Congenital coronary aneurysm and cameral fistula embolization in a teenager

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Early publication date: September 1, 2023 Coronary interventions in children are rare and mostly caused by congenital acquired anomalies, including abnormal vessel origin from the pulmonary trunk, coronary and cameral fistulas, and vasculitis (e.g., Kawasaki and Takayasu diseases or systemic lupus erythematosus with aneurysm development) [1–3] or cardiac surgeries with coronary transplantation.

A 17-year-old girl was referred for cardiac evaluation due to a heart murmur found on auscultation in a pediatric office. In medical history, the patient was asymptomatic without symptoms of chest pain or easy fatigability. A diagnosis of right coronary artery (RCA) aneurysm with a cameral fistula was established in a cardiology department on transthoracic echocardiography (TTE), computed tomography, and coronary angiography, with a negative treadmill exercise stress test. The patient was referred for heart surgery. On admission to the cardiac surgery center, the vital signs were normal with a regular heart rate of 72 beats per minute and blood pressure of 100/60 mm Hg. Myocardial biomarkers (NT-proBNP and troponin) were within the normal range. An electrocardiogram (ECG) showed normal sinus rhythm without features of ventricular hypertrophy or myocardial ischemia. TTE revealed normal myocardial contractility, dilated proximal RCA (6 mm) with turbulent flow over the right ventricular (RV) wall. The child was qualified for initial interventional RCA aneurysm embolization and in case of failure, surgery was an option.

Aortic root angiography showed proximal RCA dilation, critical pre-aneurysmal stenosis (1 mm), large right coronary aneurysm $(10 \times 7 \text{ mm})$ with a cameral fistula stealing the blood into the RV (Figure 1A–C, Supplementary materials, *Videos S1–S3*). The aneurysm continued into the distal RCA supplied abundantly from the circumflex artery (Cx) collateral circulation. A balloon occlusion test of the RCA aneurysm with a 4 mm Tyshak balloon catheter was performed with simultaneous ECG evaluation. It showed normal ECG tracings indicating sufficient Cx collateral circulation.

An arterio-venous wire loop was established with a multipurpose catheter over a 0.014-inch guidewire and an Amplatz 6 mm Goose NeckTM snare system (ev3, Plymouth, MN, US) by crossing the aorta, right coronary aneurysm, cameral fistula, RV and inferior vena cava (Supplementary materials, *Videos S4–S7*). Unfortunately, access to the aneurysm with a 4 F multipurpose catheter *via* cameral fistula was inapplicable due to the small size of the fistula (less than 2 mm).

Finally, the approach through the aortic root and, critically, RCA stenosis was established with a 2.9 F catheter. The aneurysm was successfully embolized with neurological detachable Penumbra Coil 400 system and PAC coils (Penumbra, Alameda, CA, US) (Figure 1D–F; Supplementary materials, *Videos S8–S11*). The clinical course was uneventful with normal ECG and myocardial contractility on TTE. The troponin level was transiently elevated up to 90 ng/l (normal range <26.2 ng/l) with normalization within 3 days.

In 1 year follow up the girl was in good condition with normal TTE and magnetic resonance imaging (LVEF 60%, RVEF 59%) without features of myocardial ischemia.

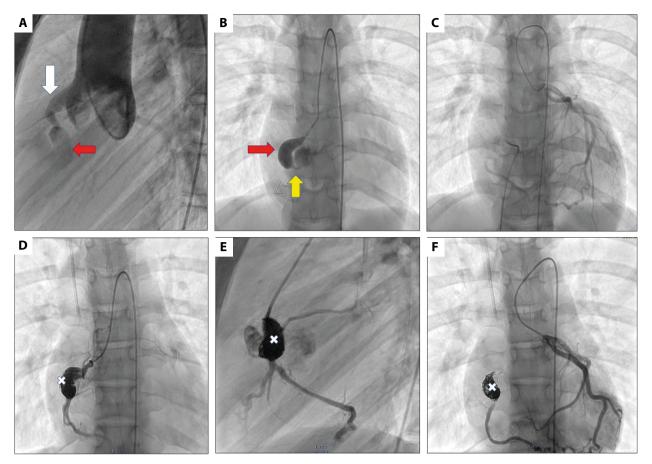


Figure 1. A. Aortography in the lateral view showing right coronary proximal dilation (white arrow), pre-aneurysmal stenosis, and a large coronary aneurysm (red arrow). B. Selective right coronary angiography (anteroposterior view) showing a large aneurysm (red arrow) and a cameral fistula (yellow arrow) draining the blood into the right ventricle. C. Selective left coronary angiography (antero-posterior view) showing collateral circulation with the right coronary artery. D and E. Selective right coronary aneurysm embolized with Penumbra coils (white cross). F. Selective left coronary angiography (anteroposterior view) showing collateral circulation without steal phenomenon *via* the embolized cameral fistula

In conclusion, we would like to underline that percutaneous or hybrid coronary interventions have become alternative options in children with coronary abnormalities [4]. The dilemma of whether to embolize an RCA in the case of a life-threatening aneurysm may be resolved with a balloon occlusion test and left coronary angiography showing sufficient collateral circulation [5].

Supplementary material

Supplementary material is available at https://journals. viamedica.pl/kardiologia_polska.

Article information

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