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Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA): echocardiographic diagnosis in a critically ill newborn.

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Abstract

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a very rare disease. Echocardiographic diagnosis can be very challenging, especially in both asymptomatic children with no history of cardiac disease or critically ill newborns in the setting of intensive care unit. We report a case of ARCAPA in a neonate with congenital pulmonary airway malformation (CPAM), whose echocardiographic diagnosis was particularly challenging due to the critical status at presentation.

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Abstract

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a very rare disease. Echocardiographic diagnosis can be very challenging, especially in both asymptomatic children with no history of cardiac disease or critically ill newborns in the setting of intensive care unit. We report a case of ARCAPA in a neonate with congenital pulmonary airway malformation (CPAM), whose echocardiographic diagnosis was particularly challenging due to the critical status at presentation.

Case report

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A neonate with prenatal diagnosis of CPAM, born from cesarean section at 35 weeks of gestation and 2.5 kg, was led in neonatal intensive care for respiratory failure at birth. He was intubated and mechanically assisted with high-frequency oscillatory ventilation (HFOV). At that time, first urgent cardiac ultrasound (CUS), with poor acoustic window, showed normal origin and course of the coronary branches and systemic right ventricular pressures (Fig 1a, b). At day2 for worsening clinical conditions he was put on veno-arterial extracorporeal membrane oxygenation (VA-ECMO). At day7 he underwent right upper and medium lobectomy for removal of CPAM III and at day 16 he was weaned-off ECMO. Serial CUS showed progressive reduction of pulmonary arterial pressure (PAP). At day52 CUS showed the presence of a small network of multiple coronary vessels in the field of the right coronary artery (RCA), a systo-diastolic jet of flow into the main pulmonary artery (MPA) at color-mapping and an inverted retrograde flow into the RCA (Fig 2a, b – video 1, 2 and 3). A more detailed examination showed a suspected abnormal origin of the RCA from the main pulmonary artery (ARCAPA) (Fig 2c – video 4). At day59 CT scan confirmed the echocardiographic diagnosis of ARCAPA origination from the medial wall of the MPA (Fig 2d and 3) and dilation of the main epicardial coronary network due to the presence of a multiple coronary collateral connections. After recovery from thoracic surgery and multidisciplinary review of the case, the patient was discharged at home with strict follow-up and future evaluation for elective coronary surgery.

Discussion

ARCAPA is an extremely rare congenital heart disease (CHD) firstly described by the Irish anatomist John Brooks in 1885 . Its incidence, based on coronary angiographic procedures, is estimated around 0.002%, but, given the usually asymptomatic behavior of this anomaly, its real prevalence in general population might be higher . Compared to the more common anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), ARCAPA is associated with lower mortality rate in infancy and childhood and has a bimodal age presentation with peaks near after birth or in the middle 40-60s . ARCAPA is often diagnosed incidentally and usually is an isolated lesion. Sometimes it is associated with other malformations : more often CHD (23.8%), less frequently extracardiac and genetic syndromes . As described, the most frequent associated CHDs are aorto-pulmonary window and ventricular septal defect , in rare cases aortic arch hypoplasia or patent ductus arteriosus with coronary fistulae . In 4% of cases ARCAPA has been found in patients with extracardiac lesions such as anal atresia, tracheoesophageal fistula, limb anomalies or other . Our case is the first described in a patient with CPAM. Historically, embryological origin of coronary anomalies is thought to be linked to defects in arterial septation, the same process that, around the 12th day of life, leads to the formation of aorta and pulmonary trunk from the common truncus arteriosus . Another theory advocates abnormal signaling in pathways that regulate implantation of the primary coronary plexus to the aorta or pulmonary trunk . Embryologic causes of CPAM, which results from the cessation of lung development during various stages of embryogenesis , seems to be different and not linked to those of coronary anomalies.

Pathology reports described the anomalous coronary artery made different for the normal coronaries: thin walled, dilated and/or vein-like . Precise origin of the abnormal vessel is not always described in literature reports but it can originate from any site off the pulmonary trunk wall: the anterior or posterior sinus of Valsalva, the anterior aspect of trunk, or more frequently distal to the pulmonary valve . In our case ARCAPA took-off from the medial wall of the pulmonary trunk, distal to the anterior Valsalva sinus. A web of small vessels (collaterals) nearby the ostium (as in our case) or stenosis of the origin of ARCAPA have been reported .

Diagnosis and pathophysiology.

Various tools are available for diagnosis of ARCPA. Conventionally, coronary angiography is the gold standard for its ability to visualize the retrograde flow through the RCA, extensive collateralization between RCA and LCA, dilation and increased flow into the epicardial coronary system and direct flow between RCA and MPA . Recently, echocardiography and computed tomography/magnetic resonance (CT/MR) can be fundamental for diagnosis of ARCPA, thanks to the improvements of these technologies. Echocardiography plays a crucial role as a screening, low-cost and diffusely used tool. It can provide first and immediate

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suspicion, and definitive diagnosis, in about 25% of cases of ARCAPA/ALCAPA , thus improving survival of asymptomatic patients. However, echocardiographic diagnosis of coronary anomaly is not simple. In our case and in other reports , it is very easy to confuse the origin of the RCA. As our figure 1 shows, also in the presence of ARCAPA the sonographer can often visualize an "apparently normal" RCA-origin for the right aortic sinus of Valsalva: this ultrasound artifact may be due to the contact of the aortic wall with the proximal part of the RCA usually coursing strictly nearby and leading to erroneous evidence of the vessel originating from the aorta. If not highly suspected, for example during a pediatric routine screening ultrasound, the operator would never search for a coronary origin from the MPA, thus leading to misdiagnosis. The evidence of retrograde (commonly "blue", away from the probe) systo-diastolic flow into the RCA, is the key diagnostic sign and should mandatorily induce the sonographer to search for ARCAPA. To be noted that, as opposed to the normal pulsatile coronary blood flow which is visible at color-doppler during diastole , the blood flow in the abnormal vessel is present both during systole and diastole because the pressure into the coronary system (derived from the left coronary artery and from the wall stress into the myocardium) is higher than the pulmonary pressure. Sometimes a proper jet of blood flow draining into the MPA (fig 2b - the amount of blood shunting from the left to the right system and increasing the Qp/Qs) can be identified at the color-doppler mapping (commonly "blue", away from the probe) and should "ring-the-bell" about the presence of ARCAPA to the sonographer. Multiple small tortuous vessels, resembling coronary fistulae (i.e. collateralizations), with low-velocity and retrograde flow in the area of the proximal RCA course are also key signs of ARCPA. However, as in this case, collateralizations do not develop soon after birth, as sometimes they may only slowly develop during infancy. Moreover, retrograde inverted (left-to-right) flow in RCA appears only after PAP declines, but this may take days, weeks or months, especially in critically ill patients, ECMO patients or newborn with persistently high PAP for several reasons. Our case is a peculiar example since several obstacles to the prompt diagnosis were faced: the neonate was on HFOV for several days, thus preventing a good acoustic window, and he was put on ECMO at day 2 and this might have delayed the PAP decrease. As showed , ECMO circuit alters the coronary flow which can become, in some cases, continuous instead of normally pulsatile . This may complicate the color-doppler analysis of the coronary flow pattern.

All the previous discussions highlight the importance of an echocardiographic revision in patients with critical acuteness, poor acoustic windows, and transient hemodynamic conditions for diagnosis of anomalies that might have been missed at first CUS. In our center our clinical practice includes a second complete screening CUS for critically ill newborns after resolution of the acute event and before discharge.

The pathophysiology of myocardial ischemia in ARCAPA is dependent from the PAP and the collateralizations. High PAP after birth drives antegrade flow of deoxygenated blood from MPA to RCA . As PAP decreases, inverted systo-diastolic retrograde flow runs from the myocardium to the MPA and causes a "steal phenomenon", the reason for progressive chronic myocardial ischemia in children. If collateralization between RCA and LCA is not sufficient symptoms and signs of myocardial ischemia occur earlier in infancy, while, if collateralization is wide enough and PAP remain low, oxygenated blood from LCA would perfuse the myocardium and the patient would reach adulthood asymptomatic (about 48% of cases). Vessel diameter, pulmonary resistances and coronary dominance also influence the amount of coronary steal and the subsequent ischemia. It has been estimated to be up to 1/1.4 L/min (20-25% of the cardiac output, increasing the Qp/Qs).

Treatment.

Ligation or percutaneous occlusion of the ARCAPA have been performed in the past with relief of the left-to-right shunt but with concerns about myocardial ischemia . The most common surgical strategy nowadays is coronary reimplantation. With good results and low risk profile in experienced centers, it guarantees antegrade and oxygenated blood flow to the myocardium, establishing dual coronary artery system and preventing the steal phenomenon .

Indications for surgery of ARCAPA are based on general consensus, since for the rarity of the disease randomized trials and observational studies are lacking. In adults, according to the 2020 ESC guidelines,

surgical intervention due to ARCAPA is needed when symptoms or ventricular dysfunction or other signs of myocardial ischemia are present as a consequence of the malformation . Since ARCAPA can lead to increased risk of myocardial infarction and sudden cardiac death even in asymptomatic patients, surgical correction is recommended in all cases also in infancy and adulthood. Symptomatic patients should receive surgical correction (RCA reimplantation onto the aorta) as soon as possible to relief myocardial damage, unless, especially older patients, deemed inoperable and good candidates for ligation or occlusion of ARCAPA . Asymptomatic patients should receive elective surgical reimplantation : this approach is practically applied in 90% of asymptomatic patients and in our center accordingly. In neonates, usually asymptomatic, elective repair should be planned when they reach a good weight in the first months of life. Our patient is planned to receive surgery in few months (around 6 kg).

Conclusion

ARCAPA diagnosis is often challenging especially in neonates, usually asymptomatic, and in critically ill patients with ventilatory mechanic assistance or poor acoustic windows. Given its peculiar pathophysiology, hemodynamic conditions (such as ECMO, high pulmonary pressure, etc ...) can make its recognition very difficult. Echocardiography is a precious tool in expert hands and subtle pitfalls, depending on hemodynamic conditions, should always be taken into consideration. Although less aggressive than ALCAPA, ARCAPA leads to myocardial ischemia and also in asymptomatic neonates or children elective surgical repair is recommended.

Figures

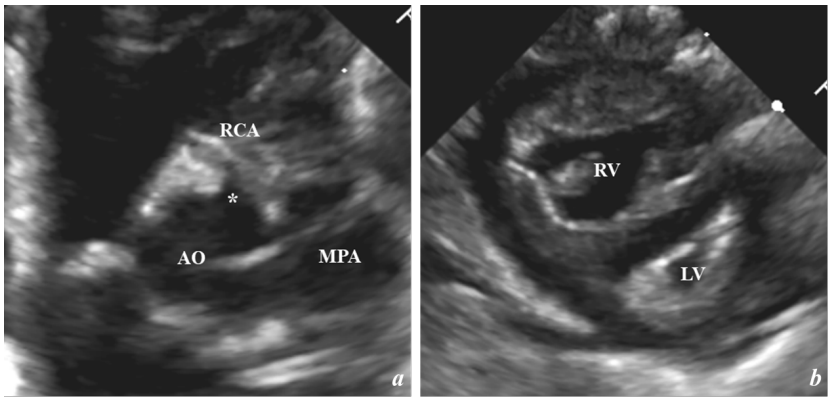

Fig 1. a. The «apparently normal» origin (*) of the RCA from the anterior aortic rightward sinus of Valsalva, a common pitfall in echocardiographic screening of coronary arteries; b. RV dilation and D-shaped LV due to the leftward shift of the interventricular septum for the high pulmonary pressures of the newborn at arrival.

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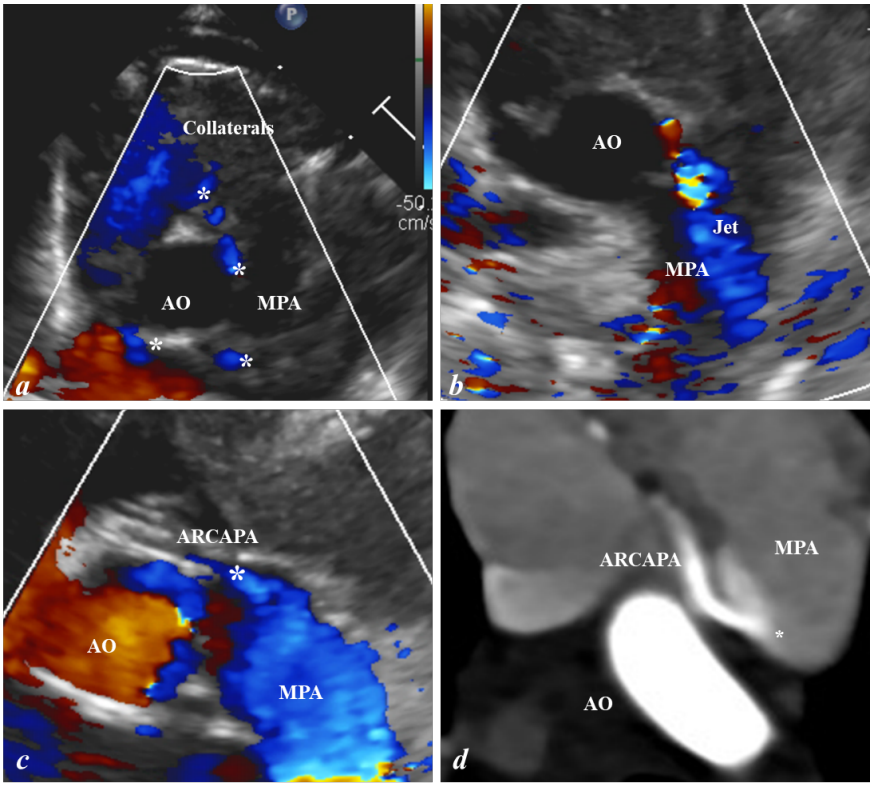

Fig 2. a. A network of small multiple coronary vessels (*) around aorta (AO), being the small initial collaterals between the left and right coronary artery; b. a systo-diastolic jet of flow into the MPA which represents the left-to-right shunt through the abnormal coronary vessel; c. the origin (*) of the RCA from the MPA (ARCAPA) with retrograde inverted «blue» flow inside, running anterior to the aorta. d. CT-scan image showing RCA arising (*) from MPA.

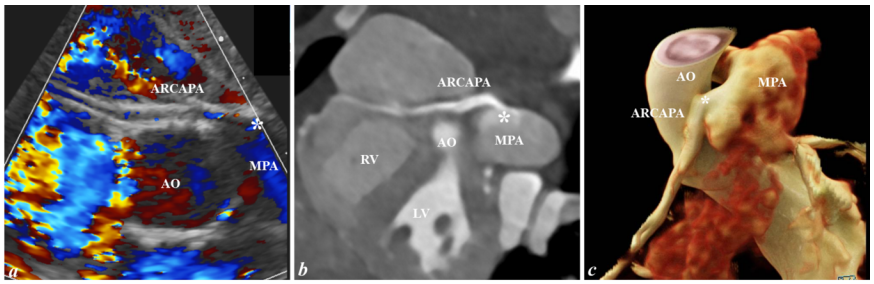

Fig 3. a, b. ARCAPA arising (*) from MPA in the echocardiographic parasternal short-axis view (a) and in the similarly oriented multiplanar reformat CT plane (b), running anterior to the aorta; c. 3D view from CT scan of the ARCAPA.

Bibliography

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