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Editorial

Hypertrophic Cardiomyopathy Is at Increased Risk of Thromboembolic Events: Deficiencies of CHA_2DS_2-VASC Score and How to Predict

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See article by He et al., pages 1800–1806 of this issue.

Patients with hypertrophic cardiomyopathy (HCM) are at increased risk of potentially catastrophic thromboembolic (TE) events that can occur without any previous clinical symptoms.[1,2] The most important therapeutic measure is lifelong anticoagulation therapy in all patients who have experienced a first episode of atrial fibrillation or TE event.[3] Unlike the risk assessment strategies used for atrial fibrillation in the general population, the Congestive Heart Failure, Hypertension, Diabetes Mellitus, Vascular Disease, Age 65 to 74 Years, and Female Sex, and 2 Points Each For Age $\geq$ 75 Years And Prior Stroke, Transient Ischemic Attack, and Thromboembolism (CHA_2DS_2-VASC) score is not usable in the setting of HCM.[3,4] Because an annual incidence of stroke or peripheral embolic events has been reported to be approximately 1% in HCM,[5] permanent anticoagulation might be considered for at-risk individuals, even in patients with sinus rhythm who have an enlarged left atrial diameter.[3,5]

Recently, the Hypertrophic Cardiomyopathy Outcome Investigators developed a model to predict TE risk in patients with HCM (HCM Risk-CVA model).[4] They demonstrated that the risk can be easily identified using several simple clinical factors. The model was developed using data from a retrospective, multicenter international cohort (Hypertrophic Cardiomyopathy Outcome Investigators) comprising 4821 patients with HCM with a follow-up period of 28,331 patient-years. The 5- and 10-year cumulative incidences of TE events were 2.9% and 6.4%, respectively. This study confirmed a high risk of TE events in patients with atrial fibrillation and a strong association between TE risk and left atrial diameter. Also, the authors demonstrated the association of patient age, heart failure, maximal left ventricular wall thickness, and vascular disease with long-term TE risk. In a later study, the same group of authors showed an association between atrial fibrillation and atrial dilation, heart failure and age.[1]

In this issue of the *Canadian Journal of Cardiology*, He et al.[6] describe the first external validation of the previously developed HCM Risk-CVA model for the long-term TE risk. They collected 417 patients with HCM and followed them for 3.5 years. Six percent of the patients experienced a TE end point, and the model demonstrated modest, but nevertheless potentially useful, discriminating capacity for TE events (C-index 0.67). Therefore, the authors suggest using it in clinical practice for individual prognosis and antithrombotic management.

Unfortunately, this study has some limitations, including a low number of patients and consequently low event rates, as well as specific management of HCM that is not fully compatible with cardiology practice in Western countries. Only 44% of patients with atrial fibrillation were treated with anticoagulation therapy, which seems to be surprisingly low.

Nevertheless, the study by He et al.[6] provides several interesting take-home messages. The most important of these is that sicker and older patients with HCM have a higher risk of TE events. Although such a statement might sound too simple, it has clinical importance and reintroduces into the discussion new information and research approaches to HCM, which has been until now mainly focused on debates about the risk stratification of sudden cardiac death or the therapeutic dilemma of alcohol septal ablation versus surgical myectomy.

As reported by Maron et al.,[7] the prognosis of patients with HCM aged $\geq$ 60 years of age is based largely on comorbidities and complications, including heart failure, atrial fibrillation, and consequent stroke. On the other hand, younger patients should be carefully stratified according to risk models for sudden cardiac death.[3,8]

Further data regarding indications for anticoagulation therapy in risky patients with HCM are urgently needed. Until then, we have to be aware that the risk of TE events in HCM is relatively high, and we should actively search for the TE risk factors validated in the study by He et al.[6] (Table 1).

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See page 1630 for disclosure information.

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Table 1. Risk factors for thromboembolic events in patients with hypertrophic cardiomyopathy

Greater age
History of vascular disease: myocardial infarction, peripheral arterial disease, aortic plaque
History of ischemic stroke, transient ischemic attack, or peripheral embolism
History of atrial fibrillation
Heart failure in functional NYHA class II-IV
Left atrial diameter
Maximal left ventricular wall thickness

NYHA, New York Heart Association.

Disclosures

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References

1. Guttmann OP, Pavlou M, ÓMahony C, et al. Predictors of atrial fibrillation in hypertrophic cardiomyopathy. Heart 2017;103:672-8.

2. Veselka J, Zemánek D, Jahnlová D, et al. Risk and causes of death in patients after alcohol septal ablation for hypertrophic obstructive cardiomyopathy. Can J Cardiol 2015;31:1245-51.

3. Veselka J, Anavekar NS, Charron P. Hypertrophic obstructive cardiomyopathy. Lancet 2017;389:1253-67.

4. Guttman OP, Pavlou M, ÓMahony C, et al. Prediction of thromboembolic risk in patients with hypertrophic cardiomyopathy (HCM Risk-CVA). Eur J Heart Fail 2015;17:837-45.

5. Haruki S, Minami Y, Hagiwara N. Stroke and embolic events in hypertrophic cardiomyopathy: risk stratification in patients without atrial fibrillation. Stroke 2016;47:936-42.

6. He S, Wang Z, Cheem TH, et al. External validation of the model of thromboembolic risk in hypertrophic cardiomyopathy patients. Can J Cardiol 2019;35:1800-6.

7. Maron BJ, Rowin EJ, Casey SA, et al. Risk stratification and outcome of patients with hypertrophic cardiomyopathy ≥60 years of age. Circulation 2013;127:585-93.

8. Cooper RM, Raphael CE, Liebregts M, et al. New developments in hypertrophic cardiomyopathy. Can J Cardiol 2017;33:1254-65.