Anomalous right coronary artery from the pulmonary artery

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The anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital heart disease, with an incidence of 0.002%. Often asymptomatic upon childhood, the diagnosis is easily missed, with the anomaly often diagnosed incidentally. Nevertheless, ARCAPA may lead to myocardial ischemia or sudden cardiac death, even in early childhood.

What is already known about the topic?
• The anomalous origin of the right coronary artery from the pulmonary artery is a rare congenital heart disease.
• ARCAPA may lead to myocardial ischemia or sudden cardiac death.
• Surgery is typically proposed upon diagnosis.

What does this article bring up for us?
This article reviews current knowledge about ARCAPA: definition, pathophysiology, recommended diagnostic procedures, and recommended therapies. In conclusion, a surgical correction is recommended even in asymptomatic patients, on the basis of a favorable risk-benefit ratio.

INTRODUCTION
The anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital heart disease (incidence of 0.002%) (1,4). The physiopathology of ARCAPA is explained by retrograde blood flow in the RCA and the risk of myocardial ischemia. After birth, an inadequate collateralization can lead to death because myocardial perfusion is ensured by collateralization from the left coronary artery to the RCA, with a steal phenomenon to the pulmonary artery (1). Often asymptomatic in childhood (5), the diagnosis is easily missed and the anomaly can be incidentally diagnosed (6). However, ARCAPA may lead to myocardial ischemia and/or sudden cardiac death even in early childhood (5,7).
We report the case of a healthy 6 year-old boy with an isolated ARCAPA, revealed in the setting of a heart murmur. A first echocardiogram suggested the diagnosis of coronary fistula or aberrant coronary artery with a preserved systolic biventricular function. The patient’s clinical history, laboratory testings electrocardiogram and chest X-ray were otherwise unremarkable and the patient was referred to our center for further evaluation.

After a second color Doppler echocardiography (Figure 1), a cardiac catheterization was performed. Selective coronary angiograms showed a dilated left coronary system and a RCA arising from the main pulmonary artery and an extensive collateral circulation between the two coronary arteries with retrograde filling of the RCA from the left system (Figure 2). This confirmed the abnormal implantation of the RCA into the pulmonary artery.

A re-implantation of the right coronary artery into the ascending aorta was performed in order to restore a normal dual coronary system (Figure 3). The procedure was uneventful and the patient was discharged on postoperative day 5.

An echocardiogram obtained at 1 year of follow-up demonstrated an unobstructed anterograde flow into the RCA.

Figure 1. Doppler echocardiogram suggesting the abnormal RCA anatomy

(A) Color Doppler suggesting retrograde blood flow in the RCA (in blue).
(B) Image suggesting a coronary fistula or aberrant coronary artery.

Figure 2. Selective coronary angiograms

Selective angiography of the left coronary artery arising from the ascending aorta in the anteroposterior (A and B2) and lateral (B1) projections.

In the latest phase of injection, a retrograde flow of contrast is seen through multiple collaterals arteries from the left coronary artery to the right coronary artery, which in turn drains into the pulmonary artery, confirming the diagnosis of ARCAPA.

A = early phase AP
B1 + B2 = late phase AP
Anomalous right coronary artery from the pulmonary artery arising from the pulmonary artery instead of the right aortic sinus of Valsalva (1,7).

The physiopathology of ARCAPA is explained by the progressive development of a retrograde blood flow in the RCA when the pulmonary vascular resistance decreases after birth with a subsequent risk of myocardial ischemia (8).

In utero, the anomalous RCA arising from the pulmonary artery is perfused adequately because of the prenatal high pulmonary vascular resistance (1,3).

Shortly after birth, the pulmonary vascular resistance drops to significantly lower levels, and a significant collateralization between the right and left coronary arteries becomes mandatory to ensure sufficient myocardial perfusion (1,3). The ventricular ischemia in ARCAPA is less frequent than in patients with anomalous left coronary artery from the pulmonary artery (ALCAPA) because the right ventricle demands less oxygen than

DISCUSSION

Congenital anomalies of the coronary arteries are rare (< 1% of the pediatric population). Some of these abnormalities are benign but others are associated with a risk of myocardial ischemia or sudden death. Among these congenital anomalies of the coronary arteries, there are:

- the anomalies of the origin of the coronary arteries;
- the anomalies of the course of the coronary arteries;
- the distal connection abnormalities mainly represented by coronary fistulas;
- the single coronary ostia (right or left) or the coronal duplication.

Isolated (70%) or associated with other heart defects (30%) like Tetralogy of Fallot or aorto-pulmonary window, ARCAPA is described as the abnormal origin of the RCA

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(A) The pulmonary artery was opened and the right coronary ostium was identified.
(B) The right coronary ostium was detached from the pulmonary artery.
(C) The ascending aorta is opened: a hole is created and the right coronary ostium from the pulmonary artery is reimplanted.
(D) Right coronary artery after reimplantation on the aorta.
* Right coronary ostium ** Ascending aorta

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ARCAPA, a rare congenital anomaly, is often asymptomatic in childhood and incidentally diagnosed. It’s associated with a risk of myocardial ischemia or sudden death. First-line investigation for the diagnosis is echocardiography. Other diagnostic modalities are: angiography, cardio-CT and MRI. Surgical correction is recommended even in asymptomatic patients based on a favorable risk-benefit ratio.

**PRACTICAL RECOMMENDATIONS**

- **TAKE HOME MESSAGE**

  - ARCAPA is often asymptomatic. Some patients present heart murmur, chest pain, palpitations, dyspnea or rarely heart failure, arrhythmias, myocardial infarction or sudden death.
  - Risk of myocardial ischemia and sudden death.
  - First-line investigation for the diagnosis of ARCAPA is echocardiography. Other diagnostic modalities are: angiography, cardio-CT and MRI.
  - Surgical correction is recommended even in asymptomatic patients based on a favorable risk-benefit ratio.


Conflict of interest
No potential conflict of interest.

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