

Anomalous Right Coronary Artery Originating from the pulmonary artery. (ARCAPA): A systematic review of literature

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ABSTRACT

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a very rare congenital heart defect. Most patients are asymptomatic and the anomaly is detected incidentally during evaluation for other problems. Occasionally, ARCAPA may lead to myocardial ischemia and/or sudden cardiac arrest. Here we highlight the presentation, diagnosis and available treatment modalities for the management of this rare anomaly.

Introduction

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital coronary anomaly. The incidence of ARCAPA is reported to be 0.002% in the literature and represents 0.12% of coronary anomalies¹. Although more frequent in younger population, some patients have gone undiagnosed till the middle age where it could be diagnosed as an incidental finding. Compared to ARCAPA, Anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) is usually discovered early on in life since these patients are usually symptomatic². ARCAPA could exist by itself or in association with congenital heart diseases like tetralogy of fallot and bicuspid aortic valve³. The timeline for symptoms is variable in patients with ARCAPA and is related to the completeness of collateral circulation. Since a good number of patients remain asymptomatic the incidence may be higher compared to what has been reported⁴. Early symptoms may be secondary to heart failure, valvular insufficiencies or myocardial infarction⁵. Anomalous origin of coronary arteries typically from the contralateral sinus of valsalva (in particular the left coming off the right sinus) was more common compared to anomalous origin of right or left coronary arteries from the PA. These anomalies were more frequently associated with cause of sudden cardiac death in athletes in the USA⁶.

The pathophysiology of ARCAPA depends on the direction of blood flow in the coronary artery and its influence on oxygen delivery to the myocardium as well as the dominance. Patients with right dominant coronary circulation do not tolerate ARCAPA well compared with those with a left dominant system⁷. Coronary steal phenomenon can occur in ARCAPA due to differences in diastolic pressure between systemic and pulmonary arterial beds, with risk of myocardial ischemia during increased myocardial oxygen demand⁸. The presentation of ARCAPA is non-uniform and symptoms vary

from effort angina to dyspnea or fatigue; however, in adulthood, ARCAPA could sometimes manifest with atypical chest pain or other nonspecific instrumental signs, such as cardiac biomarker elevation suggestive for ischemic myocardial damage or arrhythmias (premature ventricular contractions to malignant ventricular tachycardia), both potentially responsible for sudden cardiac death. Rarely, in infancy, heart failure cases have been reported which after further evaluation have turned out positive for ARCAPA⁹. EKG in ARCAPA may be normal or it may show left ventricular hypertrophy or deep Q-waves in the inferior leads. Chest X-ray may vary from looking normal to showing signs of left ventricular hypertrophy.

Diagnostic Imaging

The first case of ARCAPA diagnosed by echocardiography was reported in 1885¹⁰. Nowadays, transthoracic echocardiogram (TTE) remains the common initial step to evaluate cardiac abnormalities. It can help delineate both coronary ostia, and Doppler can provide an anatomic definition of proximal intramural course of the anomalous vessel. Although helpful transthoracic echocardiogram (TTE) is not always straightforward in adult patients as it is easier to visualize coronary ostia in infants and young children compared to adults. Color flow Doppler demonstrating intracoronary collaterals within the ventricular septum was thought to be highly indicative of ARCAPA¹¹. Echocardiogram also shows dilated coronary arteries and a retrograde flow from the right coronary artery to the pulmonary artery suggestive of a normal phenomenon given the higher pressure in the RCA system thus creating a left to right shunt. However, in cases where TTE is difficult due to poor acoustic windows given the patient size, Transesophageal echocardiogram (TEE) could be an alternative first line diagnostic approach, providing more accurate details in suspicious of anomalous coronary takeoff and proximal course. Better visual confirmation can be obtained with coronary angiography, which most commonly shows multiple collaterals from the left anterior descending artery (LAD) and left circumflex artery (LCX) to the RCA eventually draining into the pulmonary artery. Oximetry can be performed to see the level of left to right shunting in these patients where typically there is a step up of oxygenation from the right ventricle to the pulmonary artery.

More advanced imaging modalities like cardiac MRI (CMR) and CT angiography (CTA) can confirm previous echocardiographic findings. CMR provides with blood flow measurements by phase-contrast MRI per minute and per body surface area. Qp/Qs, which indicates pulmonary to systemic blood flow ratio, can be calculated using CMR. Besides assisting in quantifying the flow difference in assessment of the shunt, it can also assist with evaluation of right ventricular size and function, stress perfusion defects

and possible myocardial fibrosis³. In centers without CMR, CTA can serve as a great alternative for diagnosis of ARCAPA. It additionally provides 3D volume rendering reconstructions for adequate images¹².

Management

Treatment should be considered for patients presenting with symptoms before 35 years of age. Since the risk of sudden death decreases with age, symptomatic relief is the main goal for treatment¹³. Therapeutically, different approaches have been suggested and tried. Surgical techniques including the simple ligation of the RCA with saphenous vein bypass grafting and then reimplantation of the RCA into the aorta (Takeuchi procedure) is occasionally performed in children. The alternative method practiced was direct implantation of the RCA into the ascending aorta if possible¹³. Another alternative, usually in children, is the creation of an aortopulmonary window and establishing flow between the anomalous right coronary ostium and aorta using an intrapulmonary baffle or tunnel¹⁴.

Reported complication of thrombosis has been of concern in implanted coronary arteries. There have been three reported cases with complications of thrombosis after surgical correction of ARCAPA. Where the exact mechanism is not quite known, competitive flow including high and low flow states have been thought to be the likely explanations behind the thrombosis. Traditionally for such complications warfarin has been tried however, direct oral anticoagulants have not yet been tested or approved in this subgroup of patients. Given the low incidence and infrequent detection of these cases, larger studies are difficult to study the side effect profile of these new medications.

Conclusion

In conclusion, awareness amongst clinicians is important to make an early diagnosis of this rare anomaly. High suspicion is warranted in patients with multiple color Doppler signals detected across interventricular septum on echocardiographic evaluation. Atresia of the ostium of the RCA is a close differential of the ARCPA and can be associated with a dilated left coronary artery⁹. Echocardiogram (TTE, TEE) and coronary angiography are the common traditional methods for ARCAPA diagnosis; however, in light of advancing technology and current availability of other imaging modalities, earlier use of such resources can help making the diagnosis. Surgery remains the standard mode of correction of the anomaly and the method of correction is usually based on the surgeon's preference and experience.

Conflict of interest disclosures

The authors declare that there is no conflict of interest regarding the publication of this paper.

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