

An incidentally detected anomalous origin of the right coronary artery from the pulmonary artery in an infant

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ABSTRACT

Isolated anomalous origin of the right coronary artery from the main pulmonary artery is a rare congenital anomaly, and few cases have been reported in the pediatric age group. Here in, we report an asymptomatic case of a 2-month-old male infant who has been diagnosed as anomalous origin of the right coronary artery from the main pulmonary artery during the evaluation for cardiac abnormalities. For a suspicion on echocardiography, cardiac catheterization and coronary angiography performed to verify the diagnosis of anomalous origin of the right coronary artery from the main pulmonary artery. The patient underwent surgery and did well after two months follow up. Early diagnosis may prevent patients from cardiovascular complications.

Key words: Echocardiography, coronary artery, pulmonary artery, anomalies, cardiac catheterization.

<http://dx.doi.org/10.5546/aap.2018.eng.e102>

To cite: Ugan Atik S, Saltik L, Oztarhan K, et al. An incidentally detected anomalous origin of the right coronary artery from the pulmonary artery in an infant. *Arch Argent Pediatr* 2018;116(1):e102-e105.

INTRODUCTION

Isolated anomalous origin of the right coronary artery from the main pulmonary artery (ARCA-PA) is a rare congenital anomaly, and few cases have been reported in the pediatric age group.¹ The first case series was described by Brooks in 1885.² Based on a literature review done by Modi et al. in 2010, 20 cases were diagnosed in infants (\leq one year of age), 44 cases were diagnosed in children (\leq 18 years of age), 17 cases were in adults ($>$ 60 years of age), and in eight cases, the age was not recorded.³ Patients with

ARCA-PA are usually asymptomatic, although there are some case reports of sudden death, cardiomyopathy, angina and syncope.⁴ Here in, we report an asymptomatic case of a 2-month-old male infant who has been diagnosed as ARCA-PA during the evaluation for cardiac abnormalities.

CASE

A 2-month-old male infant (4.5 kg) was referred to our outpatient clinic for cardiac evaluation. The baby weighing 3200 g was born at 38 weeks of gestation by cesarean section and discharged within 24 h after a routine check-up. In his initial admission, his mother's complaint was perioral cyanosis but saturation was normal. Physical examination showed a nonspecific 1/6 systolic murmur on the left side of sternum. There were no other associated anomalies in his physical examination. His blood tests were in normal limits. Chest X-ray demonstrated mild cardiomegaly. An electrocardiogram showed sinus tachycardia with a right axis deviation. On echocardiography left ventricular and right ventricular functions were normal, multiple colour Doppler signals were detected within the interventricular septum highly suspicious for intracoronary collaterals (*Figure 1a*). The left main coronary artery was mildly dilated and the orifice of the right coronary artery entered the pulmonary artery few millimeters distal of the pulmonary valve (*Figure 1b*). Fistulous flow from right coronary artery orifice to pulmonary artery was detected by colour Doppler.

Cardiac catheterization and coronary angiography performed to verify the diagnosis of ARCA-PA. Selective left coronary artery angiography showed retrograde filling of the right coronary artery through collaterals from the left coronary artery and the right coronary artery ostium connected to the pulmonary artery (*Figure 2*). Diagnosis of ARCA-PA was confirmed by this angiography. After discussing the case and based on the well-known potential risk of sudden death, it was decided to send the patient to a surgical direct reimplantation of the right coronary artery. After reimplantation of the right coronary artery into the aorta which was

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Funding: None.

Conflict of interest: None.

Received: 3-25-2017

Accepted: 9-6-2017

performed in another surgery clinic, the patient did well during seven months follow-up.

DISCUSSION

Congenital coronary artery anomalies are rare malformations in general population. According to the Greenberg et al., major anomalies leading to abnormal myocardial perfusion are anomalous origin from the pulmonary artery, an origin of coronary artery from opposite or non-coronary sinus, myocardial bridging and coronary artery fistula. Among anomalous origin from the pulmonary artery, anomalous origin of left coronary artery from the pulmonary artery

(ALCA-PA) is more common.⁵ Anomalous origin of the right coronary artery from the pulmonary artery is rare than ALCA-PA (only 200 cases have been reported in total).¹ As many patients remain asymptomatic, the true incidence of this coronary anomaly might be higher.² In 70% of reported cases, it is an isolated anomaly. However, concomitant congenital cardiac defects such as aortopulmonary window and tetralogy of Fallot have been previously reported in association with the ARCA-PA syndrome.^{6,7}

Generally, these patients are asymptomatic until adulthood because of an extensive collateralization between the two coronary

FIGURE 1a: Multiple colour Doppler signals within the interventricular septum

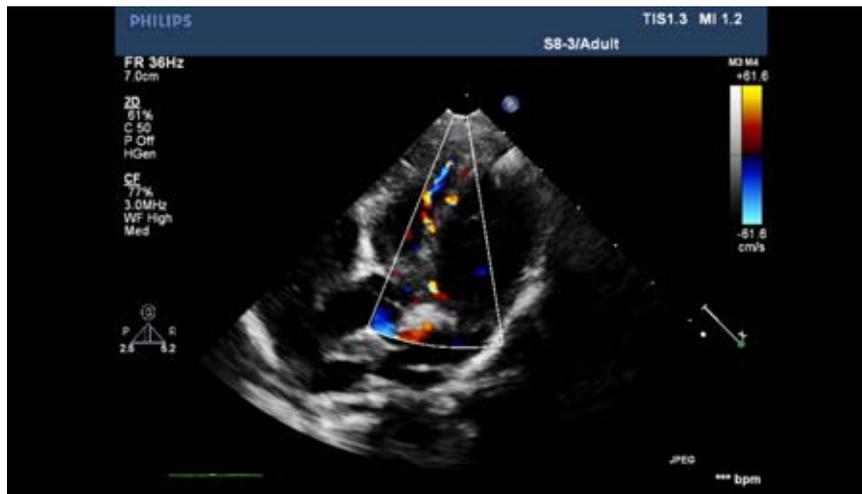


FIGURE 1b: Fistulous flow (arrows) from right coronary artery orifice to pulmonary artery detected by colour Doppler.



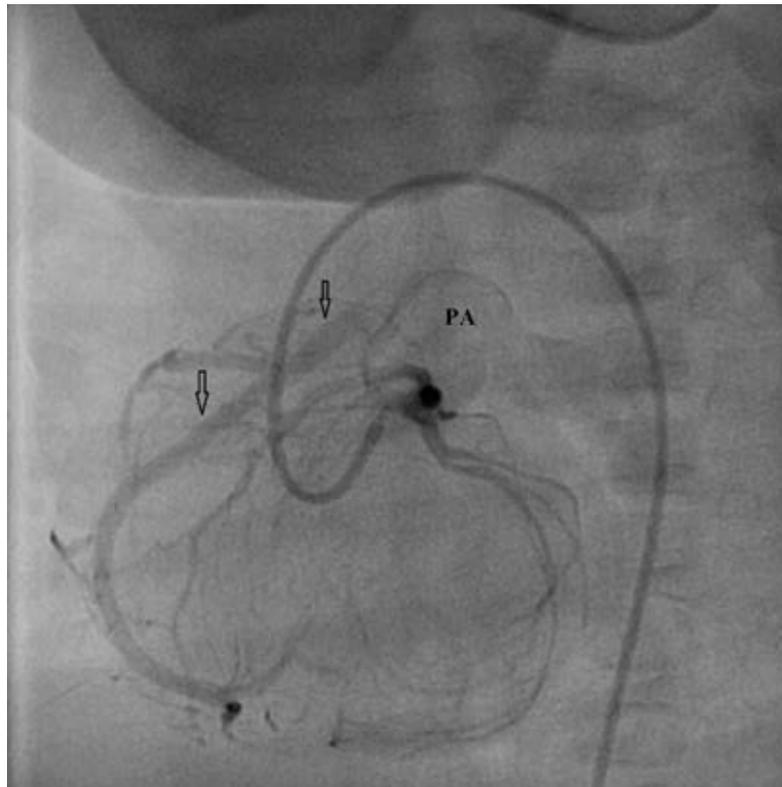
systems preserving good ventricular function.⁸ The presentation of ARCA-PA is non-uniform, patients may have nonspecific physical examination findings or dyspnea, fatigue, congestive heart failure, angina, myocardial infarction and even sudden cardiac arrest may be the first complaint.⁸ Cardiac evaluation may be triggered by a nonspecific complaint or physical examination findings such as heart murmur. In our case, although his mother was complaining about perioral cyanosis, patient's saturation was normal and the only physical finding was nonspecific 1/6 systolic murmur on the left side of sternum. On the other hand, as reported by Lin et al., the first presentation may be acute heart failure in an infant.⁹

In 1985, the first case of ARCA-PA was diagnosed by echocardiography.¹⁰ Certain echocardiographic findings, such as intracoronary collaterals within the ventricular septum visualized by color flow Doppler, has been reported to be highly suggestive of ARCA-PA.¹¹ In addition, like our patient, dilated coronary

arteries and a retrograde flow from the right coronary artery into the pulmonary artery is another characteristic echocardiographic finding for this disease.^{12,13} Coronary artery imaging with echocardiography may be difficult in some patients due to poor acoustic windows. Multislice CT and cardiovascular MRI which are non-invasive imaging modalities with high diagnostic accuracy for coronary artery anatomic anomalies may be another options for diagnosis.¹⁴

Therapeutic recommendations for ARCA-PA patients are controversial. Surgical techniques available include simple ligation of the right coronary artery, ligation of the right coronary artery with saphenous vein bypass grafting and reimplantation of the right coronary artery into the aorta.¹⁵ Many authors advocate that, a two-coronary system should be established by direct reimplantation of the RCA into the ascending aorta whenever possible and especially in children.⁵ In the light of these recommendations, we suggested reimplantation of the right coronary artery into the aortic root soon after the diagnosis

FIGURE 2: Retrograde filling of the right coronary artery (arrows) through collaterals from the left coronary artery and the right coronary artery ostium connected to the pulmonary artery



due to the potentially fatal outcome.

In conclusion, clinicians should be aware of this rare anomaly to make early diagnosis. When multiple colour Doppler signals are detected at interventricular septum in echocardiographic evaluation, one must pay attention to coronary artery orifices. When a dilated left coronary artery is observed in the echocardiogram, ARCA-PA or atresia of the ostium of the right coronary artery or a coronary fistula, which are the differential diagnoses, should be suspected. Early diagnosis may prevent patients from cardiovascular complications. ■

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