CLINICAL IMAGE

Advanced hypertrophic obstructive cardiomyopathy diagnosed in early pregnancy: successful prevention of sudden cardiac death

Janusz Kudlicki[1], Agata Frania-Baryluk[1], Anna Kania[1], Andrzej Tomaszewski[1], Andrzej Wysokiński[1], Elżbieta Czekajska-Chehab[2]

1 Medical University of Lublin, Cardiology Department, Lublin, Poland
2 Medical University of Lublin, Department of Radiology and Nuclear Medicine, Lublin, Poland

A 30-year-old woman in the 14th week of her 2nd pregnancy, was admitted to the intensive cardiac care unit in 2006 with an episode of monomorphic nonsustained ventricular tachycardia (FIGURE 1A). Her mother died suddenly at the age of 26. The first pregnancy was uncomplicated and the patient gave birth to a healthy daughter by natural labor. An electrocardiogram showed left ventricular hypertrophy. Transthoracic echocardiography showed increased left ventricular wall thickness with left ventricular outflow tract (LVOT) obstruction, with a maximum outflow gradient of approximately 200 mm Hg, velocity flow of approximately 6 to 7 m/s, systolic anterior motion of the mitral valve (FIGURE 1B–1E), and moderate mitral valve regurgitation. The patient was scheduled for implantable cardioverter-defibrillator (ICD) implantation for primary prevention of sudden cardiac death (SCD) according to the 2003 American College of Cardiology and European Society of Cardiology (ESC) guidelines on hypertrophic cardiomyopathy (HCM; 2 risk factors present: ventricular tachycardia and family history). The procedure was postponed until the postpartum period.

In the 38th week of pregnancy, an elective Cesarean section was performed for cardiovascular indications. The patient gave birth to a healthy daughter weighing 2550 g, with an Apgar score of 10 points. In the postpartum period, a dual chamber ICD was implanted (FIGURE 1F). One month after implantation, outside the hospital, the patient had an episode of cardiac arrest caused

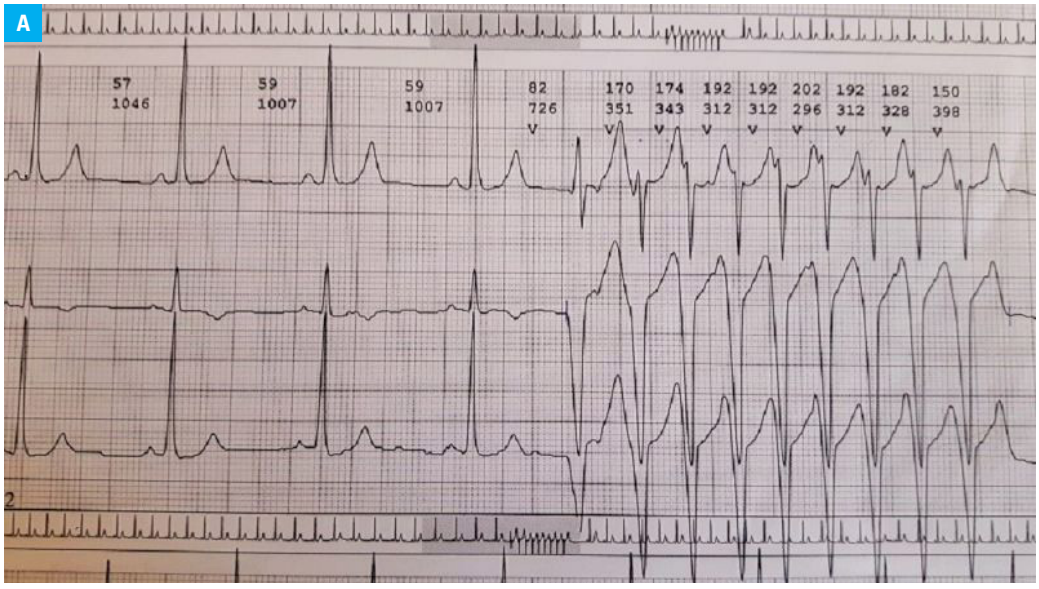

FIGURE 1 A – nonsustained ventricular tachycardia in the 15th week of pregnancy

Correspondence to:
Agata Frania- Baryluk, MD, Klinika Kardiologii, Samodzielny Publiczny Szpital Kliniczny Nr 4, ul. Jaczewskiego 8, 20-954 Lublin, Poland, phone: +48 81 724 41 51, email: a.frania@poczta.fm
Received: February 18, 2018.
Accepted: April 15, 2018.
Published online: April 15, 2018.
Conflict of interest: none declared.
Pol Arch Intern Med. 2018; 128 (4): 256-258
doi:10.20452/pamw.4245

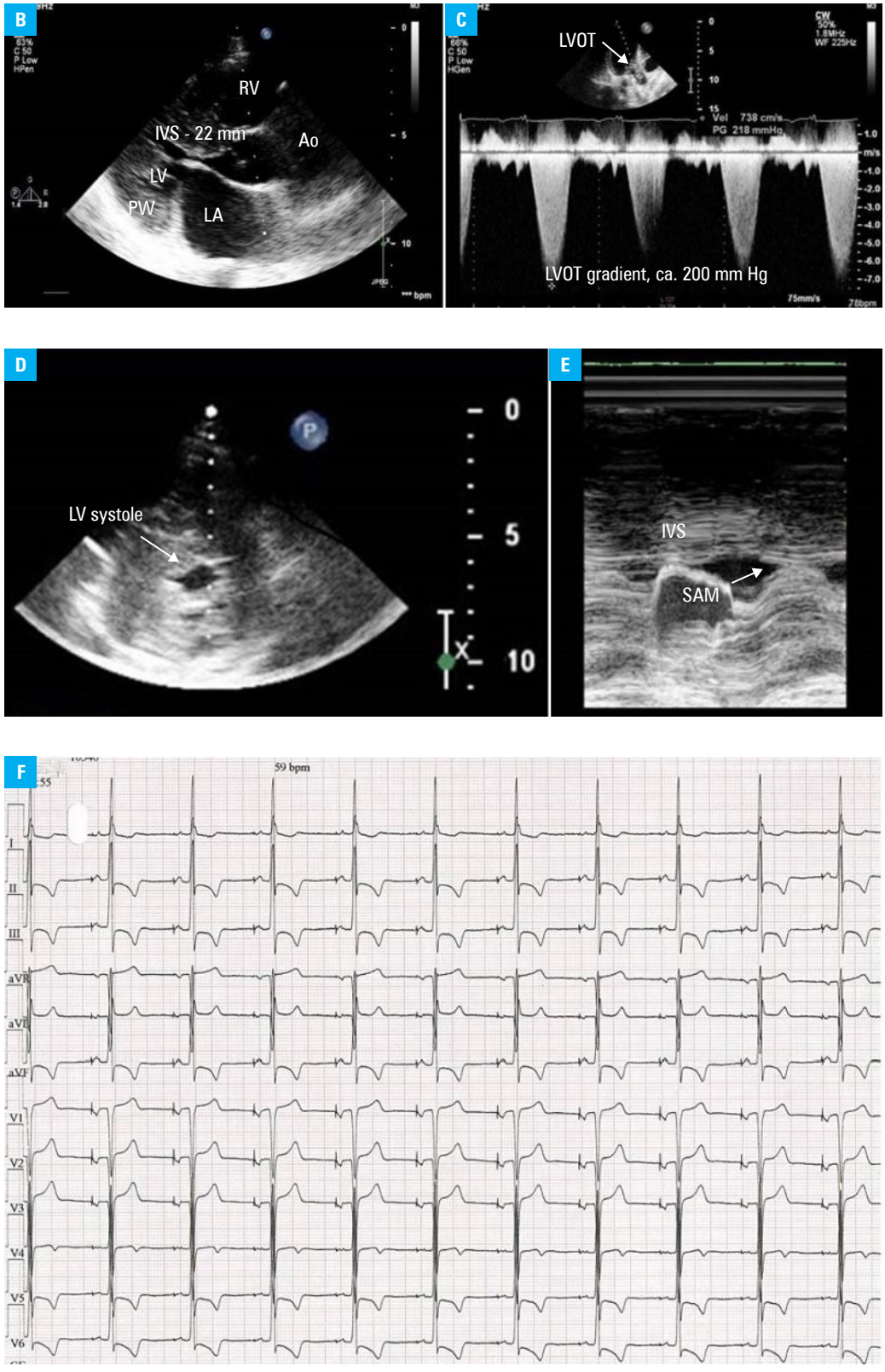

FIGURE 1
B – echocardiogram, parasternal long-axis view, massive concentric LVH; **C** – echocardiogram, continuous-wave Doppler: high maximum gradient and flow velocity in the LVOT; **D–E** – echocardiogram, parasternal short-axis view and M-mode: massive concentric LVH and systolic anterior motion of the mitral valve (arrow); **F** – electrocardiogram after implantable cardioverter-defibrillator implantation, AAI pacing, LVH and overload
Abbreviations: see on the next page

by ventricular fibrillation, with adequate ICD intervention.

Owing to inadequate coronary artery anatomy for alcohol septal ablation, septal myectomy was performed (FIGURE 1G–1I). The patient did not present any syncopal episodes in a 7-year follow-up. She received calcium channel blockers and β-blockers. On follow-up echocardiography, the maximum and mean outflow gradients were 11.5 mm Hg and 6.29 mm Hg, respectively (FIGURE 1J).

Only a limited number of pregnancies in women with HCM have been reported.[1-3] Pregnancy is contraindicated only in women with significant impairment of systolic function (LVEF <40%) or severe symptomatic LVOT obstruction, and it may be possible after relief of obstruction by surgery or alcohol septal ablation.[1] Nevertheless, pregnant women in World Health Organization (WHO) class II should be assessed every trimester, while those in WHO class III should

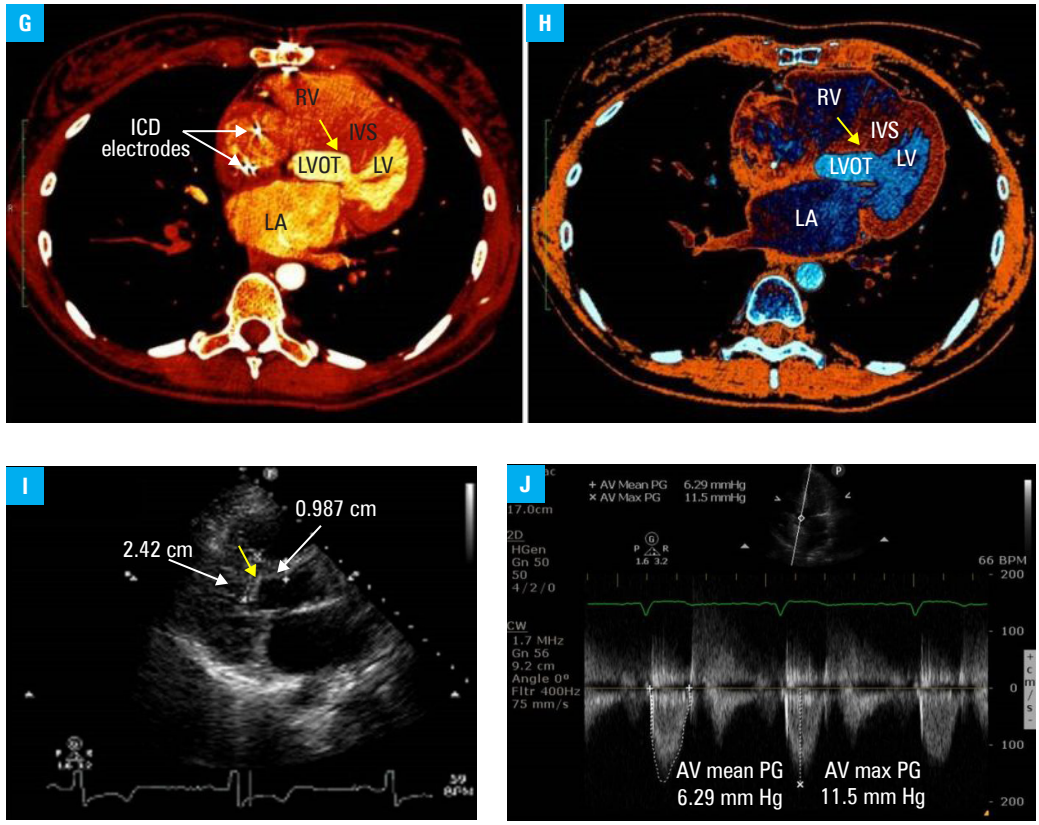

FIGURE 1 G–H – computed tomography scans of the heart performed after myectomy, resected part of the IVS (yellow arrow); I – echocardiogram after myectomy, parasternal long-axis view, resected part of the IVS (yellow arrow); J – echocardiogram after myectomy, continuous-wave Doppler: significantly reduced maximum and mean LVOT gradients, aortic regurgitation

Abbreviations: Ao, aorta; AR, aortic regurgitation; AV, aortic valve; ICD, implantable cardioverter-defibrillator; IVS, interventricular septum; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; PG, pressure gradient; PW, posterior wall; RV, right ventricle; SAM, systolic anterior motion

be followed monthly or bimonthly by a multidisciplinary team in specialized centers.[2] Cesarean section is preferred in women with severe LVOT obstruction, severe heart failure, or preterm labor while on oral anticoagulant treatment.

Patients with HCM are at increased risk of SCD. Therefore, pregnant women with HCM and history of SCD in a close family member require greater attention and careful therapeutic planning. The decision on ICD implantation and its timing should be made as soon as possible. An alternative option is a LifeVest wearable defibrillator as the bridging therapy before ICD implantation. There are optimistic data about the efficacy of this method[4]; however, there are no data for pregnant women. Considering the retrospectively estimated probability of SCD at 5 years (based on the HCM Risk-SCD calculator from 2014 ESC Clinical Practice Guidelines), our patient would be classified as high-risk (27%), which confirms the efficacy of ICD implantation.[5] In summary, septal myectomy is an effective method of decreasing the LVOT gradient.

ACKNOWLEDGMENTS The authors would like to congratulate the surgical team of the Cardiac Surgery Department, Clinical Hospital in Poznan, for completing a successful myectomy.

REFERENCES

1 Autore C, Conte MR, Piccinno M, et al. Risk associated with pregnancy in hypertrophic cardiomyopathy. J Am Coll Cardiol. 2002; 40: 1864-1869.

2 Krul SPJ, Smagt JJ, van den Berg MP, et al. Systematic review of pregnancy in women with inherited cardiomyopathies. Eur J Heart Fail. 2011; 13: 584-594.

3 Pieper PG, Walker F. Pregnancy in women with hypertrophic cardiomyopathy. Neth Heart J. 2013; 21: 14-18.

4 Chudzik M, Szyda Ł, Urbanek B, et al. The LifeVest wearable defibrillator for noninvasive prevention of sudden arrhythmic death syndrome. The first Polish case of an adequate LifeVest intervention. Pol Arch Intern Med. 2017; 127: 641-642.

5 Elliott PM, Anastasakis A, Borger MA, et al. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy: The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). Eur Heart J. 2014; 35: 2763-2767.