

Recent ESC guidelines to identify HCM patients at high risk for sudden death unreliable

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Hypertrophic cardiomyopathy (HCM), a genetic form of heart disease with thick heart walls, is the most common cause of sudden death in patients under 50. While only a minority of patients with HCM are at risk, the implantable cardioverter-defibrillator (ICD) can effectively prevent sudden death in those patients. Recently, the European Society of Cardiology (ESC) published new guidelines advancing an equation as the best way to determine which HCM patients should receive an ICD. However, a study conducted by U.S. and Canadian investigators challenges the ESC Guidelines. Their research found that the ESC sudden death risk score method did not perform effectively in reliably identifying the high-risk patients who need ICDs for the prevention of sudden death.

"Our data show that in HCM, strategies to identify implantable defibrillator candidates based on a rigid mathematical and statistical formula, as promoted by ESC, are unreliable for identifying high-risk [patients](#) who could benefit from lifesaving therapy with prophylactic ICDs," stated lead investigator Barry J. Maron, MD, of the Hypertrophic Cardiomyopathy Center, Minneapolis Heart Institute Foundation, Minneapolis, Minnesota.

The method for identifying high-risk patients is called risk stratification and it is predicated on the results of certain tests (such as ambulatory ECG monitoring, echocardiogram, and stress testing), and personal and

family histories. Syncope (fainting), extreme thickening of the left ventricle, and ventricular tachycardia, as well as extensive scarring of the wall detectable by MRI with contrast can be risk markers. This assessment method and risk markers are a prominent part of the U.S. Guidelines on management of HCM and identification of patients at unacceptably high-risk, and have been well accepted as such.

In evaluating the ESC Guidelines, researchers used the case records of more than 1600 patients and found that for the 35 patients who died suddenly, only 4 (11%) would have been considered high risk by ESC criteria. For the 41 patients who had a life-saving shock from a defibrillator, 27 (59%) would not have even received a defibrillator under the ESC Guidelines and theoretically would have been at risk for [sudden death](#).

"We have retrospectively tested the mathematical ESC risk model against a large independent external cohort of individual patients with HCM," added Dr. Maron. "We found the ESC prognostic score was unreliable in identifying most high-risk patients previously managed in accord with risk stratification practices established by U.S. HCM consensus [guidelines](#)."

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