The Turkish Journal of Pediatrics 1997; 39: 127-135

ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY*

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SUMMARY: Sarıoğlu A, Saltık İL, Sağın-Saylam G, Batmaz G, Sarıoğlu T, Aytaç A. (Pediatric Cardiology Unit, İstanbul University Institute of Cardiology, İstanbul, Turkey). Anomalous origin of the left coronary artery from the pulmonary artery. Turk J Pediatr 1997; 39: 127-135.

Anomalous origin of the left coronary artery from the pulmonary artery (ALCA-PA) is a rare form of congenital heart disease. In this report, three cases with this anomaly are described; two patients presented in infancy with heart failure from myocardial ischemia and infarction, while the third was asymptomatic and ALCA-PA was diagnosed during evaluation of a residual murmur after surgery for associated cardiac defects (ventricular septal defect and patent arterial duct). All three cases underwent aorto-pulmonary tunnel repair (Takeuchi procedure), and to our knowledge two of them are the first infantile cases reported in Turkey. *Key words: coronary artery anomalies, pulmonary artery.*

Anomalous origin of the left coronary artery from the pulmonary artery (ALCA-PA) is a rare congenital anomaly that constitutes 0.24 percent of all congenital cardiac defects, with a reported incidence of 1/300,000[1]. As the mortality rate of untreated patients in the first year of life has been reported to be as high as 93 percent, it is an important form of congenital heart disease with a wide spectrum of clinical presentation[2]. In this report, three cases of ALCA-PA presenting with different clinical features are described.

Case Reports

Case 1

A two-year-old boy known to have congenital heart disease since birth was referred to our clinic for evaluation of his heart conditon. Physical examination, echocardiography, cardiac catheterization and angiography revealed a large perimembranous ventricular septal defect (VSD) and a patent arterial duct (PDA). At the age of three years, patch closure of a 2 x 2 cm VSD and double ligation of the PDA were performed. On postoperative follow-up one year later, a continuous murmur at the 3rd left intercostal space was noticed. Two-dimensional

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echocardiography showed that the left atrium, left ventricle and pulmonary artery were enlarged and the right coronary artery (RCA) was dilated (5 mm). HPRF and color Doppler examination revealed turbulence and continuous flow at the right ventricular apex and the mid-muscular septum (Fig. 1). Repeat cardiac catheterization showed mild pulmonary hypertension (42/26 mmHg) and a step-up in oxygen saturation in the pulmonary artery (Table I). Angiography showed that the RCA was dilated, and the left coronary artery (LCA) was originating from the main pulmonary artery and filled in retrograde fashion from the RCA through collaterals (Fig. 2). The patient underwent "tunnel repair" at the age of five years. The postoperative course was uneventful, and subsequent angiographic study revealed a patent LCA filling from the aorta (Fig. 3).

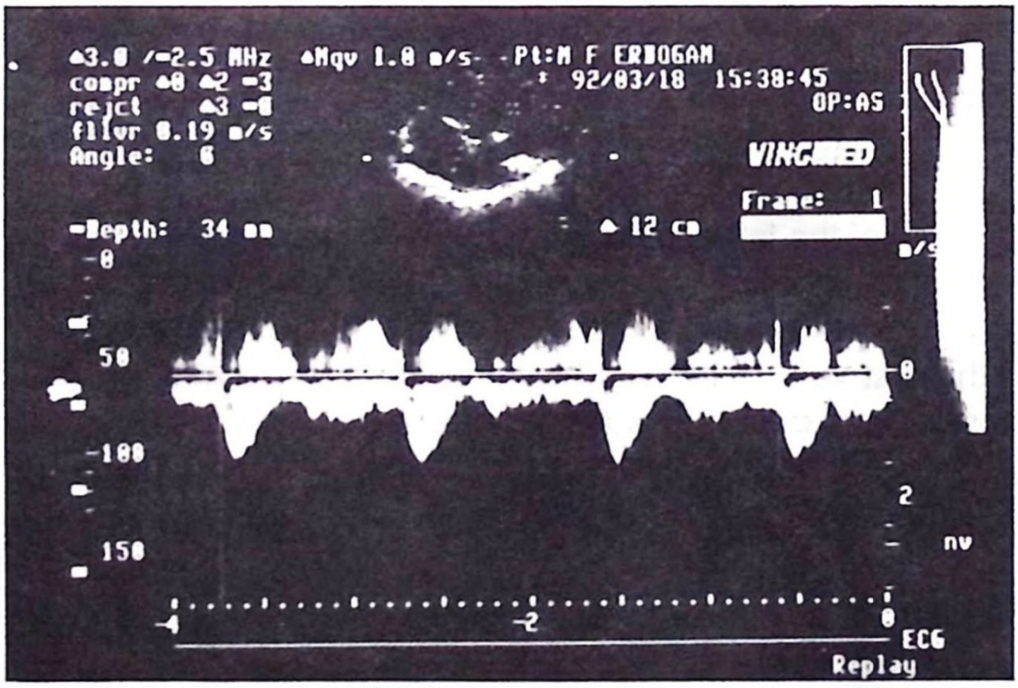


Fig. 1: Sampling by HPRF Doppler in the midmuscular septum showing the presence of continuous flow (Case 1).

Table I: Hemodynamic Findings in Three Cases with ALCA-PA

	Case 1		Case 2		Case 3*
	Pressure** (mmHg)	Oxygen saturation (%)	Pressure (mmHg)	Oxygen saturation (%)	Pressure (mmHg)
Right atrium	15/10 (13)	74.9%	11/6 (8)	27.0%	–
Right ventricle	44/6-12	73%	42/5-10	26%	–
Main pulmonary artery	42/26 (32)	82.7%	35/15 (25)	27%	–
Left ventricle	110/0-10	97.3%	90/2-15	–	75/0-20
Aorta	110/78 (94)	97.3%	90/55 (60)	–	75/38 (50)

* Right heart catheterization was not performed.
** Values in parentheses indicate mean pressures.

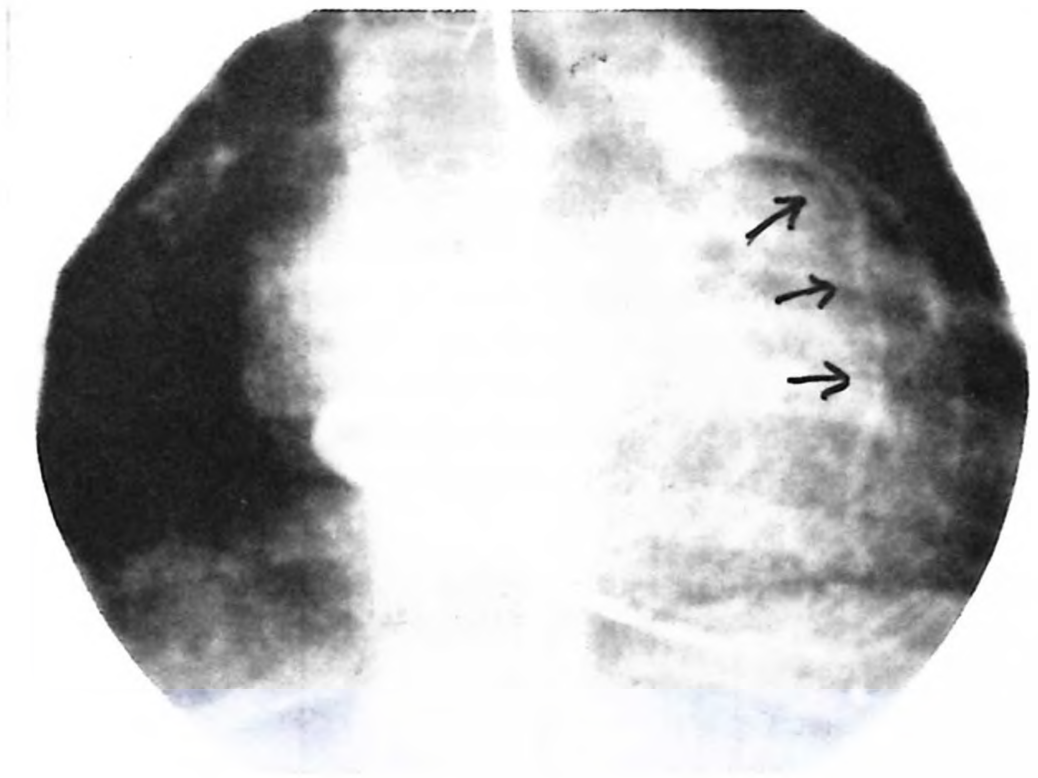
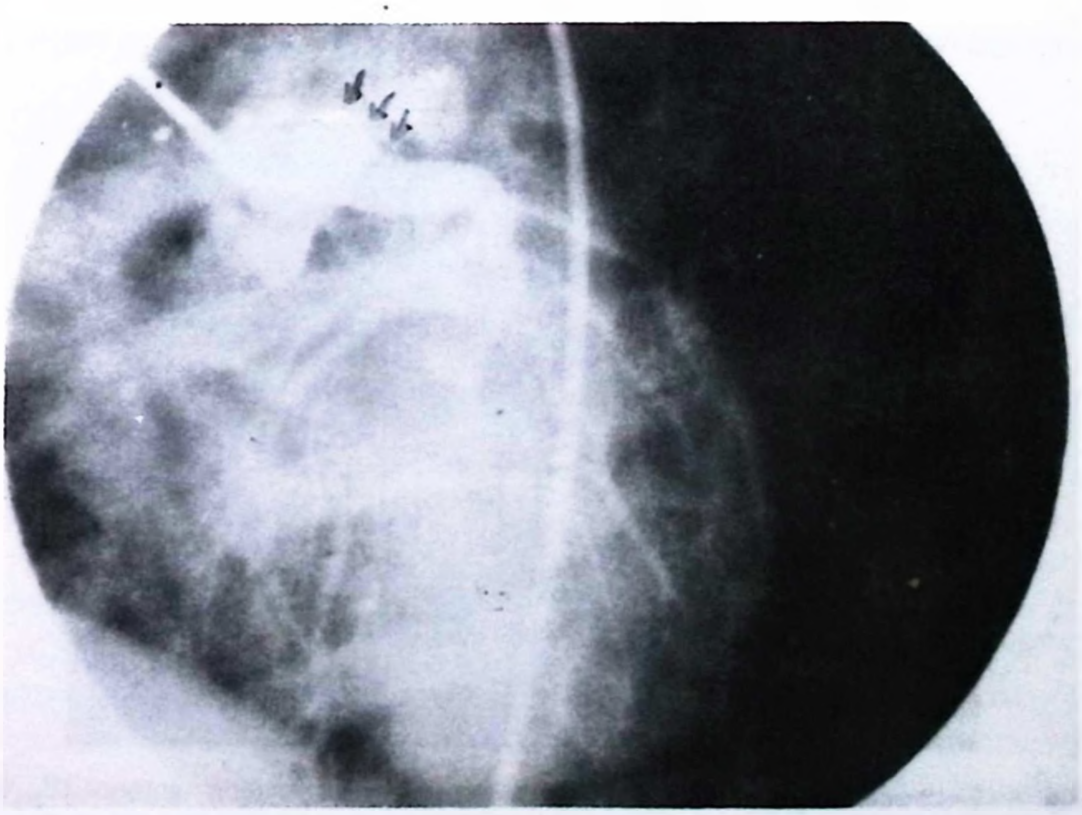


Fig. 2: Preoperative selective right coronary arteriogram in the 45° right arterior oblique view (Case 1) showing dilated RCA and LCA (arrows) filling via collaterals.



Figrue 3: Postoperative left coronary arteriogram in the 45° left anterior oblique view (Case 1) showing the aorta-pulmonary tunnel (arrows) and normal filling of the LCA.

Case 2

A three-month-old girl presented with tachypnea and failure to thrive. She was the 3300 g product of an uneventful pregnancy and delivery. She was noted to have poor feeding and respiratory difficulty since birth and was referred to our clinic for evaluation of a heart murmur. On physical examination, she had tachypnea, dyspnea and tachycardia. The femoral pulses were equal and symmetrical, a grade 2/6 short systolic murmur was audible at the 4^{th}-5^{th} left intercostal space, and the liver edge was palpable 4 cm below the right costal margin. The electrocardiogram showed left-axis deviation, left ventricular hypertrophy and an anterolateral myocardial infarction pattern (Fig. 4). Chest x-ray revealed cardiomegaly (CTR = 0.62) with pulmonary venous congestion.

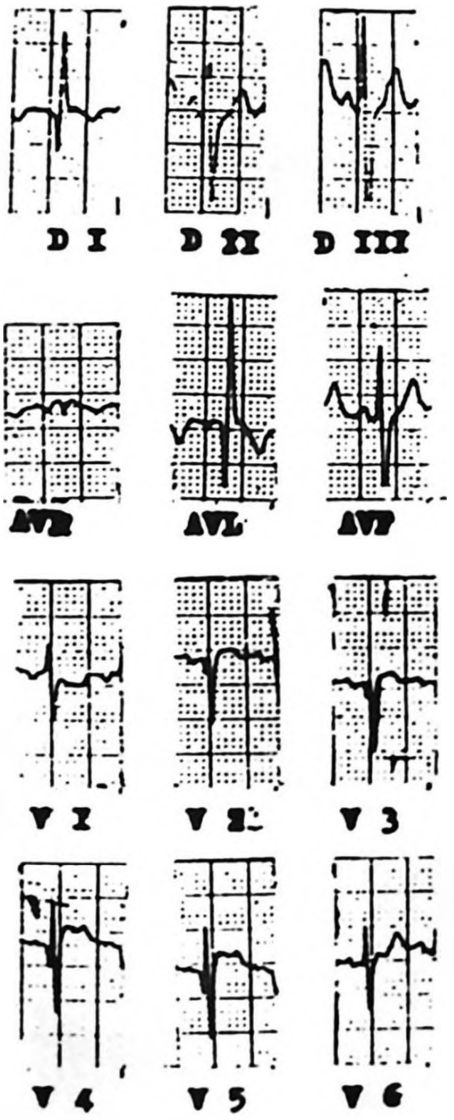


Fig. 4: Electrocardiography of Case 2 showing left-axis deviation, anterior (V_{2-5}) and lateral (lead I, AVL) myocardial infarction pattern, ST segment elevation in V_{2-5} and negative T waves in lead I, AVL.

Two-dimensional color Doppler echocardiography demonstrated an enlarged left atrium, a dilated and poorly contracting left ventricle (fractional shortening = 26%), moderate mitral regurgitation and turbulent flow in the interventricular septum. The aortic origin of the LCA could not be visualized, whereas that of the RCA was well demonstrated. Cardiac catheterization showed a step-up in oxygen saturation in the pulmonary artery (Table I), and the left ventriculogram revealed a dilated chamber with poor contractions. Aortic root injections revealed a dilated RCA, the LCA was visualized after the RCA, filling in retrograde fashion via collaterals, and the main pulmonary artery was opacified through the LCA. Tunnel repair was performed when the patient was seven months old; a zone of anterolateral myocardial infarction was noticed during surgery. The patient died of purulent meningitis and sepsis five days postoperatively.

Case 3

A one-month-old girl who presented with grunting since birth was referred from another hospital because of cardiomegaly. On physical examination, she had tachypnea, dyspnea, and tachycardia. The femoral pulses were equal and symmetrical, and a grade 2/6 short systolic murmur was audible at the 5th intercostal space. The liver edge was palpable two centimeters below the right costal margin. The electrocardiogram revealed left ventricular hypertrophy and a lateral myocardial infarction pattern. Chest x-ray showed cardiomegaly (CTR = 0.66). Two-dimensional echocardiography revealed a grossly dilated, spherical, poorly contracting left ventricle (fractional shortening = 10%), a dilated RCA (3 mm) and LCA originating from the main pulmonary artery (Fig. 5). Color Doppler examination demonstrated

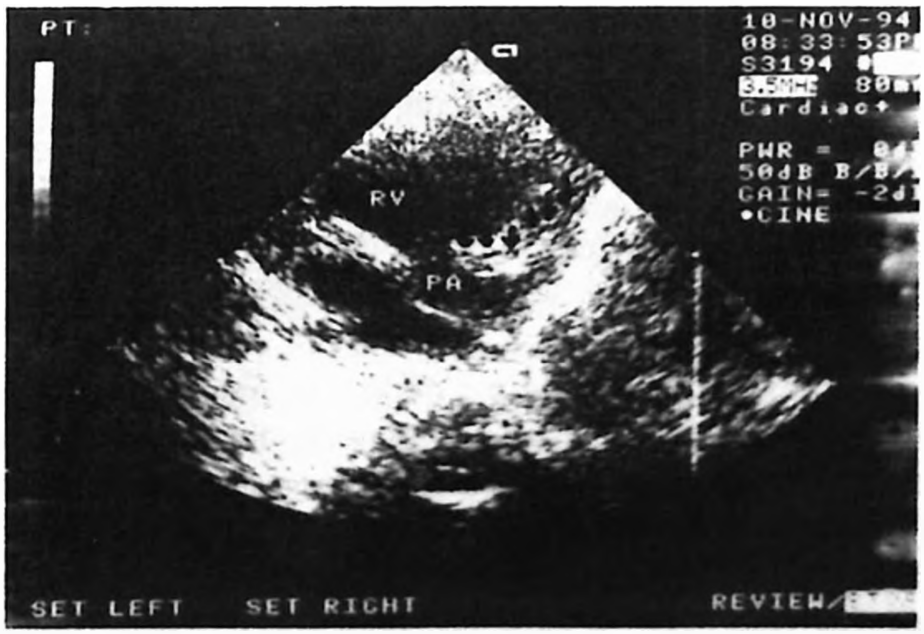


Fig. 5: Two-dimensional echocardiogram in the parasternal short-axis view (Case 3) showing the LCA (arrows) originating from the pulmonary artery.

that the direction of flow in the LCA was towards the main pulmonary artery. Aortography showed a dilated RCA, no origin of the LCA from the aortic root (Fig. 6) with late visualization of the LCA and opacification of the main pulmonary artery through the LCA. The patient underwent tunnel repair at 2.5 months of age. The postoperative course was uneventful. On follow-up nine months later, echocardiography showed that the heart chambers were of normal dimensions and left ventricular contractility was normal (fractional shortening = 38%).

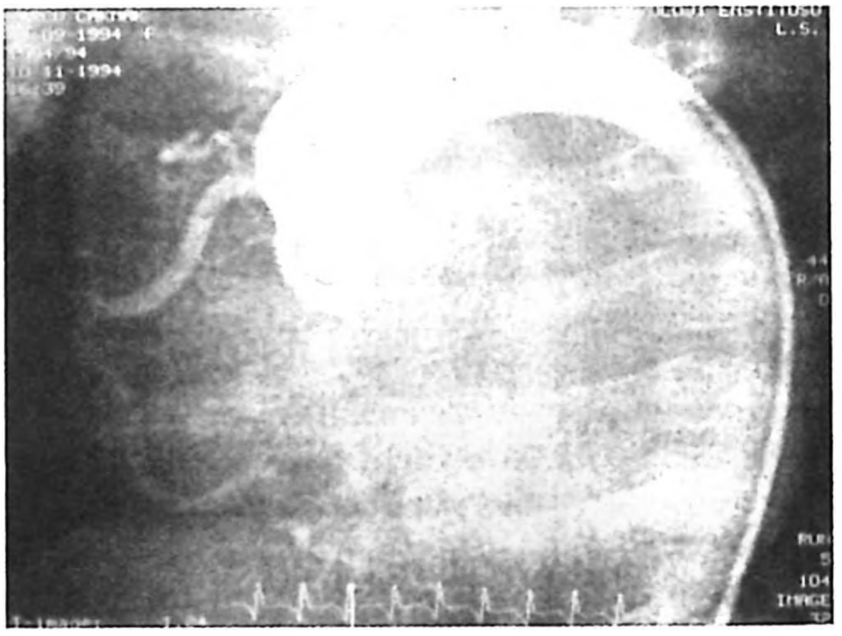


Fig. 6: Preoperative aortogram in 45° left anterior oblique view in Case 3 showing a dilated RCA filling from the aorta with no filling of the LCA.

Discussion

ALCA-PA, also known as Bland-White-Garland syndrome[3], is the most important coronoray anomaly managed by pediatricians and pediatric cardiologists. The clinical spectrum in ALCA-PA may range from the "infantile" form, presenting with signs and symptoms of myocardial infarction and congestive heart failure in infancy, to the "adult" form, which may be asymptomatic and detected in adulthood or on autopsy[4].

In intrauterine life and the early newborn period, pulmonary artery pressure and pulmonary vascular resistance are high; therefore flow in the anomalous LCA originating from the pulmonary artery is antegrade from the pulmonary artery, maintaining adequate myocardial perfusion. As pulmonary artery pressure and pulmonary vascular resistance decrease, antegrade flow from the pulmonary artery through the LCA decreases and myocardial perfusion depends on the presence of collateral circulation between the right and left coronary arteries. If collateral circulation is inadequate, myocardial infarction is likely to occur.

The clinical presentation of such patients includes dyspnea, tachypnea and signs of congestive heart failure, as in Cases 2 and 3. The majority of patients with ALCA-PA present in infancy with heart failure. Wesselhoeft et al.[2], in a review of the literature, found that 116 of 140 patients were diagnosed in infancy. Thirteen out of 15 cases reported by Askenazi and Nadas[5], and 10 out of 11 cases reported by Perry and Scott[6] were of the infantile type. On the other hand, if there is well-developed collateral circulation, as in our Case 1, the patient may be asymptomatic, the only physical sign being a continuous murmur on auscultation. This rare form constitutes only 9 of 140 patients reported in the literature review by Wesselhoeft et al[2]. The only two cases reported in Turkey[7,8] were of the adult type.

ALCA-PA is usually an isolated anomaly, although cases associated with VSD, PDA, pulmonary stenosis and Fallot's tetralogy have been reported[9-11]. In such cases the clinical picture may be dominated by the findings of the associated cardiac defect, and the diagnosis of ALCA-PA may be overlooked. All five cases reported in three articles[9-11] underwent surgery for the associated defect, but ALCA-PA was diagnosed perioperatively in one case, one year following the operation in one, and on autopsy in three. In our Case 1, ALCA-PA was detected one year after open heart surgery for VSD and PDA. The presence of VSD and PDA in this patient may have resulted in an increase in pulmonary artery pressure, thereby permitting the development of adequate collateral circulation.

The electrocardiogram is an important diagnostic tool in infants with ALCA-PA, often demonstrating anterior, anteroseptal or anterolateral myocardial ischemia and infarction patterns[12]. Our Case 2 displayed anterior myocardial infarction characterized by "Q" waves in leads I, AVL, V_{4-5}, and typical ST changes, while Case 3 had a lateral myocardial infarction pattern.

Echocardiography has proved to be useful in the definitive noninvasive diagnosis of ALCA-PA. Two-dimensional echocardiographic findings o ALCA-PA include failure to visualize the aortic origins of two separate coronary arteries, demonstration of a dilated RCA, and direct visualization of the LCA originating from the main pulmonary artery[13-15]. Retrograde flow through the anomalous LCA towards the pulmonary artery can be demonstrated by pulsed wave and color flow Doppler in the parasternal short-axis view[16-18]. Another finding that should alert the echocardiographer to the diagnosis of ALCA-PA is detection of turbulent and continous flow by pulsed wave and color flow Doppler in the interventricular septum. This finding was present in two of our patients (Case 1 and 2) in addition to the presence of a dilated RCA and failure to visualize the aortic origin of the LCA. The importance of this finding has recently been emphasized by others[19]. In Case 3, turbulent flow was not detected in the interventricular septum, but flow in the LCA towards the pulmonary artery was clearly visualized.

The ideal treatment of ALCA-PA is early surgical intervention before myocardial infarction occurs, but unfortunately all cases in infancy come to medical attention only after myocardial ischemia or infarction have occurred. Numerous operative techniques described for the treatment of patients with ALCA-PA include ligation of the LCA, subclavian-to-LCA anastomosis, reimplantation of the ostium of the LCA onto the aorta, saphenous vein by-pass grafting and creation of an aortopulmonary tunnel (Takeuchi procedure)[20-25]. All three cases in this report underwent the Takeuchi procedure.

In conclusion, we would like to emphasize that in the differential diagnosis of unexplained heart failure or cardiomyopathy in infancy and in asymptomatic children under investigation for the presence of precordial continuous murmurs, ALCA-PA should be considered and the coronary arterial anatomy should be carefully delineated by echocardiography. Failure to visualize the aortic origin of the LCA, together with demonstration of a dilated RCA turbulent and continuous flow in the interventricular septum with pulsed wave and color flow Doppler, are invaluable echocardiographic fingins for the diagnosis of ALCA-PA.

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