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Hypertrophic cardiomyopathy in asymptomatic 24-year-old pregnant woman: management according to ESC guidelines

Kardiomiopatia przerostowa u bezobjawowej 24-letniej kobiety w ciąży — postępowanie według wytycznych ESC

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Abstract

We present a case of 24-year-old asymptomatic pregnant woman in 18 hbd with hypertrophic cardiomyopathy (HCM). An echocardiogram revealed the hypertrophy of all walls of the left ventricle (LV), except for the posterolateral wall, from 21 mm to 31 mm and septal hypertrophy up to 36 mm. During the first 48-h electrocardiogram (ECG) monitoring, five episodes of slow ventricular tachycardia (VT) consisted of three ExV up to 108/min were recorded. The 5-year HCM sudden cardiac death (SCD) risk score revealed the low risk of 2.25% — implantable cardioverter-defibrillator (ICD) not indicated. After a C-section delivery in 37 hbd, the control echocardiography revealed the enlargement of the LV wall hypertrophy up to 38 mm. In the 48-h ECG monitoring, two episodes of asymptomatic non-sustained VT consisted of four and seven ExV up to 162/min were registered. The 5-year HCM SCD risk came up to the intermediate level of 5.91% (ICD may be considered, class IIb B of recommendations). Based on the clinical and echocardiographic findings with dynamic progress in the LV hypertrophy, exacerbation of ventricular arrhythmias and increase of N-terminal natriuretic propeptide type B, the ICD was implanted. As presented by Maron & Maron at the European Society of Cardiology Congress in London 2015, magnetic resonance scanning with the late gadolinium enhancement (LGE) estimation may be helpful in making the decision on the ICD implantation, especially within the group of intermediate 5-year risk of SCD (4–6%) with massive LV hypertrophy. The Authors suggest the extensive LGE (≥ 15%) as a primary SCD risk factor as well as a potential risk factor when conventional evaluation of the ICD implantation indications is ambiguous.

Key words: hyperthrophic cardiomyopathy, late gadolinium enhancement, ESC guidelines, implantable cardioverter-defibrillator

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Introduction

Hypertrophic cardiomyopathy (HCM) is a primary disease of myocardium that occurs in the increased left ventricular (LV) wall thickness with myocardial fibrosis. Most patients suffering from HCM have an elevated risk of sudden cardiac death (SCD). Thus, the precise evaluation of implantable cardioverter-defibrillator (ICD) implantation indications with estimation of the 5-year HCM SCD risk is necessary.

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Case report

A 24-year-old asymptomatic pregnant woman in 18 hbd was admitted due to LV hypertrophy registered accidentally during the pregnancy ultrasonography (USG).

The patient presented with no abnormalities in physical examination, denied dyspnoea, chest pain, syncope, heart palpitations and the occurrence of HCM or SCD in relatives. In the resting electrocardiogram (ECG), non-specific anomalies of QRS-complex and ST-T were found. The echocardiogram revealed hypertrophic cardiomyopathy without left ventricular outflow tract obstruction (LVOTO) or mid-cavity obstruction also during the Valsalva manoeuvre. Except for the posterolateral wall, the hypertrophy of LV from 21 mm to 31 mm and septal hypertrophy up to 36 mm were registered (Figure 1). During the first ECG monitoring, five episodes of slow ventricular tachycardia (VT) consisted of three ExV up to 108/min were recorded. The level of N-terminal natriuretic propeptide type B (NT-proBNP) was 4606 pg/mL. The 5-year HCM SCD risk score revealed the low risk of 2.25% (ICD not indicated, class IIIB). The beta-blocker was administered.

In a third day after a C-section delivery in 37 hbd, the echocardiography revealed the enlargement of LV hypertrophy up to 38 mm. In the 48-h ECG monitoring, two episodes of asymptomatic non-sustained ventricular tachycardia (nsVT) consisted of four and seven ExV up to 162/min were registered (Figure 2). An increase of the

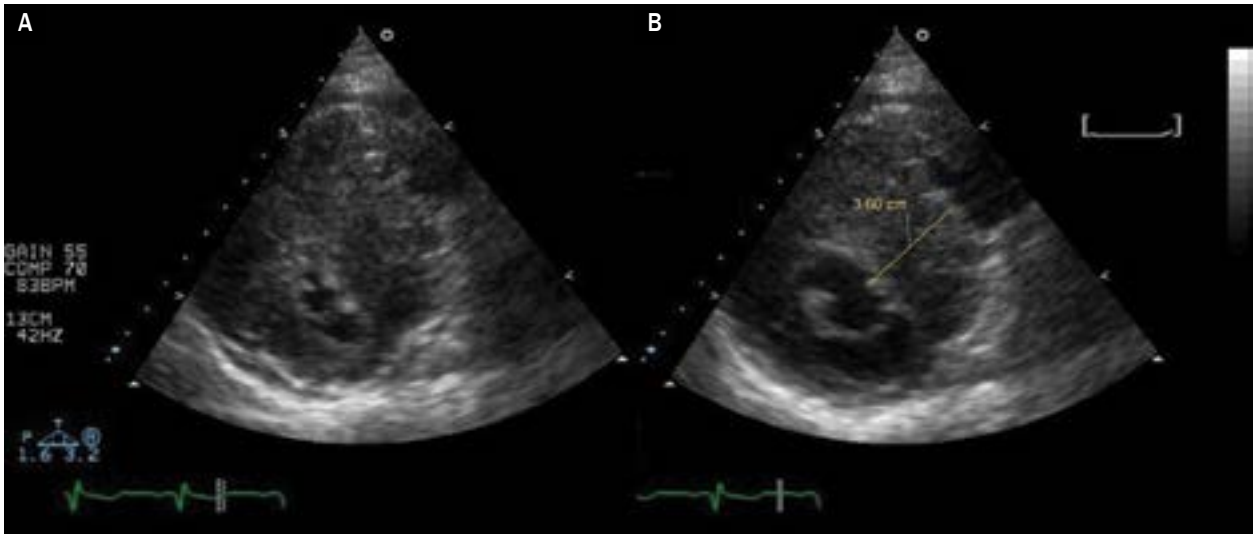

Figure 1A, B. Echocardiographic view

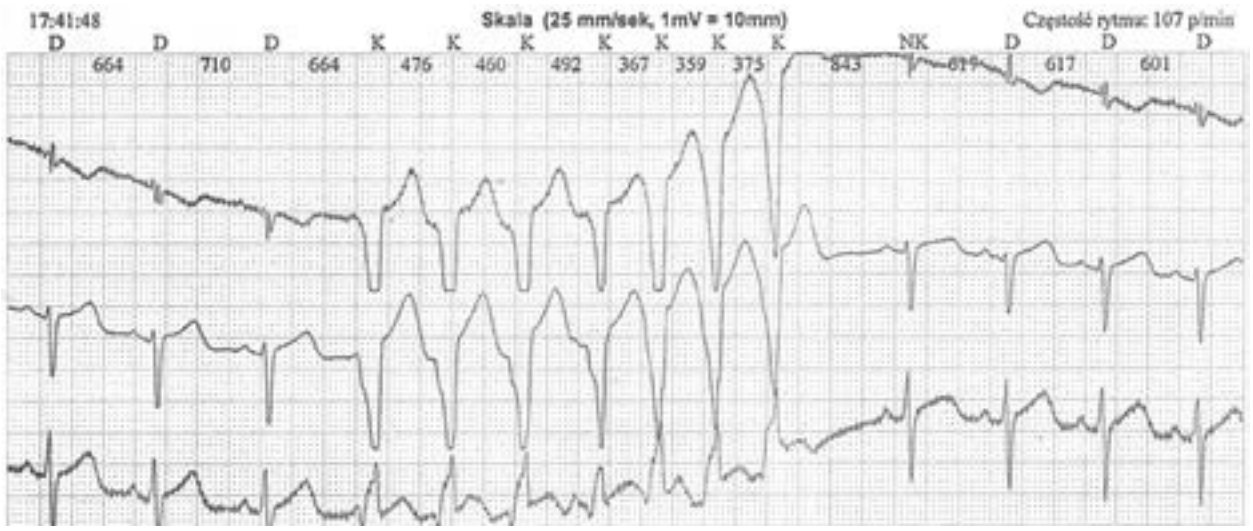

Figure 2. Non-sustained ventricular tachycardia in 48-h electrocardiogram monitoring

NT-proBNP to 7663 pg/mL was found. The 5-year HCM SCD risk came up to the intermediate level of 5.91% (ICD may be considered, class IIb B of recommendations). Based on the dynamic progress in the LV hypertrophy, the exacerbation of ventricular arrhythmias and the increase of NT-proBNP, the ICD was implanted.

Discussion

According to the European Society of Cardiology (ESC) guidelines, HCM (2014) [1] is a primary disease, that occurs in the increased left ventricular (LV) wall thickness with myocardial fibrosis. HCM by definition can be diagnosed when a left ventricle wall thickness ≥ 15 mm in at least one LV myocardial segments, measured by any imaging technique, while it cannot be explained by abnormal LV loading conditions. The decision of an ICD implantation as a primary prophylaxis of SCD is a very important part of treatment strategy in HCM. According to the newest ESC guidelines (2014), the main method of estimating the indications for ICD is the 5-year HCM risk-SCD. The risk of SCD should be assessed in every patient with HCM, when the disease is diagnosed and re-evaluated every 1–2 years or in case of changes in the clinical status (class I B of recommendations) [1].

As reported by Maron et al. [2], in the group of 239 HCM patients, massive LV hypertrophy was more often at the age of 18–39 than over 75 years (p = 0.03). In another research Maron et al. [3] report SCD in 44 cases from 744 HCM patients, while in the subgroup with LV hypertrophy > 30 mm, SCD occurred in 50% (9/18) of the patients. In the research by Spirito et al. [4] on the group of 480 HCM patients, from 43 with LV hypertrophy > 30 mm, over 95% were asymptomatic or with poor symptoms and the risk of SCD was directly related to LV hypertrophy (p = 0.001). Elliot et al. [5] reported that from 630 HCM patients, SCD occurred in 39 cases and 10 of them had LV hypertrophy > 30 mm. The patients with LV hypertrophy over 30 mm had higher risk of the SCD (p = 0.049). Summarising, young patients with massive LV hypertrophy are especially endangered with SCD. Still, the presented patient is in the IIa class of recommendations for an ICD implantation [1].

In patients with the intermediate 5-year HCM SCD risk (4–6%), as presented by Maron & Maron at the ESC Congress in London 2015 [6], magnetic resonance imaging (MRI) scanning with the late gadolinium enhancement (LGE) estimation may be helpful in making a decision on the ICD implantation. The LGE reflects the focal fibrosis of myocardium, increasing the risk of severe arrhythmias and SCD. Maron & Maron suggest the extensive LGE (≥ 15%) as a primary SCD risk factor as well as a potential risk factor when the conventional evaluation of the ICD implantation indications is ambiguous (Figure 3).

Described case highlights the role of the 5-year HCM SCD risk score and MRI scanning to guide treatment strategy in HCM. The question is, if the subcutaneous ICD (S-ICD) might be a better option in this case according to 2015 ESC guidelines (class IIa C) [7]. In such cases, the main cause of the ICD implantation is the primary prevention of SCD without the potential need of heart pacing, while an S-ICD implantation is less invasive. Also the risk of endocarditis and thromboembolic complications is lower and the risk

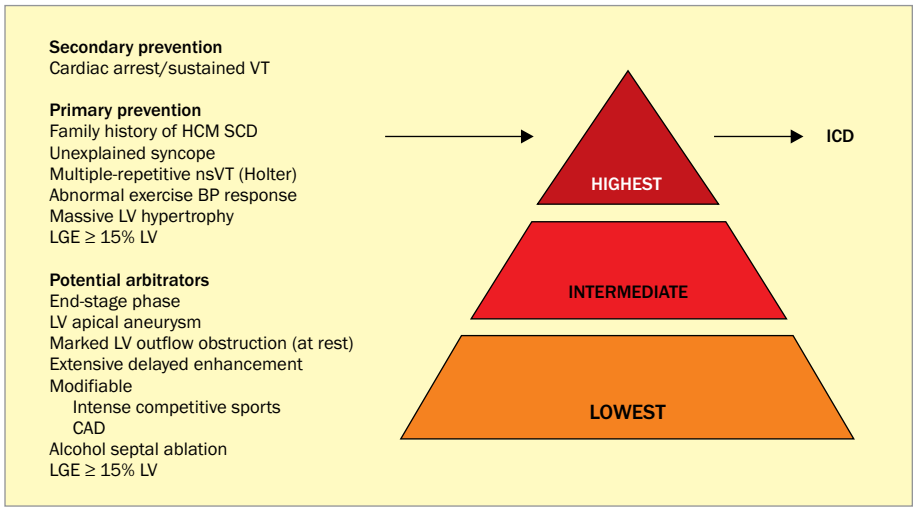

Figure 3. Risk stratification model for an implantable cardioverter-defibrillator (ICD) implantation as presented by Maron & Maron at the European Society of Cardiology Congress, London 2015 [6]; VT — ventricular tachycardia; HCM — hypertrophic cardiomyopathy; SCD — sudden cardiac death; nsVT — non-sustained ventricular tachycardia; BP — blood pressure; LV — left ventricle; LGE — late-gadolinium enhancement; CAD — coronary artery disease

of vessels fibrosis caused by ICD electrodes is minimalised, what simplifies the potential heart transplantation. Finally, better cosmetic effect may be important especially for young women.

Conclusions

According to the ESC guidelines, evaluation of the 5-year SCD risk in HCM is one of the basic tools in qualification to ICD implantation. Estimation of the LGE in MRI scanning may be helpful in making a decision on the ICD implantation in HCM patients, especially within the group of the intermediate 5-year risk of SCD (4–6%) with massive LV hypertrophy.

Conflict of interest(s)

Authors declare no conflict of interests to this research.

Streszczenie

Zaprezentowano opis przypadku 24-letniej kobiety z kardiomiopatią przerostową bez objawów w 18. tygodniu ciąży. W echokardiografii przezklatkowej (TTE) uwidoczniono przerost wszystkich ścian lewej komory (LV) poza tylno-boczną, od 21 mm do 31 mm, oraz przerost przegrody międzykomorowej do 36 mm. W 48-godzinnym badaniu elektrokardiograficznym (EKG) metodą Holtera stwierdzono 5 epizodów częstoskurczu komorowego złożonych z 3 pobudzeń o częstości do 108/min. Wyliczono 5-letnie ryzyko nagłego zgonu sercowego (SCD) jako niskie — na poziomie 2,25% (wszczepialny kardiowerter-defibrylator [ICD] nie jest zalecany). Po porodzie drogą cięcia cesarskiego w 37. tygodniu ciąży w TTE uwidoczniono progresję przerostu mięśnia LV do 38 mm. W 48-godzinnym badaniu EKG metodą Holtera stwierdzono 2 epizody bezobjawowego nieutrwalonego częstoskurczu komorowego z 4 oraz 7 pubudzeń o częstości do 162/min. Wyliczone 5-letnie ryzyko SCD zwiększyło się do 5,91% (klasa zaleceń IIb B dla ICD). Ze względu na całość obrazu klinicznego i echokardiograficznego, progresję przerostu ścian LV, nasilenie arytmii komorowej i zwiększenie stężenia N-końcowego propeptydu natriuretycznego typu B pacjentce wszczepiono ICD. U pacjentów z pośrednim 5-letnim ryzykiem SCD w kardiomiopatii przerostowej (4–6%), zgodnie z doniesieniem Maron i Maron z kongresu Europejskiego Towarzystwa Kardiologicznego (Londyn, 2015), ocena późnego wzmocnienia kontrastowego (LGE) w badaniu rezonansu magnetycznego może być pomocna w podjęciu decyzji o implantacji ICD. Autorzy wskazują na LGE większe lub równe 15% jako czynnik ryzyka SCD kwalifikujący do implantacji ICD, a także czynnik rozstrzygający w przypadku niejednoznacznych wskazań do wszczepienia ICD.

Słowa kluczowe: kardiomiopatia przerostowa, wytyczne ESC, wszczepialny kardiowerter-defibrylator, podskórny kardiowerter-defibrylator

Folia Cardiologica 2018; 13, 1: 55–58

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