

УДК 616.132-007.271-007.21-06:616.12-008.331.1]-055.2-053.81

<https://doi.org/10.20538/1682-0363-2021-4-205-210>

## A clinical case of secondary hypertension in a young woman with coarctation of the aorta

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### ABSTRACT

The article presents a case of diagnosis and treatment of coarctation of the aorta in a 20-year-old woman, who previously received follow-up care with the diagnosis of hypertension. This case demonstrates the importance of qualitative examination of young patients with hypertension, including tonometry in the lower extremities and transthoracic echocardiography. The peculiarities of this clinical case encompass a rarer, isolated type of coarctation of the aorta and high physical fitness of the patient, which reduced doctors' alertness regarding this anomaly. After surgical correction, a significant decrease in the blood pressure was achieved; however, such patients need long-term follow-up in order to detect complications, such as aneurysms, restenosis, or residual stenosis.

**Key words:** coarctation of the aorta, secondary hypertension, aortic diseases.

**Conflict of interest.** The authors declare the absence of obvious or potential conflicts of interest related to the publication of this article.

**Source of financing.** The authors state that they received no funding for the study.

**For citation:** Manukyan M.A., Falkovskaya A.Yu., Mordovin V.F., Saushkin V.V., Ryabova T.R. A clinical case of secondary hypertension in a young woman with coarctation of the aorta. *Bulletin of Siberian Medicine*. 2021; 20 (4): 205–210. <https://doi.org/10.20538/1682-0363-2021-4-205-210>.

## Клинический случай симптоматической артериальной гипертензии у молодой женщины с коарктацией аорты

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### РЕЗЮМЕ

Представлен случай диагностики и лечения коарктации аорты у женщины в возрасте 20 лет, ранее наблюдавшейся с диагнозом «артериальная гипертензия» (АГ). Приведенный клинический пример демонстрирует важность качественного обследования молодых больных с АГ, включающего тонометрию на нижних конечностях и трансторакальную эхокардиографию. Особенности данного клинического случая являются более редкий, изолированный тип коарктации аорты и высокая физическая тренированность пациентки, что снижало настороженность врачей в отношении этой аномалии. После хирургической

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коррекции порока достигнуто существенное снижение артериального давления, однако в дальнейшем такие пациенты нуждаются в длительном наблюдении с целью возможного выявления таких осложнений, как аневризмы, рестенозы и резидуальные стенозы.

**Ключевые слова:** коарктация аорты, вторичная артериальная гипертензия, заболевания аорты.

**Конфликт интересов.** Авторы декларируют отсутствие явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

**Источник финансирования.** Авторы заявляют об отсутствии финансирования при проведении исследования.

**Для цитирования:** Манукян М.А., Фальковская А.Ю., Мордовин В.Ф., Саушкин В.В., Рябова Т.Р. Клинический случай симптоматической артериальной гипертензии у молодой женщины с коарктацией аорты. *Бюллетень сибирской медицины*. 2021; 20 (4): 205–210. <https://doi.org/10.20538/1682-0363-2021-4-205-210>.

## INTRODUCTION

Coarctation of the aorta (CoA) is a relatively rare anomaly, accounting for 5–8% of all congenital heart defects. Its prevalence is 2.5–4 cases per 10,000 infants, with predominance in males (2:1 ratio) [1]. Despite the traditional division into preductal (infantile type) and postductal (adult type) depending on the location with respect to the arterial duct, congenital CoA is essentially periductal, and the age of its manifestation depends on the degree of aortic stenosis and the association with other defects [2]. There is currently no consensus on the etiopathogenesis of CoA; the most common theories are the abnormal genetic mutation theory [3], the flow theory (A. Rudolf, 1972), and the loop theory (J. Skoda, 1855). It should be recognized that in most cases (up to 83%), CoA is combined with various congenital anomalies, most commonly bicuspid aortic valve (up to 60%) and hereditary diseases, such as Turner syndrome and other anomalies [3]. In the past, CoA was regarded as a local anatomical narrowing, but is now considered to be a diffuse aortopathy [4].

The hemodynamic essence of CoA involves reduced blood supply to the lower body. At this time, renal hypoperfusion activates the renin – angiotensin – aldosterone system, which leads to persistent arterial hypertension (AH) of the upper body, development of left ventricular (LV) hypertrophy, pre- and post-stenotic aortic dilatation, and formation of collateral circulation due to anastomosed intercostal, internal mammary, and scapular arteries [5].

Proximal hypertension increases the risk of aneurysms in the cerebral arteries [6] and thoracic aorta, as well as early coronary artery disease. Depending on the degree of aortic constriction and compensatory capacity, the clinical presentation can range from

critically severe heart failure in infants to asymptomatic hypertension in adults [7]. The main manifestations of CoA are upper-limb systolic hypertension and lower-limb hypotension, with the arterial pressure (AP) gradient between the upper and lower extremities > 20 mmHg. In the natural course of the disease, the average life expectancy of patients, according to M. Campbell, is about 34 years, and the leading causes of death according to 304 autopsies were congestive heart failure (25.5%), aortic rupture (21%), bacterial endocarditis (18%), and intracranial hemorrhage (11.5%) [8]. In the meantime, surgical correction of the defect can dramatically change the fate of patients [9–11].

As CoA is mostly well diagnosed in childhood, it is one of the rare causes of arterial hypertension in adults, in whom its distinguishing feature is resistance to pharmacotherapy. Transthoracic echocardiography verifies the diagnosis of CoA, while computed tomography (CT) or magnetic resonance imaging (MRI) are useful tools in selecting an individual therapy, detecting complications, and managing the defect.

## CLINICAL CASE

Patient R., 20 years old, born in 1999, was admitted to the Department of Arterial Hypertension of the Cardiology Research Institute, Tomsk NRMC on April 13, 2020 with the following complaints: 1) stable elevation of AP up to 160–170 / 80–90 mmHg, with episodes of elevation up to 220 / 120 mmHg, accompanied by intense headache in the occipital region, dizziness, and generalized weakness; 2) shortness of breath, discomfort in the left side of the chest during rapid walking up to 300 m; 3) leg weakness during prolonged walking.

It is known from anamnesis that the patient has been actively involved in sports since childhood (basketball, biathlon), and her first elevation of AP to 160 / 100 mmHg was recorded at the age of 16 years (2015) during a physical examination. Due to her good health, the patient had not consulted a doctor until 2018 and had not taken any antihypertensive medications. In January 2018, leg weakness, headache, and dizziness appeared for the first time with a rise in AP to 220 / 120 mmHg. During the examination at the local district hospital, renovascular and endocrine hypertension were excluded and angiotensin-converting enzyme inhibitors, beta-blockers, and diuretics were prescribed to correct the AP. The patient was referred to the Cardiology Research Institute (Tomsk NRMC) for further examination and treatment in April 2020 due to failure to achieve the target AP values. In addition, it is known that the patient was the second child in the family, mother's pregnancy had no special features. Her menstruation had been regular since the age of 14. She denies the presence of chronic diseases, bad habits, aggravated heredity for cardiovascular diseases and malformations.

Upon physical examination: satisfactory general condition, clear consciousness, regular physique.

Body mass index – 27.1 kg / m<sup>2</sup>. The left border of the relative cardiac dullness is along the midclavicular line. Heart sounds are clear and rhythmic, second heart sound has accentuation over the aorta, heart rate is 76 beats per minute. A coarse systolic murmur at the Botkin – Erb's point, is noted passing to the vessels of the neck. Blood pressure in the arms: left – 203 / 115 mmHg, right – 206 / 115 mmHg; blood pressure in the legs: right and left – 140 / 90 mmHg. Other organs and systems had no clinically significant abnormalities. No significant abnormalities were detected according to the laboratory tests. ECG revealed Cornell voltage criteria for LV hypertrophy (R in aLV + S in V3 = 26 mm). According to transthoracic echocardiography, ejection fraction was 67%, heart chambers were of normal size, concentric LV hypertrophy was observed (LV myocardial mass 204 g, LV myocardial mass index 114 g/m<sup>2</sup>), valves were without evident structural changes, aortic regurgitation up to grade I was noted, the remaining systems function normally. Systolic pressure in the right ventricle was normal. No interchamber shunts were detected. CoA with a gradient of 80 / 32 mmHg was documented at the transition of the arch to the descending aorta (Fig. 1, 2).

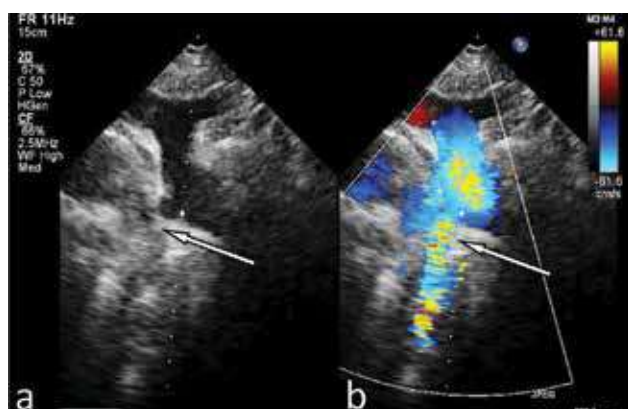


Fig. 1. Patient R., 20 years old: transthoracic echocardiography. Suprasternal position along the long axis: *a* – B-mode; *b* – color Doppler; white arrow indicates the site of stenosis

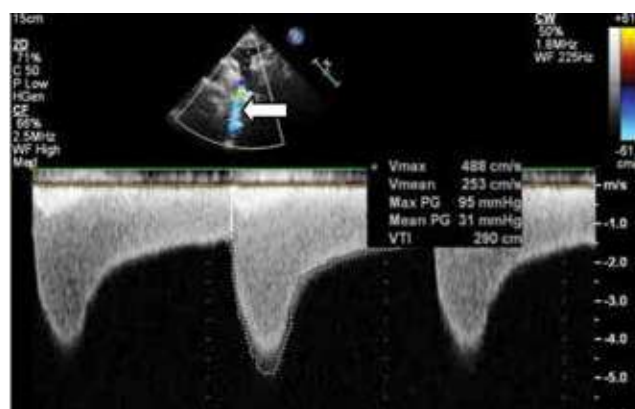


Fig. 2. Patient R., 20 years old: suprasternal position along the long axis. Continuous-wave Doppler; the arrow shows stenotic flow at the site of stenosis

MSCT aortography findings on Fig. 3 and Fig. 4 confirmed CT signs of aortic coarctation in the typical location, as well as collateral blood flow through the dilated intercostal and mammary arteries.

On the basis of the comprehensive clinical and instrumental examination, a diagnosis of congenital heart disease was made with aortic coarctation at the transition of the arch to the descending aorta, gradient 80/32 mmHg. Symptomatic arterial hypertension was

detected. Left ventricular hypertrophy, risk group 4. One week after admission to the hospital, surgical repair of aortic coarctation was performed, the arch and descending aorta were repaired with 20 mm Gore-Tex 20 suture, the aortopulmonary window was dissected. The postoperative period was without complications, the patient was discharged 10 days after the intervention with the recommendations of continuous administration of metoprolol 100 mg twice a day, torasemide

5 mg /day. The entire hospital stay, including surgery, took 17 days. In 3 months after discharge from hospital, AP stabilized at 130 / 80 mmHg, and torasemide

was discontinued due to a dramatic drop in AP. Taken into account tachycardia, a beta-blocker was preferred for further treatment.

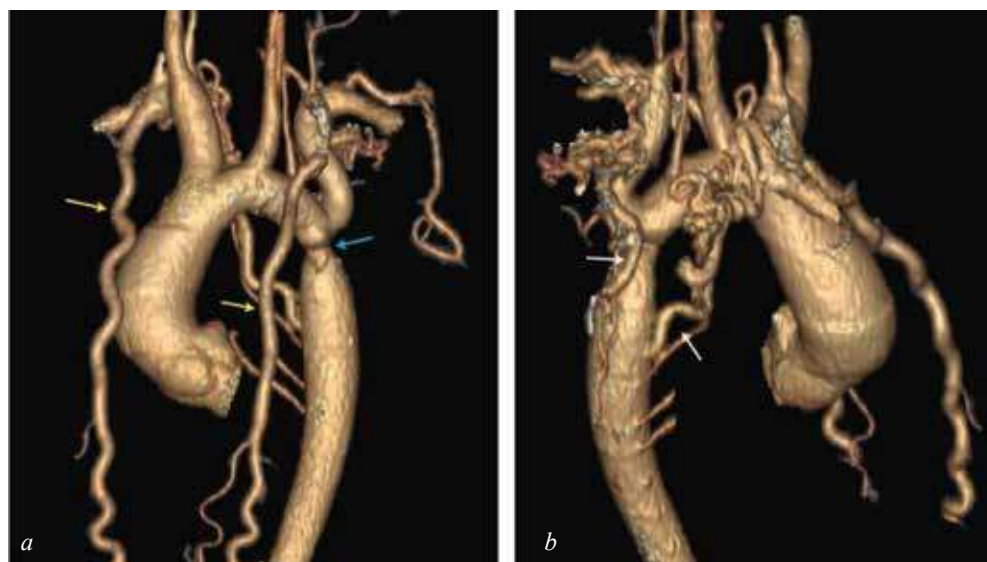


Fig. 3. Patient R., 20 years old: 3D reconstructions of the aorta in the anterior left oblique projection (a) and in the posterior right oblique projection (b). The blue arrow shows coarctation of the aorta; dilated tortuous intercostal arteries (white arrows) depart from the descending aorta; dilated mammary arteries depart from the subclavian arteries (yellow arrows).

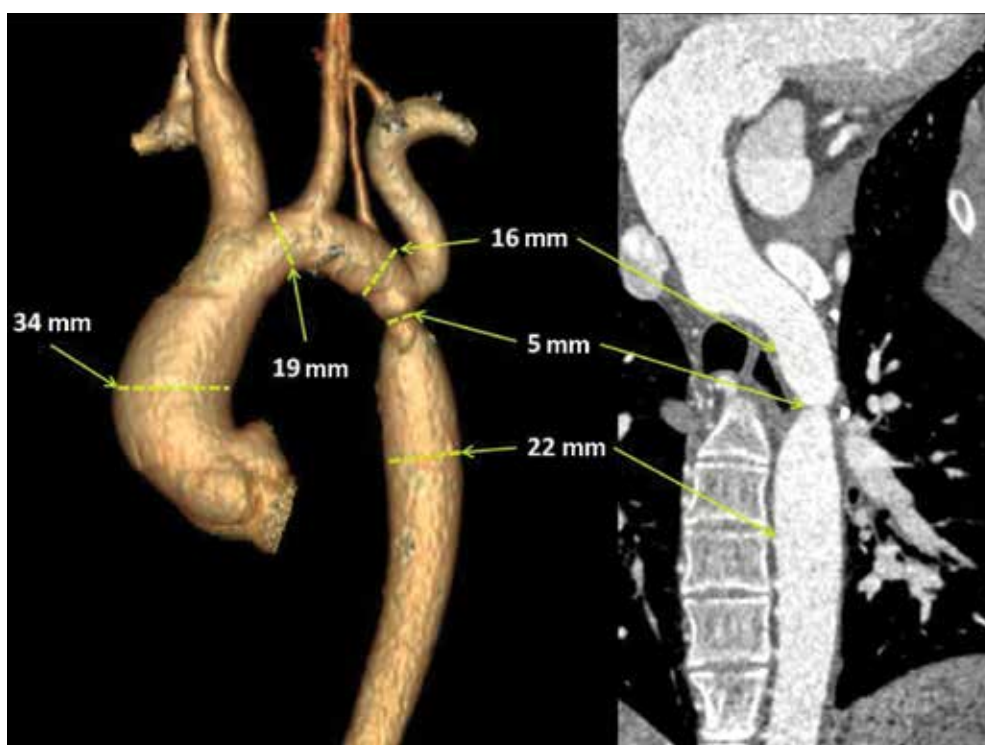


Fig. 4. Patient R., 20 years old: 3D reconstruction (left) and curvilinear reconstruction of the thoracic aorta with coarctation of the aorta. Severe narrowing of the aorta is identified distal to the origin of the left subclavian artery. Stenosis of the lumen is 70%. Post-stenotic dilatation of the aorta is detected distal to the narrowing. The root and ascending thoracic aorta are dilated. The left vertebral artery branches off the aortic arch and its orifice is proximal to the left subclavian artery

## CONCLUSION

A rarer, isolated type of aortic coarctation, asymptomatic course of the anomaly till the age of 16, and high physical fitness of the patient were the peculiarities of this clinical case, which disoriented primary care physicians during the examination, including that after manifestations of AH. Nevertheless, given the characteristic complaints and auscultation findings, tonometry on the lower extremities would have greatly facilitated the diagnosis. Referral of the patient to transthoracic echocardiography with obligatory inspection of the aorta from suprasternal access using Doppler color flow mapping and continuous-wave Doppler, as well as assessment of the abdominal blood flow would allow for timely diagnosis of this type of pathology.

It seems quite prominent that in this case, the diagnosis and correction of the defect in a specialized heart center took less than 3 weeks. However, despite a successful intervention, patients with aortic coarctation require long-term follow-up to monitor blood pressure, exclude the development of late aneurysms due to prolonged exposure of the vascular wall to increased hemodynamic pressure, and identify possible long-term postoperative complications.

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## Acknowledgements

The authors would like to express their gratitude to E.V. Tsymbaluk, Dr. Sci. (Med.), Head of the Department of Arterial Hypertension at Cardiology Research Institute, Tomsk NRMC, and E.V. Krivoschekov, Dr. Sci. (Med.), Head of the Cardiovascular Surgery Department at Cardiology Research Institute, Tomsk NRMC, for their help in preparation of clinical material.

## Authors contribution

Manukyan M.A. – drafting of the manuscript, preparation of the manuscript for publication. A.Y. Falkovskaya drafting of the manuscript, conception, critical revision of the manuscript for important intellectual content. V.F. Mordovin – advisory assistance, critical revision of the manuscript for important intellectual content. V.V. Saushkin. – carrying out of MSCT aortography, illustrations for the manuscript, consultative assistance, critical revision of the manuscript for important intellectual content. Ryabova T.R. – carrying out of ultrasound examination, illustrations for the manuscript, critical revision of the manuscript for important intellectual content.



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Received 18.08.2020

Accepted 28.12.2020