI was 0.32 ± 0.03) [12].

On the preoperative TEE, the RCA originated from the MPA and the LCA dilation and course were shown (Fig. 7). On the intraoperative TEE, communication between the AAO and the RCA was found in four cases of reimplantation of the RCA into the aorta (Fig. 8), a residual leak was found in one case of simple ligation of the RCA, and no residual shunt was found in four cases with atrial septal defects.

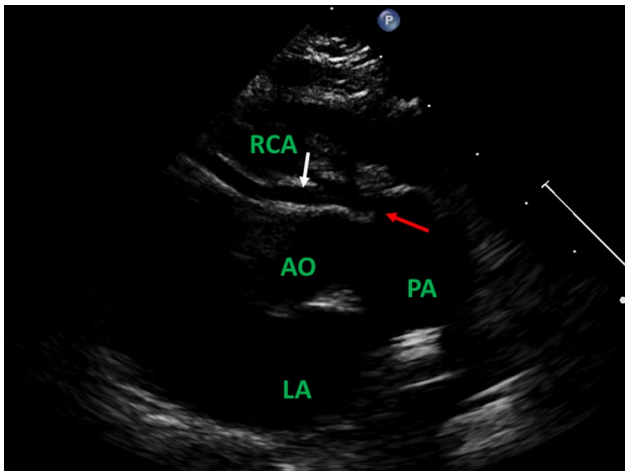

Fig. 4 In case 4, parasternal short-axis view showing the RCA arising from the PA (red arrow) and coursing rightward and anteriorly to the AO (white arrow). *AO* aorta; *LA* left atrium, *PA* pulmonary artery, *RCA* right coronary artery

The median follow-up duration was 16 months (range 5–34 months). There were no cases of early mortality, no children required further surgery, and there were no obstructions between the RCA and the AAO and no cases of situs thrombosis. A residual leak was found in one case of simple ligation of the RCA (RV MPI = 0.31). Deep Q waves disappeared at the V1 and III leads on the ECG in all three patients.

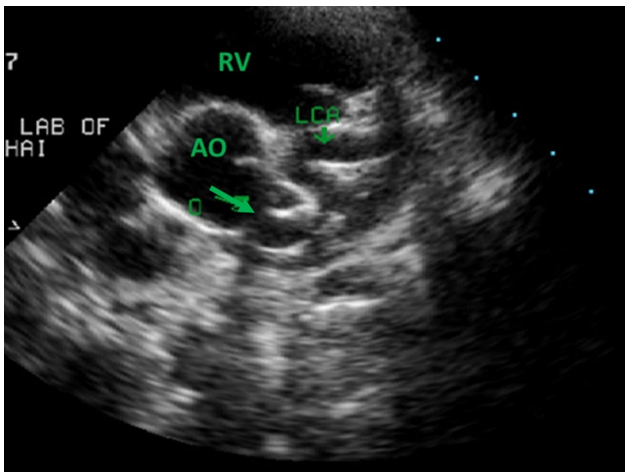

Fig. 3 In case 6, parasternal short-axis view demonstrating the dilated and circuitous LCA normally arising from the aorta. The origin of the right coronary artery could not be identified. *AO* aorta, *LCA* left coronary artery, *O* orifice of left coronary artery, *RV* right ventricle

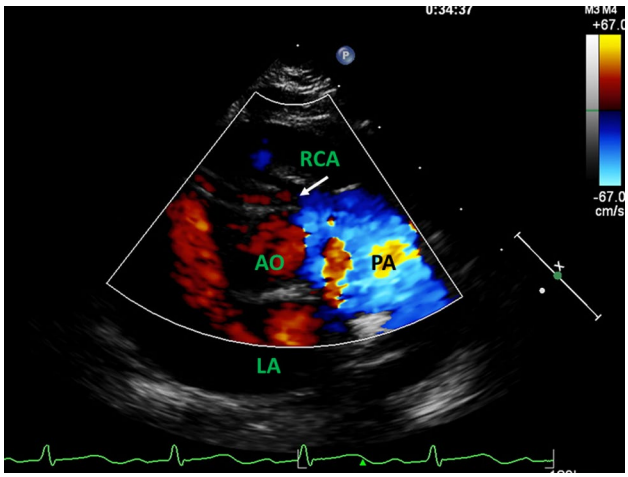

Fig. 5 In case 4, parasternal short-axis view showing a systolic anterior red flow signal from the PA to the RCA (white arrow). *AO* aorta, *LA* left atrium, *PA* pulmonary artery, *RCA* right coronary artery

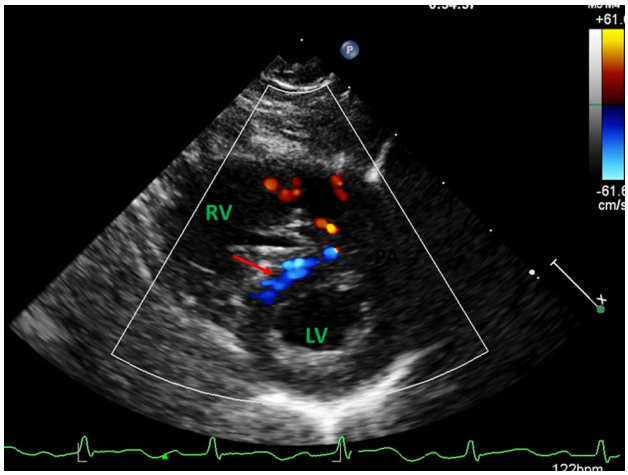

Fig. 6 In case 4, parasternal leftventricular short-axis view color Doppler echocardiography showing prominent intracoronary collaterals within the interventricular septum (red arrow). *LV* left ventricle, *RV* right ventricle

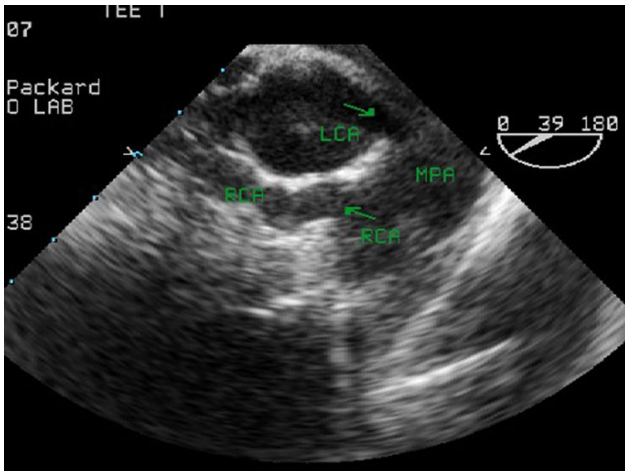

Fig. 7 In case 4, transesophageal echocardiogram clearly showing the anomalous RCA arising from the MPA root and dilated LCA normally arising from the aorta. *LCA* left coronary artery, *MPA* main pulmonary artery, *RCA* right coronary artery

Discussion

Embryology

Abnormalities in embryological development lead to positional anomalies of the coronary arteries. The coronary artery buds appear at about the 12th day of life after division of the truncus arteriosus that leads to the separation of the aorta and the PA, while coronary artery anomalies can arise secondary to either malrotation of the spiral septum dividing the truncus or malpositioning of the coronary

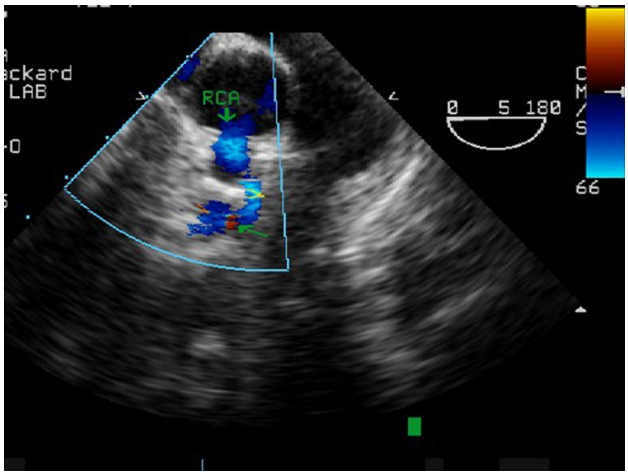

Fig. 8 In case 4, transesophageal echocardiogram showing a widely opened orifice of the RCA to the aorta and color Doppler echocardiography showing antegrade flow from the aorta to the coronary artery. *RCA* right coronary artery

buds themselves [13, 14]. Anomalous origin of the LCA from the PA (ALCAPA) is rather well known, whereas only a few cases of ARCAPA have been described. The higher incidence of ALCAPA compared to ARCAPA may be explained by the proximity of the left coronary bud to the PA sinus [15]. It affects 0.002% of all patients with congenital heart disease. Most ARCAPA cases are isolated anomalies, but approximately 25–30% are associated with other structural heart defects. In our study, the prevalence of ARCAPA was 0.0032% of all cases of congenital heart disease: There were two cases of isolated ARCAPA and four cases associated with atrial septal defects.

Pathophysiology

The pathophysiology of ARCAPA depends on the direction of blood flow in the coronary artery and its influence on oxygen delivery to the myocardium. The severity of ventricular ischemia is determined by shunt size, degree of collateral circulation, and myocardial oxygen demands [3]. Because RV oxygen demands are lower than left ventricular oxygen demands, ventricular ischemia in ARCAPA is less prominent than that in ALCAPA. The clinical diagnosis of ARCAPA is difficult due to a lack of typical signs or symptoms; rather, it is discovered incidentally at the time of surgery or catheterization. Unlike ALCAPA, most cases of ARCAPA are generally not considered lethal defects in infancy or childhood, but it may cause myocardial ischemia and sudden cardiac death, so reimplantation of the RCA into the aorta has been recommended to prevent an adverse outcome.

Table 1 Characteristics of 6 children with ARCAPA

No.	Sex	Age (years)	Preoperative echocardiography diagnosis	Surgical findings	Surgical procedure	Postoperative TEE examination
1	M	11.00	Fistula between PA and LCA	ARCAPA	Reimplantation	Communication between RCA and AAO; no obstruction
2	F	9.00	ARCAPA	ARCAPA	Reimplantation	Communication between RCA and AAO; no obstruction
3	F	18.00	ASD	ASD, ARCAPA	Ligation, ASD repair	No leak of PA ligation; no residual shunt
4	F	0.33	ASD, ARCAPA	ASD, ARCAPA	Ligation, ASD repair	Leak of PA ligation; no residual shunt
5	M	0.38	ASD, ARCAPA	ASD, ARCAPA	Reimplantation, ASD repair	Communication between RCA and AAO; no obstruction; no residual shunt
6	F	10.00	ASD, ARCAPA	ASD, ARCAPA	Reimplantation, ASD repair	Communication between RCA and AAO; no obstruction; no residual shunt

AAO ascending aorta, *ARCAPA* anomalous origin of the right coronary artery from the pulmonary artery, *ASD* atrial septal defect, *LCA* left coronary artery, *RCA* right coronary art

Coronary angiography/CT/echo examination

There are currently several methods that can be used to diagnose ARCAPA. The ECG in cases of ARCAPA may be normal or show left ventricular hypertrophy or deep Q waves in the inferior leads, but it does not appear to be useful for diagnosing ARCAPA. Coronary angiography is the "gold standard" for the preoperative diagnosis of ARCAPA because of its excellent spatial and temporal resolution, but it is not recommended for children with severe conditions since it is invasive and requires the intravenous administration of a contrast agent. Cardiac CT provides the best means by which to demonstrate detailed anatomic information about the origin, course, and relationship of the anomalous coronary artery; however, it is expensive and exposes the child to ionizing radiation [16]. Echocardiography provides a noninvasive, inexpensive, safe, and portable method to delineate the origin of the anomalous coronary artery and the course of the proximal coronary artery; it is possible to diagnose ARCAPA [17]. In our study, four of the six cases were diagnosed correctly, indicating that ARCAPA could be diagnosed by echocardiography.

Although the noninvasive diagnosis of ARCAPA using two-dimensional echocardiography and color Doppler echocardiography is feasible, the size and the course of distal coronary arteries are often difficult to visualize because of poor image quality due to poor acoustic windows and their flexural course [18]. In our study, two cases were misdiagnosed, indicating that it is prone to misdiagnosis.

Differential diagnosis

When a dilated LCA and diastolic or continuous retrograde flow from the abnormal vessel into the main PA are observed by echocardiography, ARCAPA or a coronary fistula should be suspected. In our study, one case was misdiagnosed as a fistula between the PA and the LCA because the origin of the RCA was not clearly visualized. In our cases of ARCAPA, the LCA was dilated and the RCA origin could not be found, and an anomalous diastolic upgrade flow pattern in PA that indicates a "steal effect" from the coronary circulation into the PA and inter-coronary collaterals within the ventricular septum were shown on color Doppler echocardiography. In the cases of coronary fistula, the origin of the RCA from the right sinus of Valsalva could be shown.

When a dilated LCA and inter-coronary collaterals within the ventricular septum are observed on echocardiography, ARCAPA or atresia of the RCA should be suspected. In our cases of ARCAPA, the LCA was dilated and the RCA origin from the PA could be visualized; in contrast, in cases of atresia of the RCA, the origin of the RCA from the PA was not visualized.

Transesophageal echocardiography

The advantages of TEE include the proximity of the transducer to the area of interest as well as a higher-frequency transducer that allows better spatial resolution and a detailed assessment. In our study, TEE clearly showed that the RCA anomalously arose from the main pulmonary root and the dilated LCA normally arose from the aorta, while intraoperative TEE clearly showed a new widely opened orifice of the RCA to the aorta and antegrade flow. However, TEE also has some limitations, including that it is difficult and time-consuming to visualize the entire course of the coronary arteries due to the curvilinear nature of the vessels along the epicardial surface. In addition, some complications occurred during the TEE examination (Table 1).

Follow-up

Reports describing the follow-up of ARCAPA after surgical correction are rare. The follow-up period of our cases was > 6 months; a residual leak was found in one case of simple ligation of the RCA, while no cases of mortality or dysfunction of the right ventricle were detected during the follow-up period. Our study shows that reimplantation techniques for ARCAPA are the most physiological procedures and have good results. However, the risk for potential late complications exists, and long-term patient follow-up is warranted. It may be recommended to perform an annual postoperative echocardiographic screening due to some studies reporting clotting in the reimplanted RCA [19].

Limitations

The most important limitations of the present study were the small number of patients included and its single-center experience. In addition, the follow-up period was relatively short. Further studies of large cohorts are required to enable a more comprehensive analysis of patients with ARCAPA.

Conclusion

In conclusion, if ARCAPA is suspected, an exhaustive TTE search from all echocardiographic windows should be made to determine the origin and course of the RCA and the dilated LCA, while color Doppler echocardiography should be used for detecting retrograde flow from the RCA into the main PA and inter-coronary collaterals within the ventricular septum. Based on these characteristic echocardiographic findings, ARCAPA could be diagnosed accurately. TEE was very useful in evaluating the immediate surgical results. If the origins of the coronary artery are unclear, digital subtraction angiography or CT should be performed prior to the surgical intervention.

Compliance with ethical standards

Conflict of interest The authors declare that there are no conflicts of interest regarding the publication of this paper.

Ethical considerations All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008. This study was approved by Shanghai Children's Medical Center, Shanghai Jiaotong University School of Medicine. Informed consent was obtained from all patients included in the study.

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