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CORRECTED TRANSPOSITION OF THE GREAT ARTERIES WITH COMPLETE ATRIOVENTRICULAR BLOCK AND ACCESSORY PATHWAY

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Clinical data of a 39-year-old patient with corrected transposition of the great arteries, combined with complete atrioventricular block and conduction along an accessory pathway, are presented. The features of changes in the structure of the conduction system, rhythm and conduction characteristic of this rare congenital heart disease are discussed.

Key words: congenital heart disease; corrected transposition of the great arteries; atrioventricular block; accessory pathways; electrocardiogram; Holter monitoring; permanent pacing.

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An unusual result of a patient's daily electrocardiogram (daily ECG) was published in the 2023 Journal of Arrhythmology No. 1 [1]. The readers had the opportunity to analyse the daily ECG data themselves and form their own opinion about the causes of the changes found. They were due to the presence of a rare congenital heart disease (CHD) - corrected transposition of the great arteries (CTGA) in the patient.

CTGA is less than 1% of all CHDs. The most important anatomical distinguishing feature of CTGA is the atrioventricular (AV) and ventriculo-arterial discordance, which leads to physiologically "normal" blood circulation. The patients with CTGA morphologically have a right atrium (RA) connected via a bicuspid "mitral" AV valve to morphologically the left ventricle (LV), which in turn is connected to the pulmonary artery trunk. Accordingly, the left atrium (LA) is morphologically connected via the tricuspid valve AV to the right ventricle (RV), which in turn is connected to the ascending aorta. Consequently, the LV is morphologically in the "pulmonary" position and provides for pulmonary blood flow, while the RV is morphologically in the "systemic" position and provides for systemic circu-

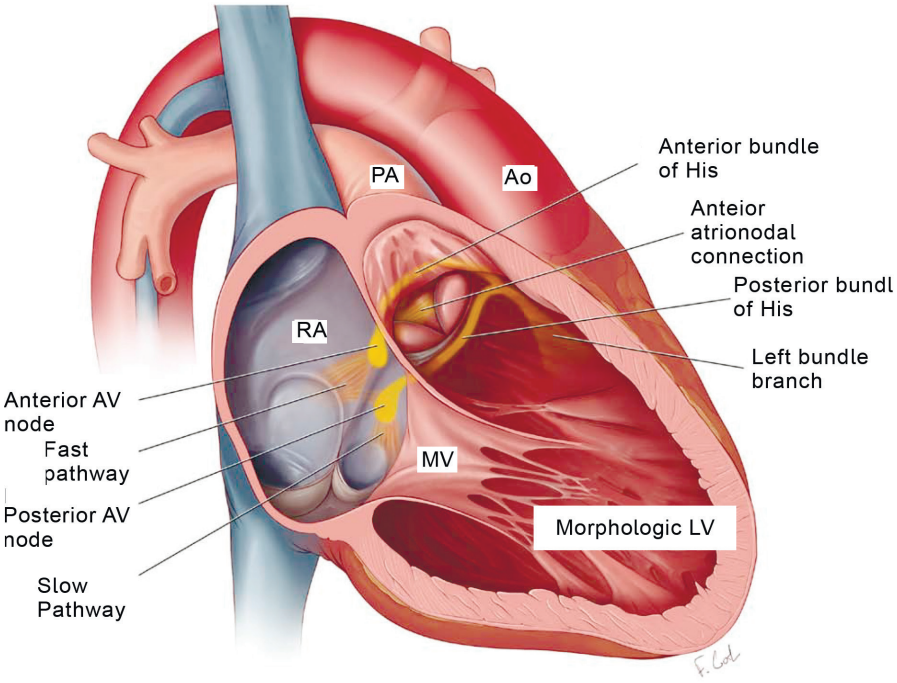

Fig. 1. Anatomy of the conductive system in congenital corrected transposition of the great arteries. The free wall of the right-hand chambers is cut out to visualize the septum. Two atrioventricular nodes (AVN) and bundle branches are presented. Note: Ao - aorta; LV - left ventricle; MV - mitral valve; PA - pulmonary artery; RA - right atrium. Modified from [8].

lation. As a result, over time, dysfunction of the systemic, morphologically RV with the development of heart failure occurs. Double discordance in CTGA can occur both in isolation and in combination with other intracardiac malformations: atrial inversion, atrial isomerism, dextrocardia (25%) [2], interventricular septal defect (70-80%) [3], pulmonary atresia and several others.

Patients with CTGA are characterized by rhythm and conduction abnormalities associated with the defect. Thus, patients with CTGA have complete AV block in 10% of cases, AV grade 1 and 2 block in 20-30% of cases, and the remaining patients have a high risk of spontaneously developing complete AV block, estimated at 2% per year over a lifetime [4]. Atrial rhythm disturbances are detected in 38% of patients with CTGA [5]. Due to double discordance, the cardiac conduction system in patients with CTGA has significant features. In patients with CTGA and normal atrial positioning, the usually located posterior AV node is hypoplastic and has no connection to other elements of the conduction system; the anterior AV node is located below the right atrial orifice near the lateral border of the fibrous contact of the right AV (mitral) and pulmonary valves (see Fig. 1). This abnormal AV node gives rise to a bundle branch that passes through the fibrous triangle, continuing into the subendocardial area of the ventricular myocardium. Upon reaching the interventricular septum, the bundle branch deviates posteriorly along the septum, giving the left bundle branch to the right side and the right bundle branch to the left side. In patients with atrial inversion, the passage of the conduction pathways resembles normal, with the AV node located posteriorly. After division into branches, it is possible to continue the conductive pathways as a blindly terminating anterior branch, not connecting to another AV node located anteriorly [6].

A 39-year-old patient consulted a cardiac surgeon with complaints of increasing dyspnea, decreased tolerance to physical activity, and presyncopal conditions over the past year. Congenital CTGA has been known since the age of 4. In childhood, cardiac cavity probing was performed at the A.N. Meshalkin Research Institute (Novosibirsk) and no CHD correction was indicated. The patient was monitored by a cardiologist at the place of residence. In 2007, ECG changes were detected, considered as WolFf-Parkinson-White (WPW) phenomenon, no surgical treatment was suggested. A history of 2 deliveries, healthy children, no CHD or rhythm abnormalities diagnosed in any of the children. When the patient applied to Krasnoyarsk Federal Center for Cardiovascular Surgery in October 2022, she was consulted by cardiac surgeons: CHD correction was not indicated due to preserved function of the systemic valve, the patient was referred to an arrhythmologist to decide if radiofrequency ablation (RFA) of the accessory pathway (AP) was necessary. According to daily ECG data, permanent complete AV block was detected, changes in the complex shape were regarded as intermittent WPW phenomenon (PQ - 100 ms, QRS - 120 ms, delta wave in V2-V6 leads), the same changes were registered on ECG.

Transthoracic echocardiography. RA: B-mode size - 4.0 cm, volume - 50 ml. LA: size in M-mode - 4.4 cm, volume - 62 ml. RV: not dilated. Systolic pulmonary artery pressure 37 mmHg. Inferior vena cava: size - 2.3 cm, collapsing on inspiration less than 50%. "Tricuspidal" valve: ring - 3.7 cm, regurgitation 2-3 st. "Mitral" valve: ring - 3.2 cm, regurgitation 1-2 st. LV: posterior wall in diastole - 0.7 cm, interventricular septum in diastole - 1 cm, final diastolic size - 3.9 cm, Simpson ejection fraction - 60%. RV: end-diastolic volume - 78 ml, end-systolic volume - 34 ml, ejection fraction - 56%. Aorta: arch - 2.5 cm, descending - 3.4 cm, abdominal - 1.7 cm, maximal pressure gradient - 8 mm Hg. Pulmonary artery: gradient - 12 mm Hg, regurgitation - 1 st. Additionally: septa intact, extrasystole on the monitor. Conclusion. Echocardiographic picture of CTGA. RV on the left, systemic, with the aorta branching off from it. LV on the right, with the PV branching off from

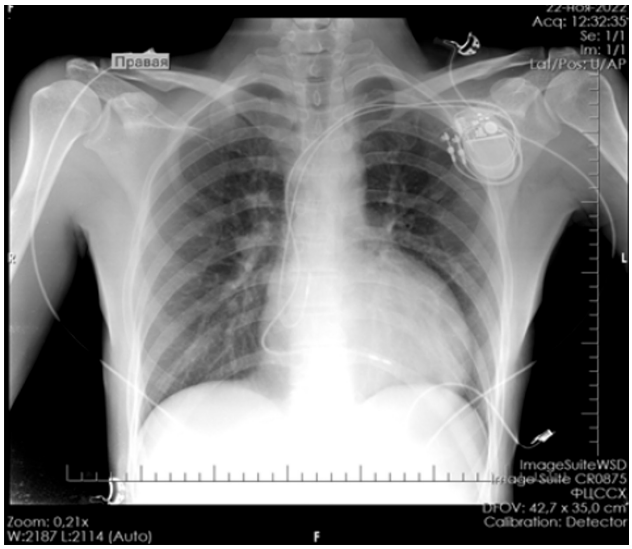

Fig. 2. Patient's X-ray.

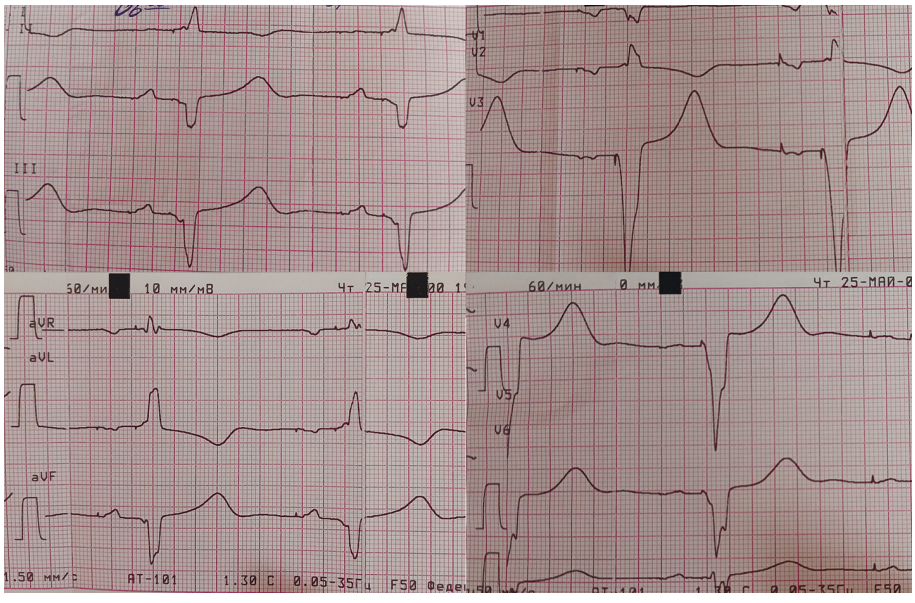

Fig. 3. ECG of a patient after pacemaker implantation.

it. Crossing of blood flow at the level of the main vessels and atria. Mitral insufficiency 1-2 degree. Systolic function of the systemic RV is preserved. Tricuspidal insufficiency of the 2-3 degree. Diastolic function of the RV is impaired by type 2. Expressed hypertrophy of the RV walls. LV myocardial contractility is satisfactory. No pericardial or pleural effusion. Pulmonary hypertension, SPPA 37 mmHg (determined by discharge on the "mitral" valve). The pulmonary artery is located in front and to the right of the aorta. The ascending aorta and arch to the right (normal).

The patient was hospitalized in the Krasnoyarsk Federal Center for Cardiovascular Surgery with the diagnosis: CHD. Corrected transposition of the great vessels. Stage 3 transient AV block. Transient AV block 2 degree, Mobitz 2. Intermittent WPW phenomenon. A follow-up examination was carried out, the results of which confirmed complete AV blockade, the sinus node function was assessed as preserved, the QRS complex shape changes were regarded as the WPW phenomenon, intermittent type. On November 18, 2022, a pacemaker (PM) was implanted with endocardial stimulation in DDD mode (Fig. 2, 3). In the early postoperative period, the patient developed a complication - left-sided pneumothorax; a pleural drain was applied. The complication resolved within 3 days.

At discharge, positive dynamics observed: the patient's dyspnea, presyncopal conditions, and tolerance to physical exertion increased. It was decided to abstain from RFA AP; dynamic monitoring is indicated. Three months after the operation, the patient noted a significant improvement of her well-being in a telephone conversation, described symptoms of heart failure, typical for functional class 1 according to NYHA. A follow-up examination is scheduled 6 months after PM implantation in the outpatient clinic of the Federal Center for Cardiovascular Surgery. The following is expected: PM programming, ECG, transthoracic and transesophageal echocardiography, cardiac magnetic resonance imaging, repeat consultation with a cardiologist at the outpatient clinic of the Federal Center for Cardiovascular Surgery to determine indications for systemic valve replacement.

The alternation of complete AV block and AP conduction has been described in patients with CTGA [7]. Interesting features of the ECG data they presented are the occurrence of second-degree AV block against a background of P-QRS-T complexes with signs of pre-excitation, and the presence of retrograde ventriculoatrial conduction against a background of complete AV block and following "narrow" QRS complexes in their rhythm [1]. It is known that patients with CTGA develop tachyarrhythmias along with AV block, including those associated with the presence of AP [8]. Such patients usually require radiofrequency catheter ablation, which can be associated with certain technical difficulties [9, 10]. The patient in question did not complain of palpitations. It was possible to assume that rhythm and conduction disturbances detected in the patient during the daily ECG [1] were associated with the presence of CHD based on high amplitude of P waves (in II lead it exceeds 300 μV) and unusual configuration of QRS complexes with signs of pre-excitation. In the compact part of these complexes (following the delta wave), the R waves prevail in the right thoracic leads and the S waves in the left leads, which can hardly be explained by anything other than the presence of CHD.

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