An asymptomatic 16-year-old girl with anomalous left coronary artery from pulmonary artery (Bland-White-Garland syndrome)

Pacjentka 16-letnia z bezobjawowym nieprawidłowym odejściem lewej tętnicy wieńcowej od pnia płucnego (zespół Blanda-White'a-Garlanda)

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Abstract

A 16-year-old girl was referred to our hospital to perform cardiac multi-slice computed tomography (MSCT) due to an unclear view of coronary arteries in echocardiography performed during a routine visit to the cardiology clinic. As an infant she was hospitalized due to signs of severe heart failure, and while in hospital she underwent cardiac arrest. However, ever since this incident she has not experienced any cardiovascular symptoms, and still remains asymptomatic. On physical examination we found systolic murmur localized to the left sternal border. In echocardiography the dimensions of cardiac chambers and ventricular contractility were normal. Cardiac MSCT demonstrated markedly dilated tortuous right coronary artery (RCA), giving very well-developed collaterals crossing the interventricular septum toward the left coronary artery (LCA). The LCA was also dilated but not as much as the RCA. The images showed an anomalous origin of the LCA from the pulmonary artery trunk (ALCAPA) and suggested retrograde filling of the LCA through collaterals from the RCA and emptying into the pulmonary artery trunk (PA). Coronary angiography and aortography confirmed the diagnosis of ALCAPA (Bland-White-Garland syndrome). It showed a very big tortuous RCA, the LCA filling retrogradely through collaterals from the RCA and draining into the PA. Anomalous left coronary artery from pulmonary artery is a congenital anomaly associated with early infant mortality, very rarely diagnosed in asymptomatic adults, with uncertain but probably poor prognosis. MSCT is useful for diagnosis. The patient still remains asymptomatic and so far she and her parents have not agreed to surgical treatment.

Key words: anomalous left coronary artery from pulmonary artery, Bland-White-Garland syndrome, cardiac multi-slice computed tomography

Streszczenie

Dziewczynka 16-letnia została skierowana do szpitala w celu wykonania wielorzędowej tomografii komputerowej (*multi-slice computed tomography* – MSCT) serca z powodu niejasnego obrazu tętnic wieńcowych w echokardiografii wykonanej podczas wizyty kontrolnej w poradni kardiologicznej. Jako niemowlę była ona hospitalizowana z powodu objawów ostrej niewydolności serca, przebyła wtedy zatrzymanie krążenia. Później rozwijała się dobrze, nie zgłasza dotąd żadnych dolegliwości ze strony układu krążenia. W badaniu przedmiotowym stwierdzono szmer skurczowy wzdłuż lewego brzegu mostka. W badaniu echokardiograficznym wymiary jam serca i kurczliwość mięśnia lewej komory były prawidłowe. W badaniu metodą MSCT uwidoczniono znacznie poszerzoną, krętą prawą tętnicę wieńcową (*right coronary artery* – RCA), dającą bardzo dobrze rozwinięte krążenie oboczne przez przegrodę międzykomorową do lewej tętnicy wieńcowej (*left coronary artery* – LCA). Lewa tętnica wieńcowa była także poszerzona, lecz w mniejszym stopniu niż RCA. Uwidoczniono nieprawidłowe odejście LCA od pnia płucnego (ALCAPA), obraz sugerował wsteczne wypełnianie się LCA przez kolaterale od RCA i opróżnianie LCA do pnia płucnego (*pulmonary artery* – PA). Koronarografia i aortografia potwierdziły ALCAPA (zespół Blanda-White'a-Garlanda). Uwidoczniono bardzo dużą, krętą RCA oraz LCA wypełniającą się wstecznie przez kolaterale od RCA i opróżniającą się do PA. Zespół Blanda-White'a-Garlanda jest wrodzoną patologią związaną z dużą śmiertelnością we wczesnym okresie niemowlęcym, bardzo rzadko diagnozowaną u dorosłych bez objawów, z niepewnym, a prawdopodobnie złym rokowaniem. Badanie metodą MSCT jest przydatnym narzędziem diagnostycznym w takich przypadkach. U pacjentki nie występują żadne objawy choroby; wraz z rodzicami nie zgadza się ona na leczenie operacyjne.

Słowa kluczowe: nieprawidłowe odejście lewej tętnicy wieńcowej od pnia płucnego, zespół Blanda-White'a-Garlanda, MSCT tętnic wieńcowych

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Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (anomalous left coronary artery from pulmonary artery [ALCAPA] or Bland-White-Garland syndrome) is a rare congenital disease. It is present in 1 out of 300 000 live-born children and constitutes 0.24% to 0.46% of congenital heart diseases [1-3]. Untreated disease leads to death in over 90% of children before the first year of age. Adult patients with this disease are very rarely found and asymptomatic patients are completely unique [3]. For the last 10 years diagnoses of ALCAPA in adult patients have been made with the use of multi-slice computed tomography (MSCT) [3, 4].

Case report

We present a case of a 16-year-old girl referred to our hospital from the Paediatric Cardiology Outpatient Clinic for computed tomography of the heart. The reason for referral was the unclear picture of coronary arteries found on echocardiography (large proximal segment of the right coronary artery with a turbulent flow) and multiform echoes – turbulences in the interventricular septum. Echocardiographic examination was performed routinely during the patient's control visit at the Cardiology Outpatient Clinic. The patient was hospitalized between the 6th and 9th month of age because of severe heart failure with cardiogenic shock and cardiac arrest. The hospitalization lasted 3 months and ended with a diagnosis of myocarditis. The patient's growth has been normal since then, she has not required any further hospitalizations and has been free from pharmacological treatment since 18th months of age. Currently the patient is free from symptoms with good exercise tolerance. Physical examination shows the presence of a systolic murmur along the left sternal margin. Echocardiographic examination demonstrated (apart from the features described above) normal dimensions of the heart chambers, normal left ventricular contractility and mild mitral valve regurgitation. Multi-slice computed tomography of the coronary arteries was performed. The right coronary artery (RCA) had a typical origin from the right coronary aortic sinus, but with an atypical morphology – it was very wide (9 mm of diameter in the proximal and middle segment) and tortuous (Figure 1). The left coronary artery (LCA) does not originate from the aorta, but from the pulmonary artery (PA). There was a highly developed collateral circulation between the RCA and LCA through the interventricular septum. There were also signs of retrograde filling of the LCA from the RCA with the LCA emptying into the pulmonary trunk (Figure 2). Because of the findings described above the patient was qualified for an invasive examination including coronary angiography, aortography and right heart catheterization. The anomalous origin of the LCA from the PA (Bland-White-Garland syndrome, ALCAPA) was confirmed. The very wide and tortuous RCA provides highly developed collateral circulation to the LCA, which is also wide, but less so than the RCA (Figures 3, 4). The LCA is filled in a completely retrograde manner from the RCA and empties into the PA (retrograde flow). The patient was consulted surgically, but did not agree (she and her parents) to surgical treatment. Exercise test did not show signs of myocardial ischemia with a load of 12 METs.

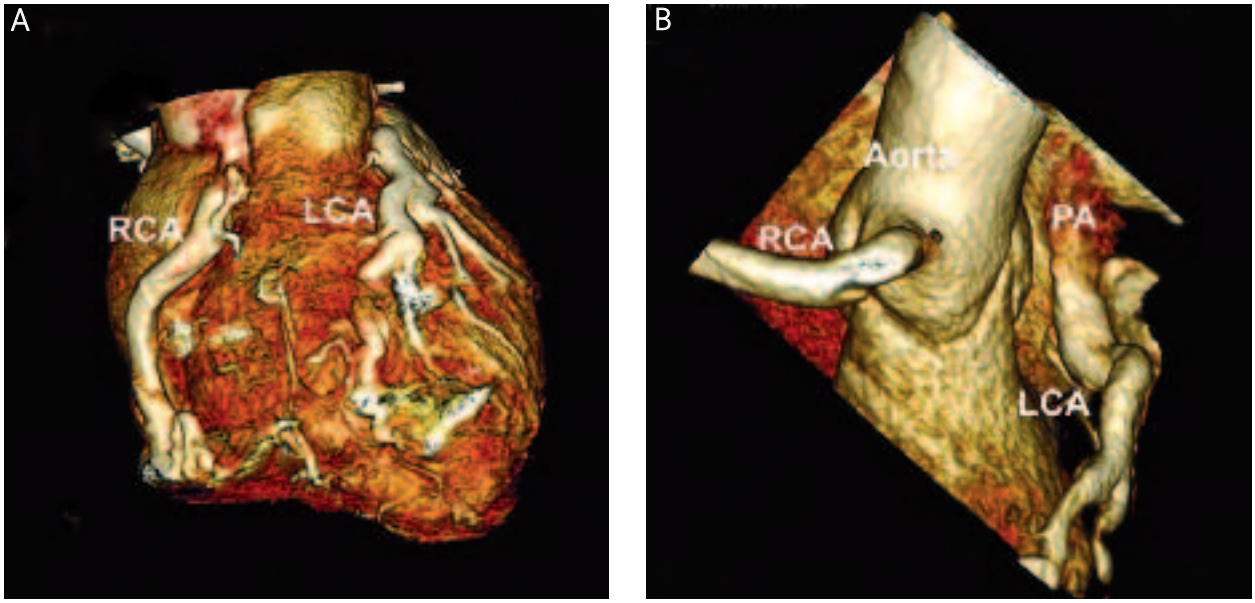

Fig. 1. Coronary arteries in MSCT (volume-rendering technique – VRT): origin of the right coronary artery (RCA) from the right coronary sinus of the aorta (**A**) and the left coronary artery (LCA) from the pulmonary artery trunk (PA) (**B**)
Ryc. 1. *Obraz tętnic wieńcowych w MSCT (rekonstrukcja objętościowa): odejście prawej tętnicy wieńcowej (RCA) od prawej zatoki wieńcowej aorty (**A**) i lewej tętnicy wieńcowej (LCA) od pnia płucnego (PA) (**B**)*

Discussion

In 85% of cases the symptoms of Bland-White-Garland syndrome occur in the first two months of life [3, 5]. Similar pressure in the aorta and pulmonary trunk existing during fetal life enables a similar blood flow in the RCA arising from the aorta and the LCA originating from the PA. After birth there is a decrease in blood pressure and blood oxygenation in the PA and the LCA, which causes myocardial ischemia of the segments supplied by the LCA. The situation is worsened by the closure of the ductus arteriosus, which occurs up to 3 months of age, and at that time the infants usually present with symptoms of severe heart failure caused by ischemia, and if they are left untreated they usually die.

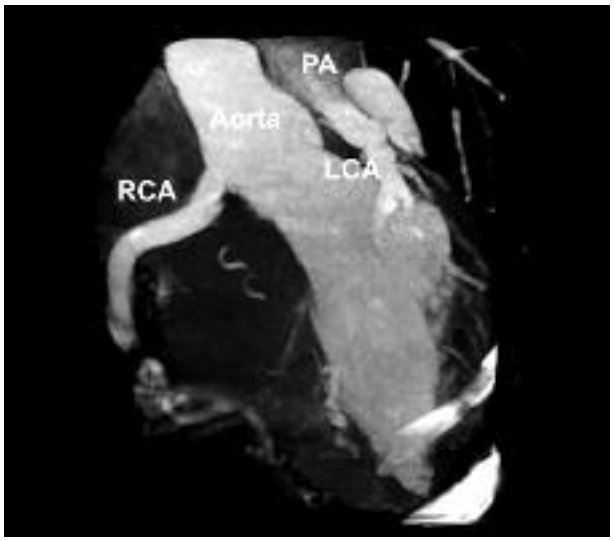

Fig. 2. Coronary arteries in MSCT (multiplanar reformation – MPR): origin of the right coronary artery (RCA) from the aorta and the left coronary artery (LCA) from the pulmonary artery trunk (PA), emptying the LCA (jet) into the PA
Ryc. 2. *Obraz tętnic wieńcowych w MSCT (projekcja wielopłaszczyznowa): odejście prawej tętnicy wieńcowej (RCA) od aorty i lewej tętnicy wieńcowej (LCA) od pnia płucnego (PA), widoczne opróżnianie LCA („jet") do PA*

Survival is possible only after development of potent collateral circulation between the RCA and the LCA with reversal of blood flow in the LCA (in the direction of the PA) [3-5]. Such a situation is present in adult patients with Bland-White-Garland syndrome described in the literature: a large, widened and tortuous RCA providing many collaterals to the LCA with reverse flow of blood in the LCA into the PA. This picture was present in the described case in MSCT and coronary angiography. Typical echocardiographic features of this disease consist of widened RCA, reverse flow in the LCA to the PA and highly developed flow in the collaterals present in the interventricular septum [6]. This picture was partially present in our patient.

Prognosis in adult patients with ALCAPA is uncertain or even unfavourable [3-5]. The asymptomatic period is often followed by the onset of heart failure or sudden cardiac death [5, 6]. The authors of a systematic review of data published in 2011 analysed 152 cases of adult patients with ALCAPA and found only 14% of patients who were completely asymptomatic [3]. Other patients presented symptoms of heart failure or angina, heart palpitations and syncope. In most of the patients physical examination

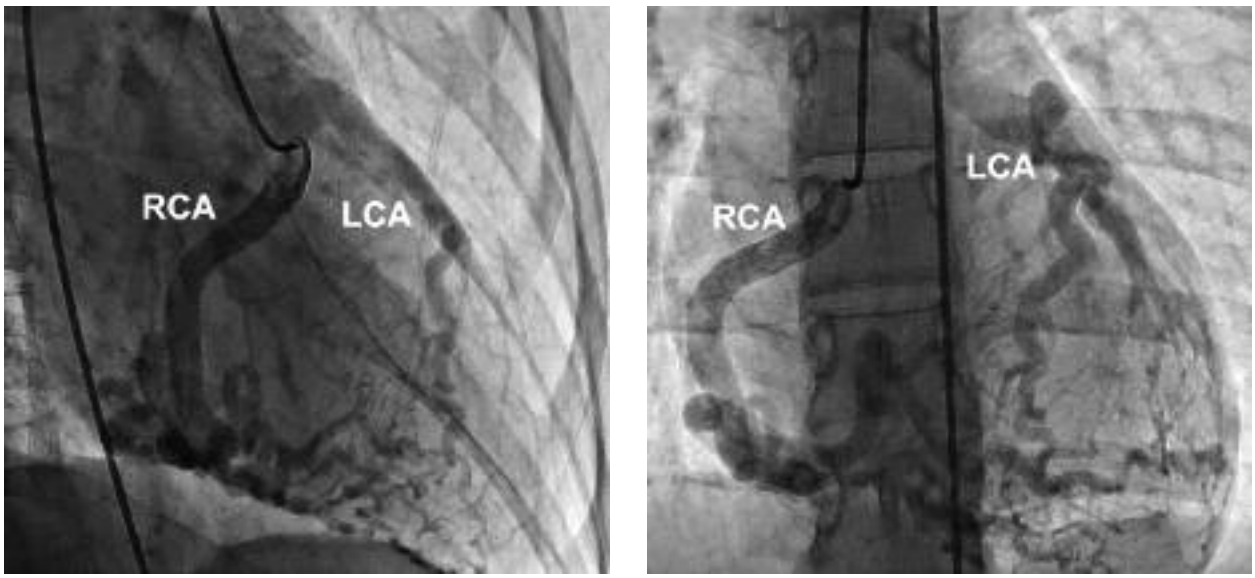

Fig. 3. Coronary angiography of the right coronary artery (RCA): it shows retrograde filling of the left coronary artery (LCA) through collaterals from the RCA and drainage of the LCA into the PA; RAO 30 (**A**) and AP projection (**B**)
Ryc. 3. *Angiografia prawej tętnicy wieńcowej (RCA): widoczne wsteczne wypełnianie się lewej tętnicy wieńcowej (LCA) przez krążenie oboczne od RCA i opróżnianie LCA do pnia płucnego; projekcje RAO 30 (**A**) i AP (**B**)*

demonstrated the presence of a systolic murmur along the left sternal margin (hyperkinetic flow of blood in the functional fistula formed by the connection between the systemic and pulmonary circulation through the collaterals between the RCA and LCA), as observed in our patient.

A particularly important issue is the risk of sudden cardiac death. This type of incident occurred in 7% of patients in the presented review. However, the frequency was higher in younger patients (< 50 years of age) (9%) when compared to patients > 50 years of age (2%). Mean age of patients who died suddenly was 31 years and most of them were completely asymptomatic before that event [3].

These observations lead to the conclusion that all adults with ALCAPA should be qualified for surgical treatment [3-5]. The diagnosis is made at various age of the patients by means of echocardiography (infants), coronary angiography or, more frequently, MSCT. Surgical treatment also depends on the age at which the disease is diagnosed. In infants the LCA is most frequently transplanted from the PA to the aorta, while adults are more frequently treated with bypass to the LCA with ligation at its origin from the PA or with Takeuchi surgery involving the formation of a tunnel through the pulmonary trunk linking the LCA and the aorta [5, 7, 8].

In conclusion, it should be noted for cardiological practice that Bland-White-Garland syndrome might also be present in adults. MSCT examination is an excellent diagnostic tool for this heart defect.

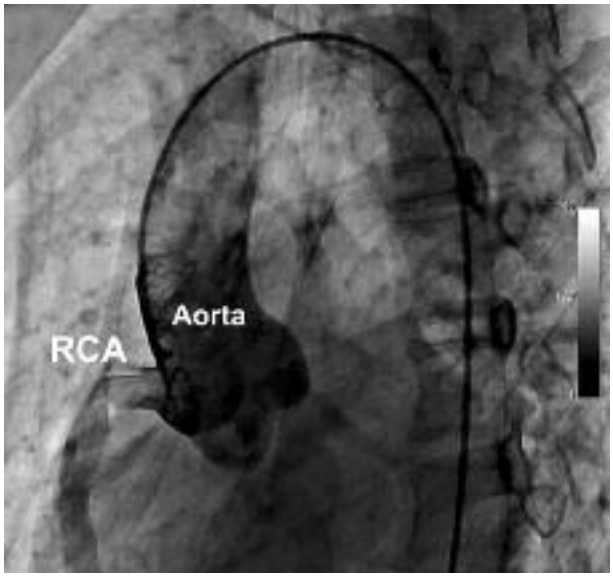

Fig. 4. Aortography in LAO 60 projection: it shows the right coronary artery (RCA) and lack of the left coronary artery

Ryc. 4. *Aortografia w projekcji LAO 60: widoczne kontrastowanie prawej tętnicy wieńcowej (RCA) i brak lewej tętnicy wieńcowej w miejscu typowym*

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