Ultra-Low Risk of Sudden Cardiac Death Seen in HCM Challenges Prevailing Views

Steve Stiles

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The risk for sudden cardiac death (SCD) in adolescents and young adults with hypertrophic cardiomyopathy (HCM) is "an order of magnitude lower" than most recent estimates would indicate, a report based on comprehensive autopsy data suggests.

The analysis of people experiencing SCD, aged 10 to 45 years, identified only 44 cases of "definite" HCM-related SCD and nine more considered "probable" or "possible" over 12 years in Ontario, Canada, with its population of 13.6 million people, say researchers.

That's far lower than the 0.5% to 1.0% annual rate often cited for SCD in patients with the genetic disease based on registries and referral populations, which were likely subject to selection bias, Paul Dorian, MD, St. Michael's Hospital, Toronto, told theheart.org | Medscape Cardiology.

"If an individual patient is identified with HCM, their risk of dying suddenly is very likely less and maybe much less than previously believed," he said.

"A second thing that our research indicates is that if you have HCM and you die suddenly, sudden death is much less often precipitated by exercise than previously believed."

SCD related to HCM struck during exercise that was at least moderate in less than one-fifth of cases in the analysis, published October 21 in Circulation, with Dorian as senior author and Adaya Weissler-Snir, MD, St. Michael's Hospital, as lead author.

It may have largely avoided the kind of selection bias typical of autopsy studies because in Ontario, "coroners are required to investigate any death that is sudden, unexpected, or from nonnatural causes," the report notes.

"By protocol, every unexpected sudden death in an individual <45 years of age is subject to autopsy except for cases with a well-documented cardiac history."

That may explain why HCM-related SCD rates in the current analysis are strikingly lower than most estimates based on the literature, Dorian explained. Those tend to come from "observational studies in cohorts of patients that have by definition been referred to and are participants in a registry."

Probably, he said, "these patients don't represent the universe of patients with HCM; they represent a selected subgroup."

The current analysis indeed challenges presumptions about the prevalence of HCM-related SCD on which most current risk estimates are based, agreed Jil C. Tardiff, MD, PhD, University of Arizona College of Medicine, Tucson.

"The original estimates of sudden death in HCM were really high," she observed for theheart.org | Medscape Cardiology. Selection and referral bias "has really haunted the field for many years."

But the higher estimates have been falling in recent years, she noted, and the current analysis — with which she was not involved — continues the trend. Its advantage is that "the autopsies were performed in all comers. There was no selection."

It's key point for patients with HCM, she said, "is not that sudden cardiac death is not important, it's just much rarer than we thought."
And that "fits with the message we're trying to send to patients, that most of them will live with normal life spans, and that [HCM] is not some sort of dreadful death sentence."

That exercise seems to play a smaller than appreciated role in eliciting HCM-related SCD is also in line with other research, Tardiff said, and "is actually, to my mind, a hugely important part of this study."

SCD in the autopsy series struck during moderate to vigorous exercise in only 17% of cases; it was during sleep or rest in 64%, the report notes.

"Patients are still told all the time not to exercise," Tardiff said. "This should really start to put that to rest."

However, the association of SCD to exercise seemed to vary by age. HCM-related SCD was linked to exercise in 77.8% of cases in people 20 years or younger but in only 4.8% of those older than 20 years \(P < .001\).

Also consistent with the literature, Tardiff noted, is the finding that HCM-related SCD was more than five times as likely in males as in females.

| Rate of SCD per 1000 HCM Person-Years by HCM Determination at Autopsy at Age 10 to 45 Years |
|---------------------------------|-------------|---------|----------|
| HCM Determination         | Overall     | Male    | Female*  |
| Definite           | 0.31        | 0.53    | 0.098    |
| Definite or probable | 0.33        | 0.63    | 0.11     |
| Definite, probable, or possible | 0.38 | 0.63    | 0.13     |

\*\(P < .001\) vs male for all rate differences

The analysis has further implications for the use of implantable cardioverter defibrillators (ICDs) in patients with HCM, in that the devices are probably less protective than suggested by current risk calculators, Dorian observed.

But that would likely vary by how the HCM diagnosis came about, for example, whether after referral to a specialist based on symptoms or from sports preparticipation screening.

"In general our work would suggest that, depending on how that patient was identified, the risk of sudden death is probably lower than most doctors believe, and therefore the benefit from an ICD is probably less than most doctors believe," he said.

The current analysis indeed argues for less use of ICDs in patients with HCM, Tardiff agreed. "The bottom line is, if sudden cardiac death is this rare, then we really need to think very deeply about who gets them."

She said what patients with HCM are told about the risk for SCD tends to be based on old assumptions that don't reflect contemporary understanding. SCD remains a concern for them, but "I think sudden cardiac death has taken an outsized importance in how we think of the disease and how it's portrayed."

Weissler-Snir, Dorian, and the other authors report no conflicts. Tardiff said she has no relevant disclosures.

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