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Multifaces of hypertrophic cardiomyopathy: a case of transformation of hypertrophic phenotype into dilated

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ABSTRACT

The article presents a clinical case of a rather rare course of hypertrophic cardiomyopathy with the transformation of a hypertrophic phenotype into a dilated phenotype against the background of the "burned-out phase" phenomenon, ventricular and supraventricular rhythm disturbances, and multiple genetic mutations. Timely started disease-modifying therapy (quadruple therapy) for chronic heart failure led to reverse positive remodeling of the left chambers of the heart.

Keywords: hypertrophic burnout cardiomyopathy; non-sustained ventricular tachycardia; atrial fibrillation; global longitudinal strain; left atrial strain; quadruple therapy.

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Многоликая гипертрофическая кардиомиопатия: случай трансформации гипертрофического фенотипа в дилатационный

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АННОТАЦИЯ

Представлен клинический случай довольно редкого течения гипертрофической кардиомиопатии с трансформацией гипертрофического фенотипа в дилатационный фенотип на фоне феномена «выгорания» (burned-out phase), желудочковых и суправентрикулярных нарушений ритма, множественных генетических мутаций. Своевременно начатая болезнь-модифицирующая терапия (квадротерапия) хронической сердечной недостаточности привела к обратному позитивному ремоделированию левых камер сердца у пациента.

Ключевые слова: гипертрофическая кардиомиопатия выгорания; неустойчивая желудочковая тахикардия; фибрилляция предсердий; глобальная продольная деформация; продольная деформация левого предсердия; квадротерапия.

Как цитировать

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INTRODUCTION

The 2023 ESC Guidelines for the Management of Cardiomyopathies have largely changed the approaches to the classification of cardiomyopathies (CMPs) [1]. Currently, the following types of CMPs are distinguished: hypertrophic, dilated, Non–dilated left ventricular cardiomyopathy (NDLVC), arrhythmogenic, and restrictive cardiomyopathy.

The recommendations are based on a phenotypic approach to CMP classification and diagnosis. Importantly, family members may have different phenotypic manifestations, and in the same patient, as the disease develops and progresses, a phenotypic variant of CMP may transform into another. The predominant cardiac phenotype at the time of initial diagnosis should guide the diagnosis and follow-up of the patient. The case of using this approach in real clinical practice is presented herein.

CLINICAL CASE

Patient R., aged 62 years, has been under the care of the clinic since 2002. Regarding his medical history, he actively participated in various sports in his youth, including alpine skiing and wrestling. The suspicion of heart pathology first arose in 1995 (at the age of 34), when, for the first time in his life, an electrocardiogram (ECG) was taken before a competition (the results were not provided). ECG changes detected led to the diagnosis of an acute myocardial infarction, despite the absence of any complaints at the time. The patient was hospitalized for 3 weeks, followed by rehabilitation. After 2 months, the polyclinic at the patient's place of residence again detected signs of acute myocardial infarction on the control ECG and suggested rehospitalization. However, the patient categorically refused this suggestion due to the absence of complaints and good health. Complaints of palpitations and chest discomfort during moderate physical activity were first documented in 2002, at the age of 41. The patient was subsequently referred to the city antiarrhythmic center for further evaluation. Upon examination, he was diagnosed with hypertrophic CMP (HCMP) without left ventricular (LV) outflow tract obstruction. Daily ECG monitoring revealed a high premature ventricular complexes(PVCs) count, with approximately 10.000 events per day. Coronarography was performed for the pain syndrome. The subepicardial coronary arteries were free of hemodynamically significant stenoses. In addition, coronary systolic reverse flow, previously described in the literature in patients with HCMP and identified as a potential cause of subendocardial myocardial ischemia, was observed [2, 3]. A slow-release metoprolol succinate (Betaloc 30K®) at a dose of 100 mg per day was prescribed. The number of PVCs decreased during the therapy, although they did not entirely disappear. An attempt to increase the dose of beta-blocker was accompanied by a decrease in systolic blood pressure to 85 mmHg. In 2007 (at the age of 46), sustained hemodynamically significant ventricular tachycardia (VT) accompanied by a drop in blood pressure was recorded for the first time; in addition, paroxysmal and persistent forms of atrial fibrillation (AF) occurred. For several months, in addition to slow-release metoprolol succinate, the patient received amiodarone, which demonstrated a positive clinical effect. However, amiodarone was discontinued because of the occurrence of cordarone-induced thyrotoxicosis. In the same year, the patient received a dual-chamber implantable cardioverter-defibrillator (ICD) that can control ventricular rhythm disturbances and had function of cardioversion of AF. Owing to the frequent painful triggering of the ICD in response to AF and the deterioration of the patient's quality of life, the patient and his attending physician decided to disable the function of cardioversion of AF. In 2008, pulmonary vein cryoisolation was performed because of the frequent symptomatic episodes of AF. During several years after of pulmonary vein cryoisolation AF did not recur. Considering the high risk of AF recurrence and structural changes in the left chambers of the heart, the patient took propafenone 150 mg 2 times a day and warfarin in addition to metoprolol succinate sustained release. A gradual increase in the dosage of metoprolol succinate (increases of 12.5 mg at intervals of no more than 2 weeks) allowed us to reach the maximum dose (200 mg per day). The therapy was effective in preventing AF recurrence, although recurrent sustained VT led to ICD activation several times a year. Because the ICD battery depleted, the ICD was replaced in 2012. The newly installed ICD did not has the function of cardiovercion of AF but had a function of antitachycardia pacing (ATP) as a way to terminate VT. Recurrent sustained VT persisted and led to ICD activation several times a year. Since 2018, rare AF recurrence have been documented. In 2018 the patient was diagnosed with type 2 diabetes mellitus and dyslipidemia. In addition to rhythm disturbances, the patient reported chest discomfort and dyspnea during moderate exercise, with progressive worsening of exercise tolerance. Since 2018 patient took slow-release metoprolol succinate (200 mg daily), apiksaban (5 mg twice daily), dapagliflozin (10 mg daily), and rosuvastatin (20 mg daily).

In 2020, the ICD was replaced once more due to battery exhaustion. Repeated coronary angiography was performed before ICD replacement. Coronary angiography did not register coronary systolic reversed flow phenomenon. A 60% stenosis of diagonal branch of the left coronary artery

was detected. Fractional flow reserve (FFR) was assessed (>0.8). So the stenosis did not require revascularization. No significant changes were observed in the other coronary arteries.

The patient's condition markedly decline in February 2021. He presented to the clinic with complaints of dyspnea and chest discomfort, not only by exercise, but also at night. Echocardiography revealed a markedly different picture than that observed during the initial treatment in 2002. LV exhibited slight dilatation, with an end-diastolic volume index (EDVI) of 75 mL/m^2. The left atrium (LA) demonstrated pronounced dilatation, with a left atrial volume index (LAVI) of 68 ml/m^2. Diffuse hypokinesia of all LV walls, a reduction in myocardial thickness of the interventricular septum (IVS) from 19 to 12 mm, and an decrease in LV ejection fraction (EF) up to 40% were observed. An elevation in pulmonary artery systolic pressure (PASP) up to 52 mm Hg was also noted. In addition to severe systolic dysfunction, the patient had severe diastolic dysfunction (grade 3):

$$VE / VA = 2{,}1;\ VE / Em \text{ (mean)} = 16.3,$$

where *VE* is the velocity of the transmittral blood flow in the rapid filling phase, *VA* is the velocity of the transmittral blood flow at LA systole, and *Em* is the mean value of the sum of the velocities of the septal and lateral segments of the mitral valve annulus.

Contrasted chest computed tomography (CT), which was performed to exclude pulmonary embolism as the cause of the rapid deterioration in the condition, confirmed dilatation of the left heart chambers, fluid in the posterior parts of the pleural cavities on both sides (not more than 150 mL on each side), and signs of interstitial pulmonary edema. No data supporting pulmonary embolism were obtained. Brain natriuretic peptide level were elevated up to 1,683 pkg/mL. The patient did not undergo cardiac magnetic resonance imaging (MRI) because he had an ICD. Torasemide (10 mg daily), eplerenone (25 mg daily with subsequent increase in dose to 50 mg daily after 1 month), and valsartan + sacubitril (Uperio®) dose titration from 50 mg twice daily were added to the current therapy with metoprolol succinate (200 mg daily), apixaban (5 mg twice daily), dapagliflozin (10 mg daily), rosuvastatin (20 mg daily). Propafenone was discontinued because the EF decreased. After stabilization of the condition, stress echocardiography (stress EchoCG test) was performed. The patient's condition was considered transformation of a hypertrophic phenotype into a dilated phenotype with a burned-out phase and ventricular and supraventricular rhythm disturbances [3, 4]. EF dynamics for 2008–2021 are shown in Table 1.

Genetic testing was performed owing to the atypical disease course. Massive parallel sequencing of a panel of 17 genes associated with HCMP revealed two mutations: Glu163del mutation in *TNNT2* (heterozygous carrier), which codes for the synthesis of the troponin T protein. This mutation is clearly associated with HCMP and has a high penetrance. In addition, the identified mutation was associated with a high risk of sudden cardiac death (SCD). The second mutation, a truncating variant located in the M-band, was found in *TTN* (heterozygous carrier), which codes for the synthesis of titin protein. It is likely to be pathogenic. *TTN* mutations are associated with several types of cardiac and skeletal myopathies (hypertrophic, dilated, and restrictive CMPs, LV hypertrabecularity, distal Myoshi muscular dystrophy, Salih myopathy, etc.).

The patient has two children: the older daughter has confirmed HCMP but has not been genotyped, and the younger son has no genetic or phenotypic manifestations of HCMP.

During therapy, the patient's condition improved significantly, and the dyspnea stopped. Rhythm disturbances persisted as rare non-sustained atrial tachycardia and VT. The maximum well-tolerated daily dose of valsartan and sacubitril did not exceed 200 mg when titrated, and persistent hypotension occurred when further increases were attempted. The patient was then seen in May 2022, and an ECG was performed (Fig. 1).

The control EchoCG demonstrated positive dynamics, with an increase in EF to 49%, decrease in EDVI to 45 mL/m^2, LAVI remained the same (68 ml/m^2), PASP decreased to 29 mmHg. Diastolic function parameters exhibited improvement, with *VE/VA* = 1.1 and *VE/Em* ratio (mean) = 10.3 (grade 1 diastolic dysfunction). Longitudinal deformation of the LV myocardium was determined (in the 2D-strain mode). It was significantly impaired, with a value of –15% (which is below the normal range of –18%). Maximum myocardial deformation disturbances were observed in the hypertrophied IVS (the zone is colored pale pink in Fig. 2).

Table 1. Dynamics of ejection fraction for 2008–2021
Таблица 1. Динамика фракции выброса за 2008–2021 годы

Year	2008	2009	2010	2014	2015	2018	2020	2021
EF, %	68	64	61	60	59	59	51	40

Note: ФВ — ejection fraction.
Примечание: ФВ — фракция выброса.

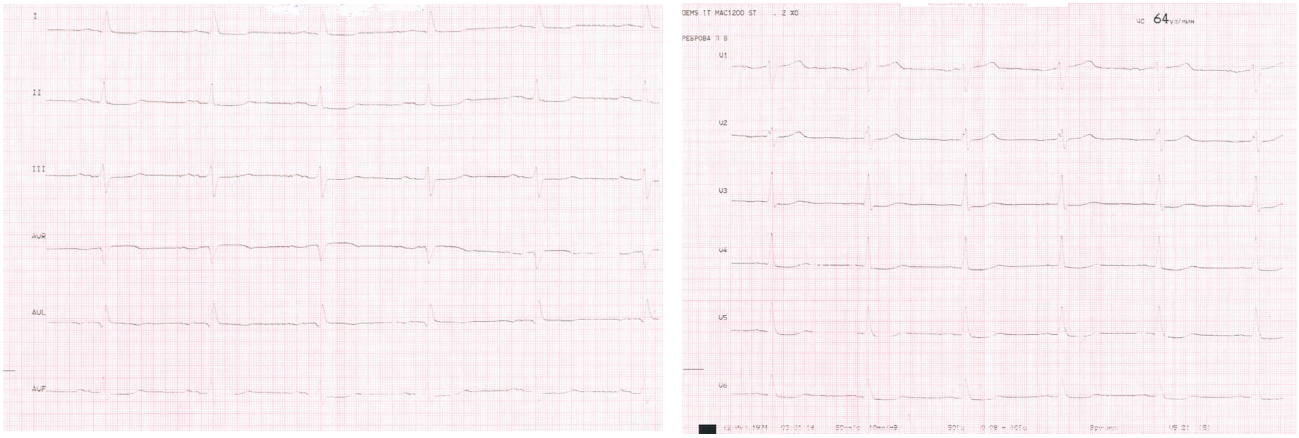

Fig. 1. Patient's ECG in May 2022 (recording speed is 50 mm/s)
Рис. 1. Электрокардиограмма пациента в мае 2022 года (скорость записи 50 мм/с)

In consideration of the notable LA dilatation, LA function was evaluated in the longitudinal strain mode (2D strain) (Fig. 3). As in the case of longitudinal LV myocardial strain, LA myocardial strain parameters were abnormal. The deformation index during the reservoir phase was equal to 13% (with a mean normal value of 39%, 95% confidence interval [CI] 38%–41%). During the conduit phase, it was 5% (with a mean normal value of 23%, 95% CI 21%–25%), whereas during the contractile phase, it was 8% (with a mean normal value of 17%, 95% CI 16%–19%) [6]. Notably, the strain indices during the conduit and contractile phases were negative, as the LA myocardium shortens during these phases. For the convenient comparison of indices, it is customary to discard the minus sign. In our patient, the LA function during all three phases was significantly impaired.

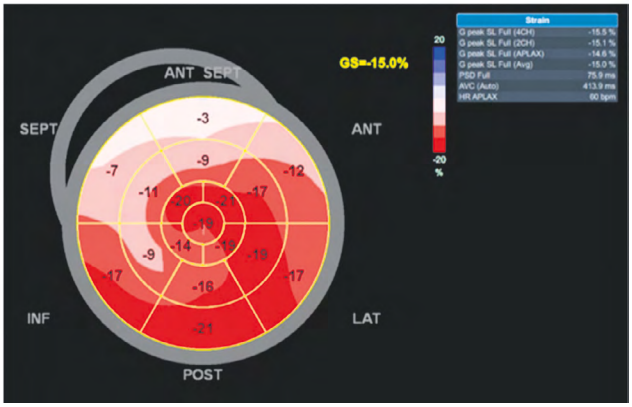

Fig. 2. Bull's eye format of peak global longitudinal strain of the left ventricular myocardium. Explanation in the text
Рис. 2. Пиковая глобальная деформация миокарда ЛЖ в формате «бычий глаз». Объяснение в тексте

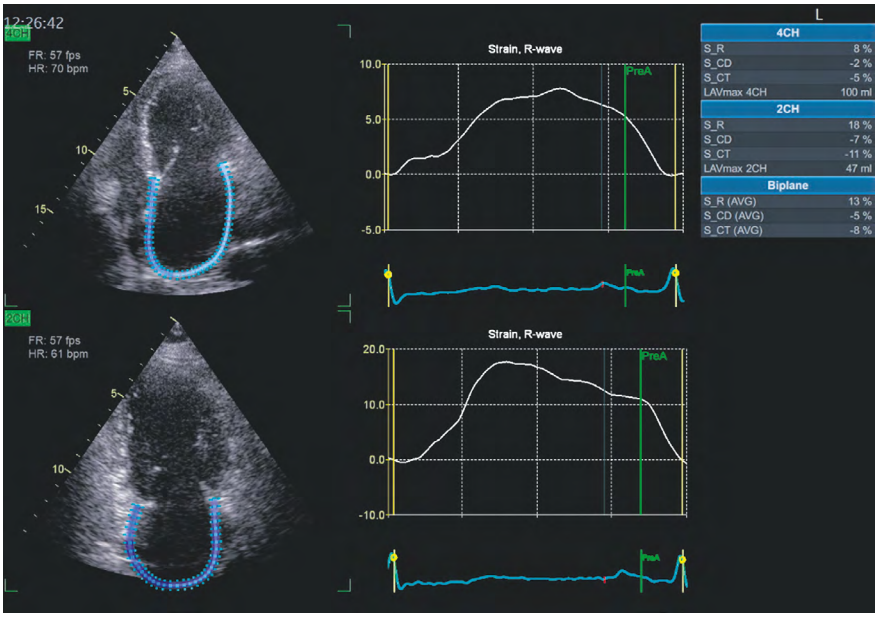

Fig. 3. Assessment of left atrial function in 2022 in 2D-Strain mode. Explanation in the text
Рис. 3. Оценка функции левого предсердия в 2022 году в режиме деформации. Объяснение в тексте

Because the assessment of LA myocardial strain was performed for the first time, it was not possible to ascertain the extent to which LA function had been disturbed previously.

During Holter ECG monitoring, 1,337 (1.5%) PVGCs, 22 runs of non-sustaned VT, and 215 (0.2%) atrial premature complexes were recorded.

Continued therapy was necessary. In September 2023, the patient presented to our facility for the recurrence of chest discomfort during physical exertion exceeding household loads. Stress EchoCG testing was performed repeatedly, with negative results. In accordance with the ESC Guidelines for the Management of Cardiomyopathies, ranolazine 500 mg twice a day [1, 3] was prescribed to control the pain syndrome. The chest discomfort was successfully managed during the medication therapy.

A control EchoCG was performed in February 2024. For other parameters the following results were obtained: EDVI (48 mL/m^2), LAVI (63 mL/m^2), PASP (25 mmHg), *VE/VA* = 1.1, and *VE/Em* (mean) = 10.0 were noted. The dynamics of key EchoCG indices are presented in Table 2. Unfortunately, the global longitudinal LV myocardial strain did not improve, remaining at the level observed in 2022.

In addition to LV diastolic function improvement, the LA volume tended to decrease (Fig. 4). Indices of LA function have improved. Consequently, the proportion of LA myocardial strain increased from 13% to 17% during the reservoir phase, from 5% to 8% during the conduit phase, and from 8% to 9% during the contractile phase. The dynamics of the indices are presented in Table 3.

During Holter ECG monitoring, 73 (0.1%) PVCs, 1 run of non-sustained VT (Fig. 5), 792 (1.0%) atrial premature complexes, and 1 run of non-sustained atrial tachycardia were recorded. The dynamics of the number of ventricular and atrial rhythm disturbances are presented in Table 4.

In this case, the therapy should be continued in accordance with national and international recommendations for the treatment of chronic heart failure, i.e., the therapy, which demonstrated a positive outcome, should be continued

Table 2. Dynamics of key echocardiography parameters during quadruple therapy
Таблица 2. Динамика ключевых эхокардиографических показателей на фоне квадротерапии

Indices	Before quadruple therapy	During quadruple therapy	
	February 2021 года	May 2022 года	February 2024 года
EF, %	40	49	55
EDVI, mL/m^2	75	45	48
LAVI, mL/m^2	68	68	63
PASP, mmHg	52	29	25
VE/VA	2.1	1.1	1.1
VE/Em	16.3	10.3	10.0

Note: ФВ — ejection fraction; ИКДО — end-diastolic volume index; ИОЛП — left atrium volume index; СДЛА — pulmonary artery systolic pressure; *VE* — transmitral blood flow velocity during the rapid filling phase; *VA* — transmitral blood flow velocity at the moment of left atrium systole; *Em* — the average value of the sum of the speeds of movement of the septal and lateral segments of the mitral valve anulus.

Примечание: ФВ — фракция выброса; ИКДО — индекс конечно-диастолического объема; ИОЛП — индекс объема левого предсердия; СДЛА — систолическое давление в легочной артерии; *VE* — скорость трансмитрального кровотока в фазу быстрого наполнения; *VA* — скорость трансмитрального кровотока в момент систолы левого предсердия; *Em* — среднее значение суммы скоростей движения перегородочного и бокового сегментов кольца митрального клапана.

Table 3. Dynamics of indicators of left atrium function during quadruple therapy
Таблица 3. Динамика показателей функции левого предсердия на фоне квадротерапии

Indices	May 2022	February 2024	Normal, mean value, (95% CI) [6]
LASr, %	13	17	39 % (95% CI 38%–41%)
LAScd, %	5	8	23 % (95% CI 21%–25%)
LASct, %	8	9	17 % (95% CI 16%–19%)

Note: ДИ — confidence interval; ЛП — left atrium; *LASr* — left atrium reservoir strain; *LAScd* — left atrium conduit strain; *LASct* — left atrium contractile strain.

Примечание: ДИ — доверительный интервал; ЛП — левое предсердие; *LASr* — деформация левого предсердия во время резервуарной фазы; *LAScd* — деформация левого предсердия во время кондуитной фазы;, *LASct* — деформация левого предсердия во время сократительной фазы

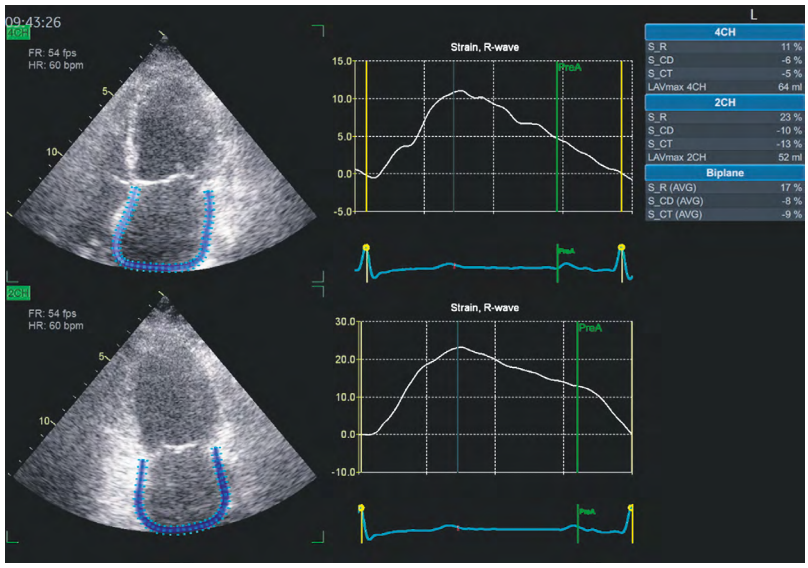

Fig. 4. Assessment of left atrial function over time in February 2024 in 2D-Strain mode. Explanation in the text
Рис. 4. Оценка функции левого предсердия в динамике в феврале 2024 года в режиме деформации. Объяснение в тексте

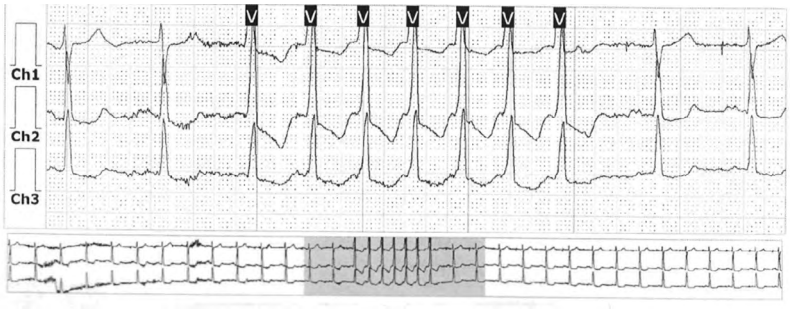

Fig. 5. Fragment of ECG monitoring. Episode of non-sustained ventricular tachycardia
Рис. 5. Фрагмент суточного мониторирования ЭКГ. Эпизод неустойчивой желудочковой тахикардии

Table 4. Dynamics of indicators of ECG monitoring for 2021–2024
Таблица 4. Динамика показателей суточного мониторирования ЭКГ за 2021–2024 годы

Indices	February 2021	May 2022	October 2022	June 2023	February 2024
Number premature ventricular complexes, *n* (%)	5698 (4.4)	1337 (1.5)	520 (0.6)	119 (0.1)	73 (0.1)
Number premature atrial complexes, *n* (%)	9341 (7.3)	215 (0.2)	1380 (1.6)	1833 (2.1)	792 (1.0)
Number of на non-sustained ventricular tachycardia	7	2	1	0	1
Number of non-sustained atrial tachycardia	9	0	0	1	1

to prevent any deterioration in the case of the withdrawal of any [7, 8].

DISCUSSION

Most HCMP cases have genetic causes. The identification of typical genetic mutations facilitates the diagnosis of HCMP. The disease is most often the result of mutations in genes encoding sarcomeric proteins, with mutations in β-myosin heavy chain (MYH7), myosin regulatory light chain (MYL2), essential myosin light chain (MYL3), myosin-binding cardiac protein C (MYBPC3), troponins (TNNI3 and TNNT2), and other proteins accounting for 40%–60% of cases [1, 3, 9].

In the present case, one of the two identified mutations (TNNT2) is typical and relatively common in HCM, whereas the second mutation is more characteristic of dilated CMP (TTN).

The HCMP is diagnosed based on the presence of LV wall thickening, which is extremely rare on the right ventricle. This thickening cannot be explained by increased hemodynamic load, which includes arterial hypertension and valvular heart diseases. In the proband, the quantitative criterion of myocardial thickness of ⩾15 mm is considered diagnostic [1, 3, 9]. Since 2002 until 2021 the patient exhibited the classic phenotype of nonobstructive HCMP, which is characterized by pain syndrome and various rhythm disturbances. HCMP-related heart failure is more often associated with diastolic dysfunction, primarily in the early disease stages. In some patients (5%–8% of those suffering from HCMP), a systolic component may be added due to the burnout phenomenon, which is characterized by a decrease in LV EF ⩽50%, LV wall thinning, and LV cavity dilation [3, 4]. The term "burnout HCMP" has been proposed to distinguish this phenotype, which is novel for the patient and emerged during disease progression, from the phenotype at the time of diagnosis [4]. Currently, no clear criteria can be used to predict the transition from the hypertrophic stage to the burnout phase. However, several potential factors have been identified, including certain mutations of genes encoding the synthesis of sarcomere proteins, family history of a terminal dilated stage of HCMP, AF, and degree of late signal enhancement by gadolinium, which reflects the severity of fibrosis on cardiac MRI [3, 4]. EF is not an optimal method for the early detection of the burnout phenomenon. Changes of myocardial longitudinal strain over time allows an unfavorable prognosis assumed. In recent years, echocardiographic assessment of myocardial longitudinal strain has become a widely used diagnostic tool. D.M. Adamczak et al. revealed an association between myocardial longitudinal strain and burnout [10]. Longitudinal LV myocardial strain was assesed in the patient at the stage of EF restoration in 2022 and 2024. It appeared to be reduced and did not improve significantly in 2024 compared with 2022, whereas EF increased from 49% to 55%. Unfortunately, this parameter was not determined before 2022.

Our patient experienced a transformation of hypertrophic phenotype into dilated phenotype due to several reasons. These include multiple mutations (one of which may lead to both hypertrophic and dilatational phenotypes), cardiac rhythm disorders, including AF, and, possibly, longitudinal deformation disorders of the LV myocardium.

Before the introduction of quadruple therapy in clinical practice, the burnout phase was assumed to represent a terminal stage of HCMP, and patients entering this phase are deemed potential candidates for heart transplantation. Our example illustrates the reversibility of changes with the timely initiation of quadruple therapy involving angiotensin II receptor type 1 and neprilysin ingibitor with sodium-glucose cotransporter type 2 inhibitor, mineralocorticoid receptor antagonist, and β-adrenoblocker. Currently, mavacamten, a selective allosteric inhibitor of cardiac myosin adenosine triphosphatase, for HCMP treatment is registered in some countries [1, 3]. However, no information is available about the effect of mavacamten on the burnout phenomenon.

In patients undergoing quadruple therapy, both systolic and diastolic indices significantly improved. This was accompanied by a tendency for LA myocardial deformation parameters to increase, reflecting an improvement in the state of the LA function.

A separate discussion is warranted regarding cardiac rhythm disorders. The most prevalent rhythm disturbance in HCMP is AF, which occurs following excessive hemodynamic load, leading to LA dilatation.

The treatment and prophylaxis of AF in HCMP are initiated according to the general principles of the recommendations for AF diagnosis and treatment, which may include certain adjustments because of the specifics of the underlying disease. Considering the high incidence of stroke in the setting of AF in HCMP, anticoagulants are recommended regardless of the presence or absence of risk scores for ischemic stroke and systemic embolism [1]. Both warfarin and direct oral anticoagulants (apixaban, dabigatran, and rivaroxaban) are used. Sinus rhythm is preferred to AF in patients with HCMP; therefore, all options should be used to maintain sinus rhythm, including pulmonary vein isolation, which was attempted in this patient with a positive effect. Amiodarone is the optimal drug for sinus rhythm control. However, in the described case, owing to the rapid development of cordarone-induced thyrotoxicosis further therapy with this drug is impossible.

Non-sustained and sustained VT is a common finding in patients with HCMP. Owing to the high risk of SCD in this pathology, risk stratification and determination of indications for ICD placement are essential [1, 3, 11, 12]. In this patient, the sustained VT accompanied by hemodynamic abnormalities necessitated ICD insertion in 2007 and replacements in 2012 and 2020 because of battery depletion. Before each replacement, the risk of SCD must be reevaluated because it may decrease over time.

CONCLUSIONS

The HCMP course in each patient is highly variable, ranging from asymptomatic to severe progressive symptoms,

including Burned-out phase of HCMP, and premature death. In the context of regular follow-up, the physician should promptly identify markers and signs of an unfavorable prognosis and initiate therapy to improve the prognosis. In the presence of risk factors for SCD, ICD implantation is indicated. In patients with signs of burned-out phase and a decreased LV EF, therapies for chronic heart failure, including quadruple therapy, may be beneficial.

ADDITIONAL INFORMATION

Consent for publication. Written consent was obtained from the patient for publication of relevant medical information and all of accompanying images within the manuscript.

Author contribution. Thereby, all authors confirm that their authorship complies with the international ICMJE criteria (all authors have made a significant contribution to the development of the concept, research, and preparation of the article, as well as read and approved the final version before its publication).

Personal contribution of the authors: T.N. Novikova — concept, design, collection and processing of materials, data analysis, text writing, literature review; A.E. Andreeva — collection and processing of echocardiography data; F.I. Bitakova — collection and analysis of daily monitoring data; V.I. Novikov — cprocessing and analysis of echocardiography data, literature review; K.A. Gladysheva — literature review; P.V. Petrova — processing of materials; P.A. Stalnova, N.A. Tokareva — collection of material.

Competing interests. The authors declare that they have no competing interests.

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Вклад авторов. Все авторы внесли существенный вклад в разработку концепции, проведение исследования и подготовку статьи, прочли и одобрили финальную версию перед публикацией.

Вклад каждого автора. Т.Н. Новикова — концепция, дизайн, сбор и обработка материалов, анализ данных, написание текста, обзор литературы; А.Е. Андреева — сбор и обработка ЭхоКГ данных; Ф.И. Битакова — сбор и анализ данных суточного мониторирования; К.А. Гладышева — обзор литературы; В.И. Новиков — обработка и анализ ЭХОКГ данных, обзор литературы; П.В. Петрова — обработка материалов; П.А. Стальнова, Н.А. Токарева — сбор материала.

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

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

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