CARDIO-SIGNAL

60

58 Y/O gentleman, K/C of HCM came for second opinion about the need for ICD implantation

Asymptomatic, no syncope or tachyarrhythmia

Neg FH for SCD

Under medical Tx. for HFrEF

Echo: Type III HCM with apical aneurysm, EF=35-40%,

LVOT gradient: 5mmHg, max wall thickness: 2.6cm

LA diameter: 3.3cm

Do you recommend ICD implantation in this patient?

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Yes, it's a correct recommendation

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Recommendations for SCD Risk Assessment
Referenced studies that support the recommendations are summarized in Online Data Supplement 11.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with HCM, a comprehensive, systematic noninvasive SCD risk assessment at initial evaluation and every 1 to 2 years thereafter is recommended and should include evaluation of these risk factors a. Personal history of cardiac arrest or sustained ventricular arrhythmias b. Personal history of syncope suspected by clinical history to be arrhythmic c. Family history in close relative of premature HCM-related sudden death, cardiac arrest, or sustained ventricular arrhythmias d. Maximal LV wall thickness, EF, LV apical aneurysm e. Nonsustained ventricular tachycardia episodes on continuous ambulatory electrocardiographic monitoring

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a. Personal history of cardiac arrest or sustained ventricular arrhythmias
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2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: Executive Summary

TABLE 7 Established Clinical Risk Factors for HCM Sudden Death Risk Stratification

Family history of sudden death from HCM	Sudden death judged definitively or likely attributable to HCM in ≥1 first-degree or close relatives who are ≤50 years of age. Close relatives would generally be second-degree relatives; however, multiple SCDs in tertiary relatives should also be considered relevant.
Massive LVH	Wall thickness ≥30 mm in any segment within the chamber by echocardiography or CMR imaging; consideration for this morphologic marker is also given to borderline values of ≥28 mm in individual patients at the discretion of the treating cardiologist. For pediatric patients with HCM, an absolute or z-score threshold for wall thickness has not been established; however, a maximal wall that corresponds to a z-score ≥20 (and >10 in conjunction with other risk factors) appears reasonable.
Unexplained syncope	≥1 Unexplained episodes involving acute transient loss of consciousness, judged by history unlikely to be of neurocardiogenic (vasovagal) etiology, nor attributable to LVOTO, and especially when occurring within 6 months of evaluation (events beyond 5 years in the past do not appear to have relevance).
HCM with LV systolic dysfunction	Systolic dysfunction with EF <50% by echocardiography or CMR imaging.
LV apical aneurysm	Apical aneurysm defined as a discrete thin-walled dyskinetic or akinetic segment of the most distal portion of the LV chamber; independent of size.
Extensive LGE on CMR imaging	Diffuse and extensive LGE, representing fibrosis, either quantified or estimated by visual inspection, comprising ≥15% of LV mass (extent of LGE conferring risk has not been established in children).
NSVT on ambulatory monitor	It would seem most appropriate to place greater weight on NSVT as a risk marker when runs are frequent (≥3), longer (≥10 beats), and faster (≥200 bpm) occurring usually over 24 to 48 hours of monitoring. For pediatric patients, a VT rate that exceeds the baseline sinus rate by >20% is considered significant.

CMR indicates cardiovascular magnetic resonance; ICD, implantable cardioverter-defibrillator; LGE, late gadolinium enhancement; LV, left ventricular; LVH, left ventricular hypertrophy; LVOTO, left ventricular outflow tract obstruction; NSVT, nonsustained ventricular tachycardia; and SCD, sudden cardiac death.

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2a	B-NR

3. For adult patients with HCM with ≥1 major risk factors for SCD, it is reasonable to offer an ICD. These major risk factors include
   a. Sudden death judged definitively or likely attributable to HCM in ≥1 first-degree or close relatives who are ≤50 years of age;
   b. Massive left ventricular hypertrophy ≥30 mm in any left ventricular segment;
   c. ≥1 Recent episodes of syncope suspected by clinical history to be arrhythmic (i.e., unlikely to be of neurocardiogenic [vasovagal] etiology, or related to LVOTO);
   d. LV apical aneurysm, independent of size;
   e. LV systolic dysfunction (EF <50%).

3. For adult patients with HCM with ≥1 major risk factors for SCD, it is reasonable to offer an ICD. These major risk factors include
   a. Sudden death judged definitively or likely attributable to HCM in ≥1 first-degree or close relatives who are ≤50 years of age;
   b. Massive left ventricular hypertrophy ≥30 mm in any left ventricular segment;
   c. ≥1 Recent episodes of syncope suspected by clinical history to be arrhythmic (i.e., unlikely to be of neurocardiogenic [vasovagal] etiology, or related to LVOTO);
   d. LV apical aneurysm, independent of size;
   e. LV systolic dysfunction (EF <50%).

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Hypertrophic Cardiomyopathy Sudden Cardiac Death Risk Calculator

Age

58 years

MLVWT

26 mm

LA Size

33 mm

Max LVOT Gradient

5 mmHg

FH SCD	No	Yes
NSVT	No	Yes
Unexplained Syncope	No	Yes
EF ≤ 50%	No	Yes
Apical Aneurysm	No	Yes
Extensive LGE	No	Yes

Risk of SCD at 5 years(%)

1.45 (*Nota bene*: This estimate may not be accurate in the setting of Apical Aneurysm and EF ≤ 50%)

Recommendation

Based on the SCD Risk factors present, this patient has a Class 2A indication for ICD (is reasonable)

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