



## Kasuistika | Case report

# Anomalous origin of right coronary artery from pulmonary artery with aneurysmal coronary arteries

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## SOUHRN

Popisujeme případ 16letého asymptomatického chlapce, odeslaného na naše pracoviště pro vyšetření srdečního šelestu. Echokardiografické i angiografické vyšetření prokázalo anomálii v odstupu pravé koronární tepny z plicnice spolu s dilatací levé koronární tepny a několika velkými kolaterálami. Cílem chirurgického řešení je vytvoření fyziologického koronárního oběhu.

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## ABSTRACT

We describe a case of 16-year-old asymptomatic boy that was referred for evaluation of a heart murmur. Echocardiography and angiography showed an anomalous right coronary artery from the pulmonary artery with dilatation of left coronary artery and several large collateral vessels. The goal of surgical therapy is establishment of a physiologic bi-coronary circulation.

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Fig. 1 – Preoperative diagnostic assessment. Selective coronary angiographic study shows the anomalous origin of the right coronary artery from pulmonary artery (\*); note the large dilated and tortuous course of LCA. The RCA is later contrasted through several large collateral vessels originating from LCA (A). Bidimensional echocardiography (short axis view) shows the aneurysm of the left coronary ostium (\*) (B) and color flow Doppler analysis confirms inversion of coronary blood flow (\*) (C).

## Introduction

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital cardiac lesion with an incidence of 0.002% [1]. The patient with ARCAPA is almost always asymptomatic. Symptomatic presentation could be with angina, arrhythmias, myocardial infarction, heart failure, sudden death or due to associated cardiac anomalies [1,2]. On occasion, an unrelated heart murmur may lead to the diagnosis being made by imaging techniques. The diagnosis is still made by echocardiography and angiography as well as CT scan. Recently MRI with stress perfusion could also provide both structural and functional information [3].

lective coronary angiography showed a dominant left coronary artery with a large dilated and tortuous course with several large collateral vessels, that later contrasted the right coronary artery.

The right coronary artery itself was dilated and drained into the pulmonary artery. The patient was scheduled for elective surgical repair in order to establish a two coronary artery system, which was successfully achieved with right coronary artery reimplantation on the ascending aorta (Fig. 2A). Postoperative transthoracic echocardiography shows the re-implanted right coronary artery arising from the aorta (Fig. 2B). The patient was discharged home on 8 post operative days with antiplatelet drug (aspirin) to avoid clot formation in aneurysmal coronary artery [1].

## Case report

A 16-year-old asymptomatic boy, with no family history of coronary artery or cardiovascular disease, was referred for evaluation because during a medical visit a continuous heart murmur at the left upper sternal border was incidentally discovered.

Transthoracic echocardiography was performed, showing a right coronary artery arising from the pulmonary artery with evident flow inversion (Fig. 1A), and an aneurysmal left coronary artery arising from ascending aorta, with a left coronary ostium diameter of 6–7 mm (Fig. 1B and 1C), with normal bi-ventricular function. A se-

## Discussion

The development of the coronary arterial system during Carnegie stage 18 is an essential event in heart formation. The arterial trunk septation (Carnegie stage 16) precedes the appearance of the coronary ostia and therefore cannot be responsible of their final position. Normally, two major coronary arteries arise from the aorta, with their ostia positioned at the left and right aortic valve sinuses. Coronary endothelial cells invade the myocardium from a variety of sources, including the pro-epicardium, endocardium and sinus venosus. An ingrowth of endothelial strands towards the aorta form a ring of capillaries

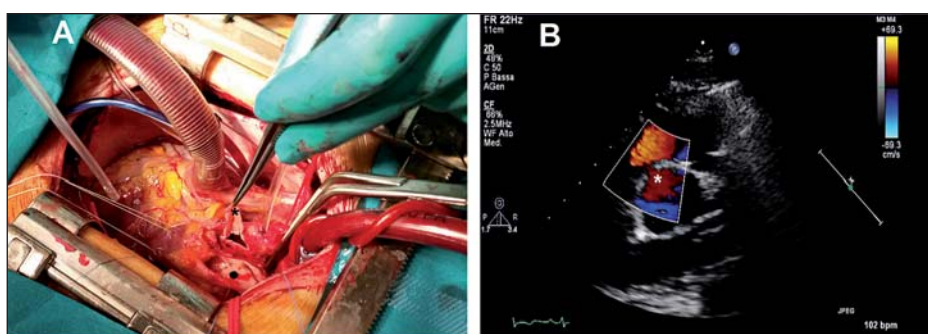


Fig. 2 – Intraoperative surgical view: reimplantation of the right coronary button (\*) from the pulmonary artery (circle) on the aorta (A). Postoperative echocardiography study shows the usual coronary blood flow in the reimplanted right coronary artery from aorta (B).

encircling the outflow tract (peritruncal plexus) and penetrate the aortic sinuses at multiple sites and ultimately remodel at only two Valsalva sinus, forming the coronary arteries buds. The molecular pathways involved in ostia formation are complex and orchestrated by vascular epidermal growth factor (VEGF), fibroblast growth factor 2 (FGF-2), platelet-derived growth factor  $\beta$  (PDGF $\beta$ ), chemokine CXCL12 and its receptor CXCR4 [1]. In particular, chemokine activity is fundamental in leading/driving endothelial cells migration to the aortic sinuses. For this reason, an alteration in chemokine activity could be considered as one of the feasible heuristic mechanisms of anomalous origin of coronary (e.g. the origin of the right coronary artery from the pulmonary artery).

Surgical correction of ARCAPA is recommended to avoid cardiac symptoms and sudden death. In a series of 70 patient with ARCAPA, 4 cases of sudden death were reported (5.7%) and four patients died as a result of severe chronic heart failure (5.7%) [2]. The onset of symptoms and severity depends on the type of dominant coronary circulation (those with right dominant system do not tolerate ARCAPA as well as those with a left system) and the grade of collateralization between right coronary artery and left coronary artery. The correction could be performed from neonatal [3–5] period to adulthood.

The repair consists of ARCAPA translocation and reimplantation to the aorta. Other technique could be coronary artery bypass graft or Takeuchi procedure (intrapulmonary tunnel), when the RCA is arising at the extreme left lateral position on the main pulmonary artery [6].

The evolution of aneurysmal coronary after surgical correction is still under investigation. Van-Meurs-van-Woezik et al. show reduction in the size of coronary artery 3 years after reimplantation of ARCAPA [7]. We suggest aspirin therapy and long-term follow-up with echocardiographic evaluation until the coronary artery returns to the normal size.

#### Conflict of interest

None.

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#### Ethical statement

I declare, on behalf of all authors that the research was conducted according to Declaration of Helsinki.

#### Informed consent

I declare, on behalf of all authors that informed consent was obtained from all patients participating in this study.

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