

Is prevention of sudden cardiac death feasible?

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Hypertrophic cardiomyopathy (HCM) has been recognized for 50 years. The risk for unexpected sudden cardiac death was an important component of the initial contemporary description of HCM (Teare; 1958).

Indeed, we now recognize HCM to be the most common cause of sudden death in the young, including trained athletes. While the implantable cardioverter-defibrillator (ICD) was introduced over 25 years ago, it was not systematically applied to HCM until year 2000. The prevention of sudden death has long been an aspiration in HCM.

Early experiences with pharmacologic strategies demonstrated that drugs (e.g., amiodarone) are not absolutely protective against sudden death.

Based on recent substantial experience, the ICD has now proved to be a safe and the only effective therapeutic intervention in patients with HCM, both for primary and secondary prevention of sudden death. The ICD intervenes appropriately to terminate ventricular tachycardia/fibrillation (VT/VF) at a rate of 5.5%/year. The ICD discharge rate is 4%/year for those patients implanted prophylactically due to one or more major risk markers.

Considerable delays up to 10 years may occur before the ICD is required to intervene to abort potentially lethal ventricular arrhythmias.

Primary prevention of VT/VF occurs with similar frequency in high-risk patients with either 1, 2 or ≥ 3 noninvasive risk markers. About one-third of patients with appropriate shocks were implanted for only one risk factor.

The ICD has proved reliable despite the extreme and complex phenotype often present in HCM with massive degrees of left ventricular hypertrophy, microvascular ischemia diastolic dysfunction, or dynamic left ventricular outflow tract obstruction.

Failure to convert life-threatening ventricular tachyarrhythmias to normal rhythm is extraordinarily rare.

BULLET POINTS:

- In high-risk HCM patients, ICDs perform in a highly effective fashion, frequently preventing sudden death by terminating primary life-threatening ventricular arrhythmias
- A single marker of high risk can be sufficient evidence to justify consideration for a prophylactic ICD in selected patients with HCM

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