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Newly diagnosed congenitally corrected transposition of the great arteries in physically active 73-year-old men

Nowo rozpoznane skorygowane przełożenie wielkich pni tętniczych u aktywnego fizycznie siedemdziesięciolatka

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Abstract

A 73-year-old Caucasian male, so far engaged in regular, recreational physical activity, was admitted to the hospital due to a progressive decline in efficiency. As a result of the performed diagnostics, a congenitally corrected transposition of the great arteries with failure of the systemic ventricle was diagnosed. Despite the coexisting congenital heart defect, the patient remained physically active for several dozen years and performed high-intensity exertions.

Key words: CCTGA, congenitally corrected transposition of the great arteries, echocardiography, heart failure

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Case report

Seventy-three-year-old Caucasian man with decades of regular physical activity was admitted to the hospital due to severe dyspnea. Since a year before hospitalization, a subjective reduction in exercise tolerance. Since 2–3 months, non-specific chest pains and a tendency to edema in legs. Several days before hospitalization, intensified exertional dyspnea followed by orthopnea. Coexisting medical conditions — several years of history of hypertension and type 2 diabetes treated with oral medications. In youth, spontaneous pneumothorax occurred during an alpine expedition, with subsequent empyema treated with drainage. The patient underwent echocardiographic examinations several times — enlargement of the heart cavities with mildly reduced or normal contractility has been reported; the difficult imaging conditions were emphasized each time.

On admission to the hospital, the patient presented fair general condition, with orthopnea and dyspnea at rest, elevated blood pressure, signs of pulmonary congestion, and pitting edema in both legs. A chest radiograph shows right-side pleural effusion, a significantly enlarged outline of the heart, mesocardia (Figure 1).

In laboratory tests, elevated values of cardiac troponin I (hsCTn I: 1321 to > 1046 ng/L; n = 34 ng/L), increased values of B-type natriuretic peptide (BNP: 10512 pg/mL; n = = 125 pg/mL), and D-dimer (DD: 1852 ng/mL; n = 730 ng/mL).

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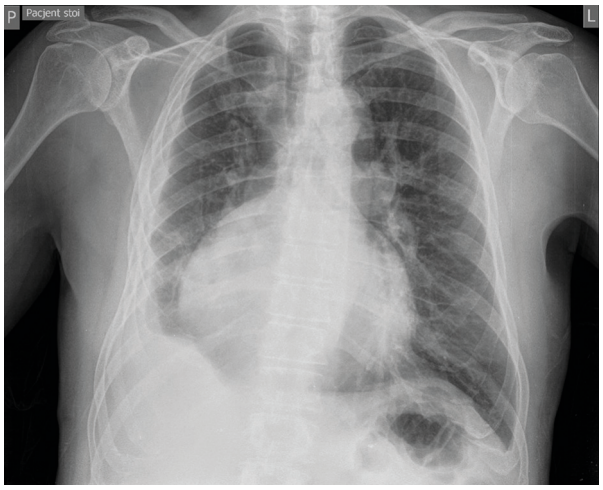

Figure 1. Chest radiography. Enlarged heart silhouette, shading in the area of the right diaphragmatic-costal sinus and along the side of the chest. Discrete parenchymal densities in the lower field of the right lung; mesocardia

Congenitally corrected transposition of the great arteries (CCTGA) was diagnosed on the basis of echocardiography. It was the first diagnosis of congenital heart disease in this patient. The heart was visualized from the sub-sternal projection (Figure 2). Examination showed an enlarged, spherical right ventricle (which is a systemic ventricle) with features of myocardial hypertrophy and significant impairment of systolic function (ejection fraction of 30%). The tricuspid valve was connected with the systemic ventricle, and mild regurgitation was present. Great arteries were extending from the heart with a parallel course characteristic of CCTGA. The pulmonary valve was with a mild regurgitation. The atria remained in a typical location. The ventricular septal defect and other coexisting heart defects were not visualized.

Coronary angiography was performed — the right coronary artery arising in a place typical for the left, left coronary artery arising in a place typical for the right; coronary arteries without atherosclerotic changes.

The Holter electrocardiogram device recorded numerous additional ventricular beats, including pairs and episodes of non-sustained ventricular tachycardia (Figure 3).

After the treatment, typical for congestive heart failure (diuretics, beta-blockers, angiotensin-converting enzyme inhibitors), a quick improvement of the general condition, reduction of BNP, and reduction of cardiac arrhythmias was achieved. In a follow-up, Holter electrocardiogram device recorded no complex forms of ventricular arrhythmia.

It should also be emphasized that the described patient remained physically active for most of his life (over 50 years), undertaking efforts of moderate and high intensity. In the last years, played squash regularly (3–4 trainings/week for 90 min) and participated in sports competitions. Additionally, about 30 minutes of brisk (5 km/h) walking a day. For about a year, resistance exercises 2–3 times a week for 60 minutes, swimming or aerobics. In his youth, the patient is actively hiking in the mountains. At 24 years of age climbed Kilimanjaro. Despite the congenital heart defect, the patient undertook intense physical activity for several dozen years and remained asymptomatic until old age.

Discussion

Congenitally corrected transposition of the great arteries is a rare cardiac malformation characterized by discordant atrioventricular (right atrium connected to the left ventricle, left atrium to the right ventricle) and ventricular-arterial

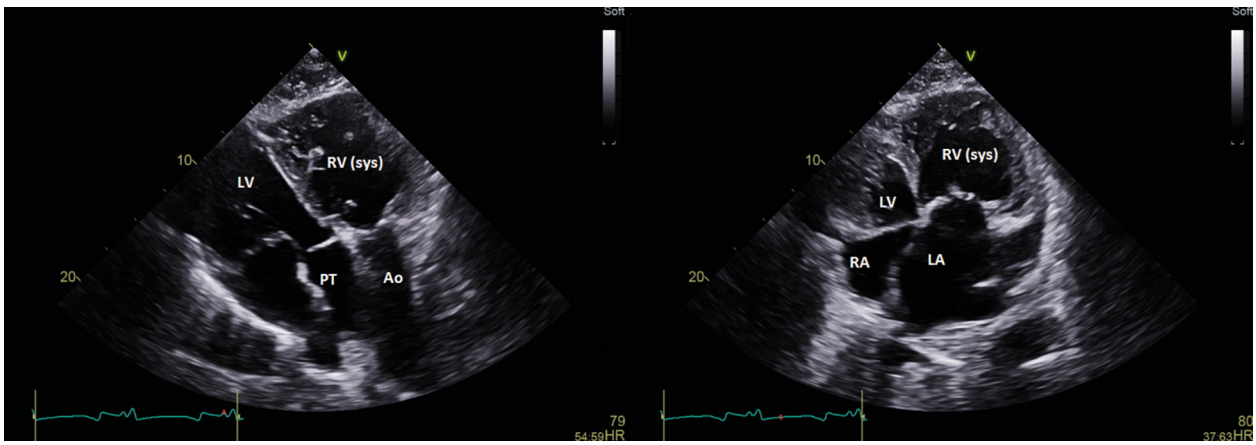

Figure 2. Echocardiographic examination, substernal projections. Enlarged, spherical right ventricle with myocardial hypertrophy, with significant impairment of contractility (ejection fraction 30%). Great arteries arising from the heart with a parallel course characteristic of congenitally corrected transposition of the great arteries; Ao — aorta; LA — left atrium; LV — left ventricle; PT — pulmonary trunk; RA — right atrium; RV (sys.) — right ventricle (systemic)

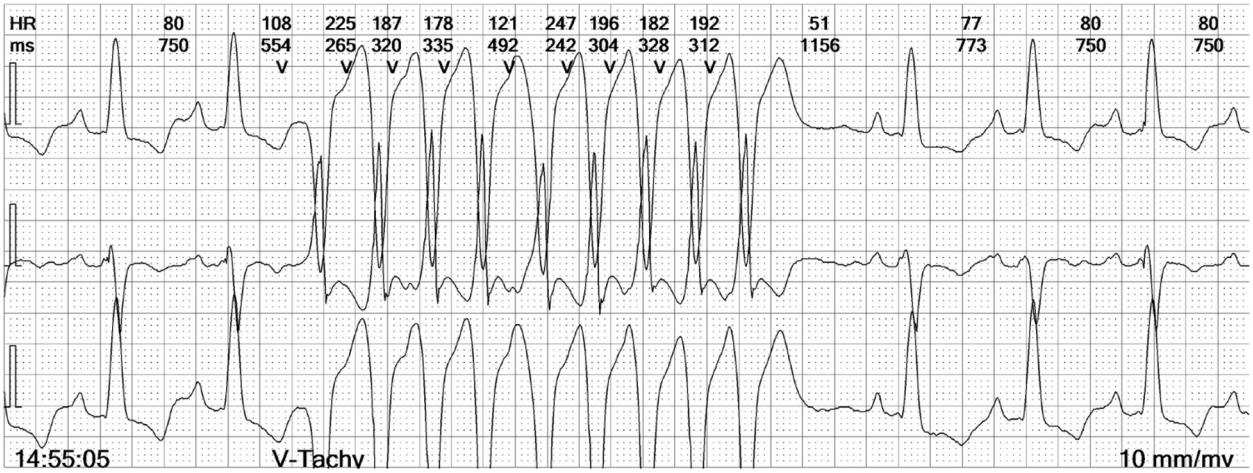

Figure 3. Holter electrocardiogram device. Non-sustained ventricular tachycardia

connection (aorta connected to the right ventricle, pulmonary trunk connected to the left ventricle). It accounts for 1% of congenital heart defects [1] and occurs in 1 in 33 thousand live births [2]. We distinguish a complex form in which the above-mentioned changes are accompanied by other anomalies, most often ventricular septal defect (70%) or valvular pulmonary stenosis (40%), and an isolated form, which may remain undiagnosed for a long time. The first symptoms appear most often in the fourth decade of life, when systemic ventricular dysfunction, tricuspid regurgitation, or complete atrioventricular block develop [3]. The average life expectancy of patients with CCTGA without associated defects is approximately 60 years, with accompanying defects — 40 years. Patients most often die of heart failure or ventricular arrhythmia [4].

One of the factors leading to progressive heart failure is the fact that the systemic ventricle is vascularized by the right coronary artery, which may lead to insufficient perfusion of the hypertrophied myocardium, and consequently to its progressive dysfunction, as muscle hypertrophy is not accompanied by an adequate proliferation of capillary vascularization. [2]

Another hypothesis is that the unfavorable shape of the systemic chamber results from remodeling aimed at maintaining adequate arterial pressure [5]. The presented patient belongs to a small group described in the literature who remained asymptomatic up to the age of 70. The oldest patient with newly diagnosed CCTGA described so far was 88 years old [6], and the oldest surviving patient was 92 years old [7].

Physical activity undertaken by people with congenital heart disease requires a systematic medical assessment: subjective and physical examination, additional tests, assessment of training intensity, and further medical supervision. Parameters of prognostic importance include ventricular function with the assessment of the ejection fraction, pulmonary artery pressure, presence of cardiac arrhythmias, the value of resting/exercise saturation, and width of the aorta [3].

Conflict of interest

None declared.

Streszczenie

Siedemdziesięciotrzyletni mężczyzna rasy kaukaskiej, dotychczas podejmujący regularną, rekreacyjną aktywność fizyczną został przyjęty do szpitala z powodu postępującego spadku wydolności. W wyniku przeprowadzonej diagnostyki rozpoznano skorygowane przełożenie wielkich pni tętniczych z niewydolnością komory systemowej. Pomimo współistniejącej wrodzonej wady serca pacjent przez kilkadziesiąt lat pozostawał aktywny fizycznie i wykonywał wysiłki o dużej intensywności.

Słowa kluczowe: ccTGA, skorygowane przełożenie wielkich tętnic, echokardiografia, wysiłek fizyczny, niewydolność serca

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