CLINICAL CASE

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A CASE OF ULTRASOUND DIAGNOSTICS OF NON-COMPACT MYOCARDIUM

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The article describes a case of ultrasound diagnostics of non-compact left ventricular myocardium. Echocardiography allowed establishing a rare congenital pathology of the left ventricular myocardium.

Keywords: non-compact myocardium, echocardiography, diagnostic criteria.

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Non-compact, or spongy, left ventricular myocardium (NLVM) is a diagnosis introduced into clinical practice relatively recently in connection with the improvement of methods for examining the heart and, therefore, is of great interest for clinicians, ultrasound diagnostics, MRI. Currently, this disease remains little known to a wide range of clinicians and causes difficulties in the correct diagnosis.

According to the literature, the prevalence of this pathology among adults is 0.014% [10]. In pediatric practice, the proportion of this pathology is 9.2% of all cases of diagnosed cardiomyopathies, ranking third after hypertrophic cardiomyopathy and dilated cardiomyopathy [9]. At the same time, experts note that real figures may significantly exceed official data.

Embryogenesis: between the 5th and 8th weeks of embryonic development, the heart muscle is organized - the fiber network is thickened and the intertrabecular lacunae are narrowed. At the same time, the formation of coronary circulation occurs, the intertrabecular spaces are reduced to the size of capillaries. In case of a violation of the normal course of this process, zones of non-compact myocardium with increased trabecularity (more than three trabeculae) remain in the heart of the newborn. In this case, deep intertrabecular spaces are formed - lacunae or sinusoids [1].

The American Heart Association classifies the non-compact LV myocardium as a primary genetic cardiomyopathy, and the WHO identifies it as an unclassified cardiomyopathy [3]. According to Benjamin et al., The non-compactness of the left ventricular myocardium is a "spongy" myocardium formed as a result of a violation of the intrauterine myocardium, characterized by a thin compact epicardial layer and a thick non-compact endocardial layer with pronounced trabecularity and slit-like spaces communicating with the cavity rather than the left ventricle coronary blood flow [4]. There is evidence of the hereditary nature of this disease. Recent studies indicate the presence of mutations in the G4.5 gene of the Xq28 locus [7].

Echocardiography is currently the method of choice for diagnosing this disease. According to the literature, there are a number of criteria that suggest that a patient has a violation of the structure of the left ventricular myocardium during ultrasound examination. The first diagnostic criteria were proposed by Chin et al., Who calculated the ratio between the distance from the epicardium to the base of the trabeculae (X) and the distance from the epicardium to the apex of the trabeculae (Y), measured at the end of diastole. At the same time, a progressive decrease in the X / Y<0.5 ratio was observed in patients with noncompact cardiomyopathy [5]. The criteria of Jenni et al are most often used in clinical practice: the ratio of the thickness of the non-compact layer to the thickness of the compact layer >2.0, measured at the end of systole, in the projection of the short axis, in the absence of other cardiac anomalies [8]. Stollberger C. et al. included in the criteria the presence of at least 3 separate trabeculae, as well as the ratio of the thickness of the compact layer and the non-compact layer 2.0 [11]. Currently, for the correct diagnosis, it is recommended to use the following diagnostic criteria: the appearance of 4 prominent trabeculae and deep intertrabecular spaces; the appearance of blood flow from the LV cavity into the intertrabecular spaces; segments of unconsolidated myocardium mainly include the apex, as well as the middle third of the inferior lateral wall of the LV; clear two-layer structure of the myocardium; the thickness of the unconsolidated subendocardial layer at the end of systole is more than twice the thickness of the compacted subepicardial layer [6].

Currently, the "gold standard" for diagnosing disorders of the left ventricular myocardium is magnetic resonance imaging of the heart, which allows visualizing the bilayer structure of the myocar-

dium with a higher spatial resolution than routine echocardiography [3]. Below is a clinical case of ultrasound diagnostics of non-compact left ventricular myocardium.

Patient B., born May 29, 2019, Sakha. The child was born from 6 pregnancies, from a 28-year-old mother with a burdened obstetric and gynecological history: the first pregnancy ended in spontaneous miscarriage at 6 weeks of gestation; a boy from 3 pregnancies died 2 days after birth.

This pregnancy proceeded with acute respiratory infections in the 1st half without fever. Ultrasound examination at 19 weeks showed the patient to have low placentation, at 27-28 weeks - complete placenta previa. Ultrasound examination of the fetus at the 31st week of pregnancy revealed cardiomegaly due to expansion of the right heart, regurgitation on the tricuspid valve, displacement of the fibrous ring of the tricuspid valve with symptoms of "atrialization" of the right ventricle. According to the ultrasound examination, pathology of the heart was suspected and the conclusion was made: the echographic picture may correspond to dysplasia of the tricuspid valve, Ebstein's anomaly. Partial abnormal drainage of the pulmonary veins, hypoplasia of the aortic arch cannot be ruled out.

The third childbirth is planned, operational for a period of 35 weeks. The birth weight of the child is 55 cm, the body length is 47 cm. The assessment of the newborn according to the Apgar scale is 8/8. With dynamic observation of the child during the first day, the condition was regarded as severe, due to cardiac pathology, prematurity, general neurological symptoms, with negative dynamics. The child was feeding through a tube, spitting up curdled milk, did not cry on examination, general pastiness was noted, spontaneous motor activity was reduced.

On the first day after birth in the ward of the Department of Intensive Care, Anesthesiology and Reanimation of Newborns, a comprehensive ultrasound examination of the heart was carried out using the “Acuson X300” apparatus manufactured by Siemens. Muscle tone and reflexes were drastically reduced. The skin is pale pink with a cyanotic tinge. Respiration rate 50 / min. SpO2 -98%. phased sensor P8-4. The study revealed: a ductus arteriosus with a diameter of 0.31 cm, an atrial septal aneurysm with blood discharge from left to right 0.36 cm. Expansion of the cavities of the right ventricle up to 1.52 cm and of the right atrium up to 2.29 cm was noted; revealed regurgitation on the tricuspid valve of grade 3,

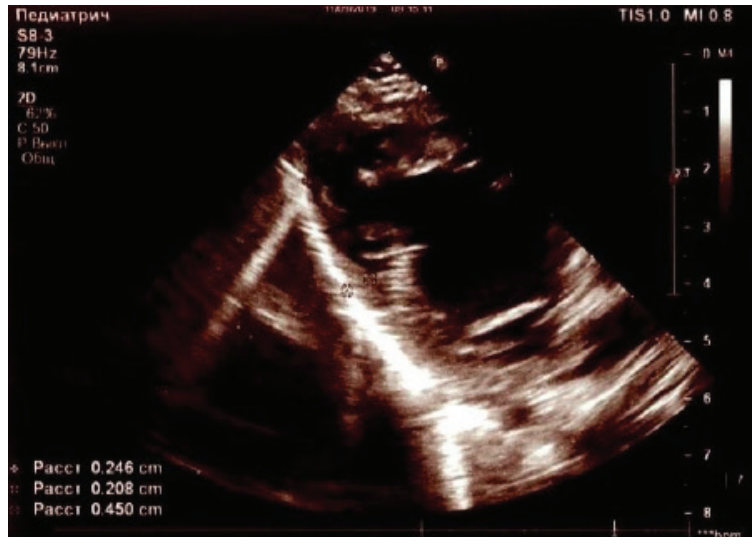

Fig. 1. Areas of myocardial thinning in the apex and areas of increased trabecularity of the left ventricle are visible

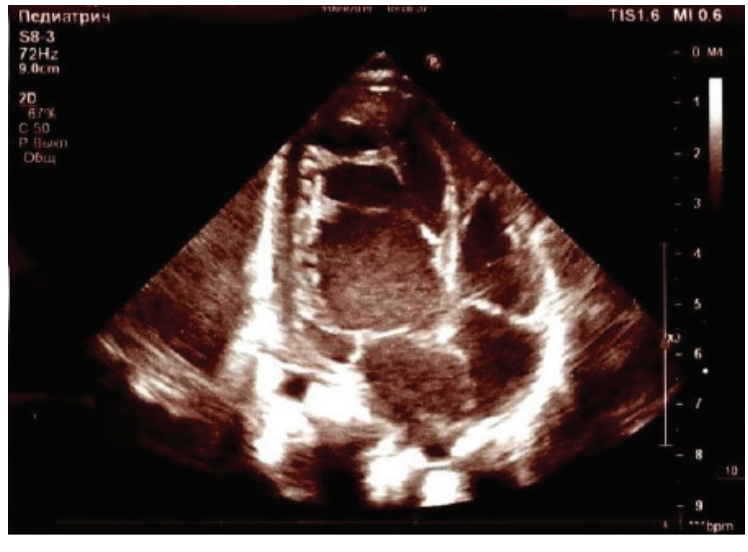

Fig. 2. Two-layer structure of the left ventricular myocardium. Dilation of the left ventricular cavity

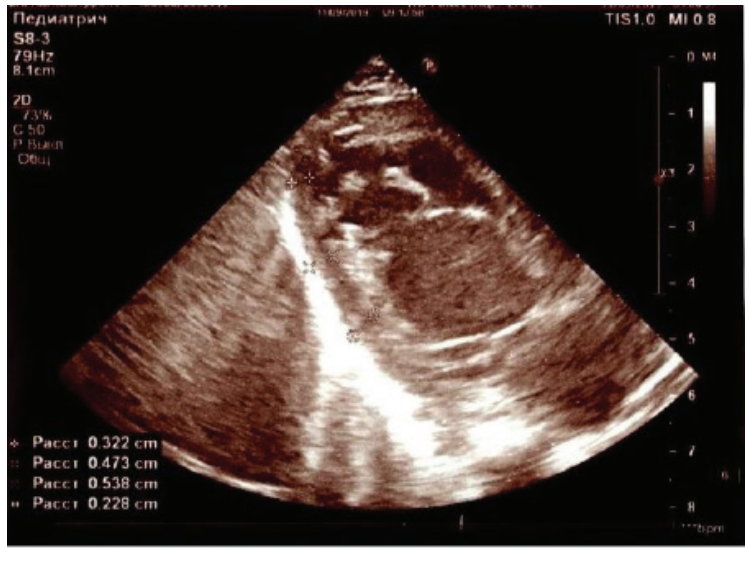

Fig. 3. Trabeculae protruding into the cavity of the left ventricle with deep intertrabecular spaces

on the mitral valve - grade 2. The child was diagnosed with grade 2 pulmonary hypertension, right ventricular myocardial hypertrophy, and expansion of the pulmonary artery trunk. At the same time, there was a decrease in the systolic function of the left ventricular myocardium, ejection fraction of 53.7%. Of course - the diastolic size of the left ventricle was within the age norm. It should be noted that during the ultrasound examination, difficulties were noted associated with tachycardia (up to 183 beats per minute) and anxiety of the child.

The child received treatment in the Department of Intensive Care and Reanimation of Newborns and the Department of Neonatal Pathology for two months, then was discharged home. Clinically, the disease proceeded according to all signs of cardiomyopathy.

Subsequently, the child was repeatedly admitted to the hospital due to the deterioration of his condition. Ultrasound examinations of the heart showed a progressive expansion of the left ventricular cavity, and increased trabecularity of the left ventricle was noted. With the passage of time, there was a change in the configuration of the heart in the form of a spherical shape.

In the fourth month of the child's life, an expert ultrasound examination of the heart was carried out using an Epiq 7 apparatus, manufactured by Phillips, with a phased transducer S8-3. From the study protocol: the left atrium is dilated to 1.9 cm.The left ventricle is dilated, the end-diastolic size is 3.5 cm.The spherical shape of the left ventricle with areas of increased trabecularity and areas of myocardial thinning in the apex area up to 0.2 cm.

A two-layer structure of the myocardium with a thinned compact and thickened non-compact layer was found, identified from the parasternal position. The N / C ratio was more than 2, where N is the non-compact layer of the myocardium, C is the compact layer of the myocardium.

The presence of numerous, excessively protruding into the cavity of the left ventricle trabeculae with deep intertrabecular spaces was noted. The right parts of the heart were not dilated. The calculated pressure in the right ventricle was 50-60 mm Hg. Attention was drawn to a diffuse decrease in the contractility of the left ventricle with areas of aki, dyskinesis in the apical segments of the left ventricle. At the time of the study, no additional echo structures were found in the cardiac cavities.

Based on the data obtained, it was concluded: Increased trabecularity of the left ventricle with areas of thinning. This echo pattern may correspond to a non-compact myocardium. Pronounced diffuse decrease in left ventricular contractility, apex dyskinesis. EF 30-35%. Expansion of the left ventricular cavity, end-diastolic size - 3.5 cm. Insufficiency of the mitral valve of the 3rd degree, probably of a relative nature. Regurgitation on the tricuspid valve of the 2nd degree. Pulmonary hypertension 2 degrees.

Subsequently, the child was consulted in absentia at the Federal State Budgetary Institution "National Medical Research Center named after Academician E.N. Meshalkin" of the Ministry of Health of the Russian Federation, where he was diagnosed with: Cardiomyopathy I42.0 - Dilated cardiomyopathy: Non-compact myocardium; Insufficiency of the mitral valve of the 2nd degree. Insufficiency of the tricuspid valve 2-3 degrees. Pulmonary hypertension 1-2 degrees. Congestive heart failure: grade 2A chronic heart failure. FC 4, threatened by the development of life-threatening arrhythmias. Threatened by sudden cardiac death syndrome. Perinatal CNS lesion of hypoxic genesis. Delayed static-motor development. Solution: An examination by a geneticist is recommended, it is necessary to take a blood test on the panel for "hereditary heart disease" to exclude microdelicia and monogenic pathology.

The parents refused to pass the analysis on the panel for "hereditary heart disease".

At the 10th month of life, the child with a worsening condition was urgently hospitalized in the Department of Intensive Care, Anesthesiology and Reanimation of the Pediatric Center. On admission, an ultrasound examination of the heart was performed. Attention was drawn to the pronounced trabecularity of the left ventricle - additional trabeculae, multiple and thickened. There was a significant expansion of the cavities of the left ventricle (EDC 3.0-3.69 cm), left atrium (3.0 cm), right ventricle (2.2 cm), right atrium (4.0 cm), pulmonary artery trunk (1 , 4-1.5 cm). The total contractility of the left ventricular myocardium was significantly reduced. EF 22.5-26%. In the cavity of the left ventricle - hyperechoic heterogeneous with uneven, indistinct contours of the echo structure - thrombi: closer to the apex, along the IVS, measuring 1.27 * 0.49 cm; in the area of the apex measuring 0.36 * 0.31 cm. Insufficiency of the tricuspid valve of the 4th degree, the mitral valve of the 2-3 degree, the pulmonary valve of the 1st degree. Pulmonary hypertension 2 degrees. Right ventricular myocardial hypertrophy.

On the X-ray imaging of the brain revealed: a large area of acute ischemia in the right parietal-temporal region, expansion of the ventricular system, convexital subarachnoid spaces on both sides. Three main clinical syndromes play a leading role in the pathogenesis of non-compact left ventricular myocardium: heart failure (73%), arrhythmic syndrome (40%), thromboembolic syndrome (33%). Patient B. had heart failure 2B, FC IV, WPW syndrome, which is most common in children with LVNM, transient ischemic attacks.

Despite the ongoing therapy, the child died three days after admission to the hospital. The diagnosis was confirmed by postmortem examination. Autopsy: left ventricular wall thickness 1.2 cm with thinning areas 0.2 cm. The myocardium is pale brown; deep intertrabecular spaces are noted in the right and left ventricles. Trabeculae are whitish, thickened, stony density. In the left ventricle in the region of the apex in the intertrabecular cavity there is a parietal red thrombus with a diameter of 0.3 cm. Histological examination: heart: stromal edema, moderate hypertrophy of cardiomyocytes, muscle fibers with foci of hyalinosis, sclerosis, fibrosis, fragmentation of muscle fibers is noted. In the left ventricle, there are sections of a thinned wall, consisting of a thickened endocardium and epicardium, which are represented by connective and adipose tissue. When examining the brain: perivascular, pericellular edema. In the tissue of the left and right hemispheres, areas with necrosis and hemorrhages are noted. Vessels along the periphery of the site are full-blooded with symptoms of erythrocyte stasis, leukocyte stasis, leukodiapedesis sites. In some of the vessels, mixed blood clots are noted. The given clinical case is presented for review, to help practitioners of ultrasound diagnostics for the correct interpretation of the echocardiographic picture of such a rare pathology as non-compact left ventricular myocardium, since literature data indicate that the actual frequency of this disease may be higher than the official data.

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THROMBOTIC COMPLICATIONS ASSOCIATED WITH THE NOVEL CORONAVIRUS INFECTION COVID-19

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We present a clinical case of a fatal thrombotic complication in a patient with coronavirus disease 2019 (COVID-19) in the recuperative period. A retrospective analysis of the patient's medical records with the chronology of laboratory and instrumental examinations, clinical course and intensive therapy was performed. The algorithm of pulmonary embolism (PE) diagnosis, validity and decision-making concerning thrombolytic therapy and extracorporeal membrane oxygenation are shown. The results of autopsy are also presented, which confirmed the diagnosis of PE and the appropriateness of medical interventions.

Keywords: COVID-19, recuperation period, myocarditis, thrombotic complications, pulmonary embolism, acute respiratory failure, artificial ventilation, extracorporeal membrane oxygenation.

Introduction. Covid-associated severe thrombotic complications develop in 17% of patients with coronavirus disease 2019 (COVID-19) and are one of the main causes of patient's death [1, 2].

Thrombosis is observed more often in people with severe course of the disease, with large area of lung lesions [4], in elderly patients, as well as in patients with severe comorbid conditions [7]. At the same time, the probability of thrombotic complications persists in patients for several months after COVID-19, which is a serious threat and explains the need to control the hemostasis system and to continue anticoagulant therapy after the patient is discharged from the hospital.

We present a clinical case of COVID-associated pulmonary embolism (PE) in the recuperative period in a young female patient without comorbid conditions, and 16% of lungs affected by COVID-19.

The **aim** of the investigation was to analyze the clinical course of and intensive therapy interventions in severe thrombotic complication in a patient with COVID-19.

Clinical observation. Patient M., female, 36 years old. On February 24, 2022 at 18:40 she was delivered by the ambulance to the Republican Hospital №1-National Center of Medicine (RHN№1-NCM) with the referral diagnosis: "Viral myocarditis? Opulent pericarditis. Bilateral polysegmental pneumonia. COVID-19 reconvalescent since February 19, 2022."

The patient complained of generalized weakness and palpitations up to 112 beats/min, elevated blood pressure (BP) up to 150/80 mmHg, dyspnea on minimal physical exertion, bilateral lower legs edema. On physical examination: height 164 cm, weight 74 kg (body mass index 27.5). General condition of the patient was severe. Patient was alert (15 points on the Glasgow Coma Scale (GCS)) and oriented. The skin was of normal color, normal moisture, turgor was preserved. Visible mucous membranes of normal color and moisture. Body temperature was 36.1˚C. Breathing unlabored, without the involvement of auxiliary muscles. On auscultation – breathing was conducted in all pulmonary fields, with increased intensity, symmetrical on both sides, no rales. Respiratory rate (RR) 18-20 breaths/min, SpO_2 - 89% on room air and 94% when insufflating 5 L/min of humidified oxygen. Cardiac tones muffled, rhythmic. BP -121/69 mmHg, heart rate (HR) 101/min. The tongue was clean and moist. The abdomen was not swollen, symmetrical, soft and painless on palpation. The liver and spleen were not enlarged. Intestinal peristalsis was active, uniform. No meteorism. No peripheral edemas. Diuresis, according to the patient, was sufficient.

It is known from the anamnesis that she considers herself sick since January 29, 2022, when she began to feel generalized weakness, vomiting, liquid stools up to 5 times a day. The patient

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