Αιφνίδιος καρδιακός θάνατος

ΣΤΗ ΧΡΟΝΙΑ ΣΤΕΦΑΝΙΑΙΑ ΝΟΣΟ ΚΑΙ ΣΤΙΣ ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΕΣ - ΜΥΟΚΑΡΔΙΤΙΔΕΣ

ΙΩΑΝΝΗΣ ΣΤΥΛΙΑΔΗΣ
ΑΝ. ΣΥΝΤ. ΔΙΕΥΘΥΝΤΗΣ ΚΑΡΔΙΟΛΟΓΙΚΗΣ ΚΛΙΝΙΚΗΣ
ΓΝΘ ΠΑΠΑΓΕΩΡΓΙΟΥ
First cardiac rhythm documented at time of sudden arrhythmic death. VT indicates ventricular tachycardia; VF, ventricular fibrillation.

Rajat Deo, and Christine M. Albert Circulation. 2012;125:620-637
Venn diagram showing interaction of various anatomic/functional and transient factors that modulate potential arrhythmogenic mechanisms capable of causing sudden cardiac death.


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Critical pathways leading to electric instability and sudden cardiac death.

Rajat Deo, and Christine M. Albert Circulation.
2012;125:620-637

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Structural heart disease in cardiac arrest survivors

Men
- CAD 80%
- VHD 5%
- DCM 10%
- Normal 3%
- Other 2%

Women
- CAD 45%
- VHD 13%
- DCM 19%
- Normal 10%
- Other 2%
- RV Dysplasia 2%
- Congenital 2%
- Spasm 5%
- Long QT 2%

Rajat Deo, and Christine M. Albert Circulation.
2012;125:620-637

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The magnitude of SCD risk

Age and cause-related
Σε ποιες συνθήκες συμβαίνει ο ΑΚΘ


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Επίπτωση & φορτίο ΑΚΘ

2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC) Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC)

Silvia G Priori, Carina Blomström-Lundqvist, Andrea Mazzanti, Nico Blom, Martin Borggrefe, John Camm, Perry Mark Elliott, Donna Fitzsimons, Robert Hatala,

ACC/AHA/HRS GUIDELINE


Sana M. Al-Khatib, William G. Stevenson, Michael J. Ackerman, William J. Bryant, David J. Callans, Anne B. Curtis, Barbara J. Deal, Timm Dickfeld, Michael E. Feld, Gregg C. Fonarow, Anne M. Gillis, Mark A. Hlatky, Christopher B. Granger, Stephen C. Hammill, José A. Joglar, G. Neal Kay, Daniel D. Matlock, Robert J. Myerburg, Richard L. Page
### 7.1.1. Secondary Prevention of SCD in Patients With Ischemic Heart Disease

**Recommendations for Secondary Prevention of SCD in Patients With Ischemic Heart Disease**

References that support the recommendations are summarized in Online Data Supplement 17 and 18.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-R</td>
<td>1. In patients with ischemic heart disease, who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R) (1-4) or stable VT (LOE: B-NR) (5) not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.</td>
</tr>
<tr>
<td>B-NR</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Value Statement: Intermediate Value (LOE: B-R)**

<table>
<thead>
<tr>
<th></th>
<th></th>
<th>2. A transvenous ICD provides intermediate value in the secondary prevention of SCD particularly when the patient’s risk of death due to a VA is deemed high and the risk of nonrhythmic death (either cardiac or noncardiac) is deemed low based on the patient’s burden of comorbidities and functional status (6).</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-R</td>
<td>3. In patients with ischemic heart disease and unexplained syncope who have inducible sustained monomorphic VT on electrophysiological study, an ICD is recommended if meaningful survival of greater than 1 year is expected (7).</td>
</tr>
<tr>
<td>B-NR</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Causes of Death in the Antiarrhythmics Versus Implantable Defibrillators (AVID) Trial
The AVID Investigators

Log rank $p = 0.0042$

Assigned Therapy

Drug

Device

Cumulative Survival

Months from Randomization
Kaplan-Meier plots depicting cumulative risk of death from any cause for treatment groups on basis of intention-to-treat analysis (P=0.142).
Figure 3. Secondary Prevention Patients With Ischemic Heart Disease

Secondary prevention in pts with IHD

SCA survivor* or sustained spontaneous monomorphic VT*

Cardiac syncope†

LVEF<35%

Ischemia warranting revascularization

Yes

No

ICD (Class I)  EP study (Class IIa)

Revascularize & reassess SCD risk (Class I)

ICD candidate‡

Inducible VA

Yes

No

ICD (Class I)  GDMT (Class I)  ICD (Class I)  Extended monitoring

Colors correspond to Class of Recommendation in Table 1.

See Sections 4.3.1 and 7.1.1 for discussion.

*Exclude reversible causes.
†History consistent with an arrhythmic etiology for syncope.
‡ICD candidacy as determined by functional status, life expectancy, or patient preference.
EP indicates electrophysiological; GDMT, guideline-directed management and therapy; ICD, implantable cardioverter-defibrillator; IHD, ischemic heart disease; LVEF, left ventricular ejection fraction; pts, patients; SCA, sudden cardiac arrest; SCD, sudden cardiac death; and VT, ventricular tachycardia.
7.1.1.1. **Coronary Artery Spasm**

**Recommendations for Patients With Coronary Artery Spasm**

References that support the recommendations are summarized in Online Data Supplement 20.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-NR</td>
<td>1. In patients with VA due to coronary artery spasm, treatment with maximally tolerated doses of a calcium channel blocker and smoking cessation are indicated to reduce recurrent ischemia and VA (1, 2).</td>
</tr>
<tr>
<td>IIA</td>
<td>B-NR</td>
<td>2. In patients resuscitated from SCA due to coronary artery spasm in whom medical therapy is ineffective or not tolerated, an ICD is reasonable if meaningful survival of greater than 1 year is expected (3-6).</td>
</tr>
<tr>
<td>IIb</td>
<td>B-NR</td>
<td>3. In patients resuscitated from SCA due to coronary artery spasm, an ICD in addition to medical therapy may be reasonable if meaningful survival of greater than 1 year is expected (3-6).</td>
</tr>
</tbody>
</table>
MORTALITY RATE REDUCTION WITH ICDs

ICD mortality reductions in primary prevention trials are equal to or greater than those in secondary prevention trials.
### 7.1.2. Primary Prevention of SCD in Patients With Ischemic Heart Disease

**Recommendations for Primary Prevention of SCD in Patients With Ischemic Heart Disease**

References that support the recommendations are summarized in Online Data Supplement 21.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>A</td>
<td>1. In patients with LVEF of 35% or less that is due to ischemic heart disease who are at least 40 days’ post-MI and at least 90 days postrevascularization, and with NYHA class II or III HF despite GDMT, an ICD is recommended if meaningful survival of greater than 1 year is expected (1, 2).</td>
</tr>
<tr>
<td>I</td>
<td>A</td>
<td>2. In patients with LVEF of 30% or less that is due to ischemic heart disease who are at least 40 days’ post-MI and at least 90 days postrevascularization, and with NYHA class I HF despite GDMT, an ICD is recommended if meaningful survival of greater than 1 year is expected (2, 3).</td>
</tr>
<tr>
<td></td>
<td></td>
<td><strong>Value Statement:</strong> High Value (LOE: B-R)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3. A transvenous ICD provides high value in the primary prevention of SCD particularly when the patient’s risk of death due to a VA is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient’s burden of comorbidities and functional status (4).</td>
</tr>
<tr>
<td>I</td>
<td>B-R</td>
<td>4. In patients with NSVT due to prior MI, LVEF of 40% or less and inducible sustained VT or VF at electrophysiological study, an ICD is recommended if meaningful survival of greater than 1 year is expected (5).</td>
</tr>
<tr>
<td>IIa</td>
<td>B-NR</td>
<td>5. In nonhospitalized patients with NYHA class IV symptoms who are candidates for cardiac transplantation or an LVAD, an ICD is reasonable if meaningful survival of greater than 1 year is expected (6-9).</td>
</tr>
<tr>
<td>III: No Benefit</td>
<td>C-EO</td>
<td>6. An ICD is not indicated for NYHA class IV patients with medication-refractory HF who are not also candidates for cardiac transplantation, an LVAD, or a CRT defibrillator that incorporates both pacing and defibrillation capabilities.</td>
</tr>
</tbody>
</table>
Figure 4. Primary Prevention of SCD in Patients With Ischemic Heart Disease

Primary prevention in pts with IHD, LVEF ≤40%  
MI <40 d  
and/or revascularization <90 d  
Yes*  
Inducible sustained VT  
Yes  
ICD (Class I)  
No  
No  
Reassess LVEF >40 d after MI  
and/or >90 d after revascularization  
NYHA class I  
LVEF ≤30%  
Yes  
ICD (Class I)*  
No  
NYHA class II or III  
LVEF ≤35%  
Yes  
ICD (Class I)  
No  
LVEF ≤40%, NSVT, inducible sustained VT or EP study  
Yes  
ICD (Class Ia)  
No  
GDMT (Class Ia)  
NYHA class IV candidate for advanced HF therapy†  
Yes  
ICD should not be implanted (Class III: No Benefit)  
No  
GDMT (Class Ib)

Colors correspond to Class of Recommendation in Table 1.  
See Section 7.1.2 for discussion.  
*Scenarios exist for early ICD placement in select circumstances such as patients with a pacing indication or syncope.  
†Advanced HF therapy includes CRT, cardiac transplant, and LVAD.  
thought due to VT. These are detailed elsewhere in an HRS/ACC/AHA expert consensus statement (24).  
CRT indicates cardiac resynchronization therapy; EP, electrophysiological; GDMT, guideline-directed management and therapy; HF, heart failure; ICD, implantable cardioverter-defibrillator; IHD, ischemic heart disease; LVEF, left ventricular ejection fraction; MI, myocardial infarction; NSVT, nonsustained ventricular tachycardia; NYHA, New York Heart Association; pts, patients; SCD, sudden cardiac death; VT, ventricular tachycardia; and WCD, wearable cardioverter-defibrillator.
### 7.1.3. Treatment and Prevention of Recurrent VA in Patients With Ischemic Heart Disease

Recommendations for Treatment of Recurrent VA in Patients With Ischemic Heart Disease

References that support the recommendations are summarized in Online Data Supplement 22 and 23.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-R</td>
<td>1. In patients with ischemic heart disease and recurrent VA, with significant symptoms or ICD shocks despite optimal device programming and ongoing treatment with a beta blocker, amiodarone or sotalol is useful to suppress recurrent VA (1-3).</td>
</tr>
<tr>
<td>I</td>
<td>B-R</td>
<td>2. In patients with prior MI and recurrent episodes of symptomatic sustained VT, or who present with VT or VF storm and have failed or are intolerant of amiodarone (LOE: B-R) (4) or other antiarrhythmic medications (LOE: B-NR) (5-9), catheter ablation is recommended (10-12).</td>
</tr>
<tr>
<td>IIb</td>
<td>C-LD</td>
<td>3. In patients with ischemic heart disease and ICD shocks for sustained monomorphic VT or symptomatic sustained monomorphic VT that is recurrent, or hemodynamically tolerated, catheter ablation as first-line therapy may be considered to reduce recurrent VA (10, 11).</td>
</tr>
<tr>
<td>III:</td>
<td>B-R</td>
<td>4. In patients with prior MI, class IC antiarrhythmic medications (e.g., flecainide and propafenone) should not be used (13).</td>
</tr>
<tr>
<td>Harm</td>
<td>C-LD</td>
<td>5. In patients with incessant VT or VF, an ICD should not be implanted until sufficient control of the VA is achieved to prevent repeated ICD shocks (14).</td>
</tr>
<tr>
<td>III:</td>
<td>C-LD</td>
<td>6. In patients with ischemic heart disease and sustained monomorphic VT, coronary revascularization alone is an ineffective therapy to prevent recurrent VT (15, 16).</td>
</tr>
</tbody>
</table>
Mortality and Morbidity in Patients Receiving Encainide, Flecainide, or Placebo — The Cardiac Arrhythmia Suppression Trial

<table>
<thead>
<tr>
<th>Study</th>
<th>Year</th>
<th>Patient population</th>
<th>Study arms</th>
<th>Findings</th>
<th>Comments</th>
</tr>
</thead>
</table>
| Substrate Mapping and Ablation in Sinus Rhythm to Halt Ventricular Tachycardia – SMASH-VT | 2007 | Prior MI with history of single ventricular arrhythmia or appropriate ICD therapy, but no prior AAD use | Substrate-based endocardial ablation versus standard medical therapy        | • Ablation decreased the likelihood of recurrent VT requiring ICD therapy (67 versus 88 %; p<0.007)  
• No significant difference in mortality rates | Exclusion of patients receiving AADs limits the clinical relevance |
| Ventricular Tachycardia Ablation in Coronary Heart Disease – VTACH    | 2010 | Prior MI, reduced left ventricular ejection fraction (<50 %), and stable VT, qualifying for a secondary prevention CD | Catheter ablation and ICD versus ICD alone. Both groups were eligible to receive AADs | • Ablation group had improved freedom from recurrent ventricular arrhythmias (47 versus 29 %; p<0.044)  
• No significant difference in mortality rates | Ablation reduced the frequency of recurrent VT; however, there was a >50 % recurrence rate within 2 years |
| Catheter Ablation for Ventricular Tachycardia in Patients with Implantable Cardioverter Defibrillator – CALYPSO | 2015 | Ischemic heart disease, no contraindication to AADs, and received >1 ICD shock or ≥3 anti-tachycardia pacing therapies for VT | Catheter ablation versus AAD (first line: amiodarone, Sotalol; second line: mexiletine, ranolazine, dofetilide) | • Lower risk of recurrent VT in the AAD arm: 64 (43 %) versus 8 62 %  
• Median time to recurrent VT was longer in the ablation arm: 75 versus 57 days | Significant difficulty enrolling patients. Underpowered to detect any differences in outcomes |
| Ventricular Tachycardia Ablation versus Escalated Antiarrhythmic Drug Therapy in Ischemic Heart Disease – VANISH | 2016 | Prior MI, ICD, and recurrent VT despite use of AADs | Catheter ablation versus escalation of AAD therapy (amiodarone, mexiletine) | • Composite endpoint all-cause mortality, VT storm, or appropriate ICD shock after 30 days occurred less in ablation versus AAD escalation (59.1 versus 88.5 %; p=0.04)  
• Adverse events more common in patients randomized to AAD escalation | For patients with recurrent VT despite AAD therapy, catheter ablation is a valuable strategy that avoids AAD-related adverse events |

AAD – anti-arrhythmic drug; ICD – implantable cardioverter defibrillator; VT – ventricular tachycardia.
Figure 5. Treatment of Recurrent VA in Patients With Ischemic Heart Disease or NICM

ICD with VT/VF recurrent arrhythmia*

Polymorphic VT/VF

Consider reversible causes

Sustained monomorphic VT

Catheter ablation as first-line therapy (Class I)

Amiodarone or sotalol (Class I)

Arrhythmia not controlled

Drug, electrolyte induced

Treat for QT prolongation, discontinue offending medication, correct electrolytes (Class I)

Revascularize (Class I)

Amiodarone (Class I)

Beta blockers or lidocaine (Class IIa)

No reversible causes

Arrhythmia not controlled

INO with frequent VT or VT storm

ANOICM

Identifiable PVC triggers

Catheter ablation (Class I)

Autonomic modulation (Class IIb)

Yes

No

Catheter ablation (Class I)

Catheter ablation (Class IIa)

Catheter ablation (Class IIa)

Colors correspond to Class of Recommendation in Table 1. See Sections 5.6, 6.7.1, and 7.2 for discussion.

*Management should start with ensuring that the ICD is programmed appropriately and that potential precipitating causes, including heart failure exacerbation, are addressed. For information regarding optimal ICD programming, refer to the 2015 HRS/EHRA/APHRS/SOLACE expert consensus statement (26).

APHRS indicates Asia Pacific Heart Rhythm Society; EHRA, European Heart Rhythm Society; HRS, Heart Rhythm Society; IHD, ischemic heart disease; ICD, implantable cardioverter-defibrillator; PVC, premature ventricular complex; NICM, nonischemic cardiomyopathy; SOLACE, Sociedad Latinoamericana de Estimulación Cardíaca y Electrofisiología; VF, ventricular fibrillation; and VT, ventricular tachycardia.
Nonischemic Cardiomyopathy
### 7.2. Nonischemic Cardiomyopathy

**Recommendations for Patients With NICM**

References that support the recommendations are summarized in Online Data Supplement 24.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-NR</td>
<td>1. In patients with suspected NICM from myocardial infiltrative processes, cardiac MRI with late gadolinium enhancement is useful for diagnosis (1-3).</td>
</tr>
<tr>
<td>IIa</td>
<td>B-NR</td>
<td>2. In patients with suspected NICM, cardiac MRI with late gadolinium enhancement can be useful for assessing risk of SCA/SCD (1-3).</td>
</tr>
<tr>
<td>IIa</td>
<td>C-EO</td>
<td>3. In patients with NICM who develop conduction disease or LV dysfunction at less than 40 years of age, or who have a family history of NICM or SCD in a first-degree relative (&lt;50 years of age), genetic counseling and genetic testing are reasonable to detect a heritable disease that may clarify prognosis and facilitate cascade screening of relatives (4, 5).</td>
</tr>
</tbody>
</table>
Figure 6. Secondary and Primary Prevention of SCD in Patients With NICM

Patients with NICM

SCA survivor/sustained VT (spontaneous/inducible)

Yes

ICD candidate*

No

Symptoms concerning for VA

Yes

No

No

No

Class II-III HF and LVEF ≤35%

Yes

No

Yes

Arrhythmogenic syncope suspected

Etiology uncertain

No, due to newly diagnosed HF (<3 mo GDMT) or not on optimal GDMT

ICD candidate*

No

ICD candidate*

ICD (Class I)

Amiodarone (Class Ib)

ICD (Class IIa)

EP Study (Class IIa)

ICD (Class I)

WCD (Class IIb)

If LVEF <35%

If positive

If LV <35% and Class II-III HF

Reassess LVEF ≥3mo

NICM due to LMNA mutation and 2nd risk factors

Colors correspond to Class of Recommendation in Table 1. See Section 7.2 for discussion.

*ICD candidacy is determined by functional status, life expectancy or patient preference.

2nd indicates secondary; EP, electrophysiological; GDMT, guideline-directed management and therapy; HF, heart failure; ICD, implantable cardioverter-defibrillator; LVEF, left ventricular ejection fraction; NICM, nonischemic cardiomyopathy; SCA, sudden cardiac arrest; SCD, sudden cardiac death; VA, ventricular arrhythmia; and WCD, wearable cardiac-defibrillator.
### 7.2.3. Treatment of Recurrent VA in Patients With NICM

**Recommendations for Treatment of Recurrent VA in Patients With NICM**

References that support the recommendations are summarized in Online Data Supplement 29.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>IIa</td>
<td>B-R</td>
<td>1. In patients with NICM and an ICD who experience spontaneous VA or recurrent</td>
</tr>
<tr>
<td></td>
<td></td>
<td>appropriate shocks despite optimal device programming and treatment with</td>
</tr>
<tr>
<td></td>
<td></td>
<td>a beta blocker, amiodarone or sotalol can be beneficial (1).</td>
</tr>
<tr>
<td>IIa</td>
<td>B-NR</td>
<td>2. In patients with NICM and recurrent sustained monomorphic VT who fail or</td>
</tr>
<tr>
<td></td>
<td></td>
<td>are intolerant of antiarrhythmic medications, catheter ablation can be useful</td>
</tr>
<tr>
<td></td>
<td></td>
<td>for reducing recurrent VT and ICD shocks (2, 3).</td>
</tr>
</tbody>
</table>

Recommendation-Specific Supportive Text
Arrhythmogenic right ventricular cardiomyopathy is an inherited cardiomyopathy that predominantly affects the right ventricle but can affect the left ventricle causing areas of myocardial replacement with fibrosis and adipose tissue that frequently causes VA and SCD.
A and B, Bipolar voltage map of the right ventricular (RV) endocardium in a patient with arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) from group 1 (A, left anterior oblique view; B, anteroposterior view).

Rong Bai et al. Circ Arrhythm Electrophysiol. 2011;4:478-485
Ventricular arrhythmia (VA)/implantable cardioverter-defibrillator (ICD) therapy-free survival by the ablation approach.

Rong Bai et al. Circ Arrhythm Electrophysiol. 2011;4:478-485
### 7.3 Arrhythmogenic Right Ventricular Cardiomyopathy

References that support the recommendations are summarized in Online Data Supplement 30.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-NR</td>
<td>1. In selected first-degree relatives of patients with arrhythmogenic right ventricular cardiomyopathy, clinical screening for the disease is recommended along with genetic counseling and genetic testing, if the proband has a disease causing mutation (1-4).</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>2. In patients with suspected arrhythmogenic right ventricular cardiomyopathy and VA or electrocardiographic abnormalities, cardiac MRI is useful for establishing a diagnosis and for risk stratification (5-8).</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>3. In patients with arrhythmogenic right ventricular cardiomyopathy and an additional marker of increased risk of SCD (resuscitated SCA, sustained VT, significant ventricular dysfunction with RVEF or LVEF &lt;35%), an ICD is recommended if meaningful survival greater than 1 year is expected (9-13).</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>4. In patients with arrhythmogenic right ventricular cardiomyopathy and VA, a beta blocker is recommended (11, 14, 15).</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>5. In patients with a clinical diagnosis of arrhythmogenic right ventricular cardiomyopathy, avoiding intensive exercise is recommended (11, 12, 16-21).</td>
</tr>
<tr>
<td>Ila</td>
<td>B-NR</td>
<td>6. In patients with clinically diagnosed or suspected arrhythmogenic right ventricular cardiomyopathy, genetic counseling and genetic testing can be useful for diagnosis and for gene-specific targeted family screening (1, 4, 22-26).</td>
</tr>
<tr>
<td>Ila</td>
<td>B-NR</td>
<td>7. In patients with arrhythmogenic right ventricular cardiomyopathy and syncope presumed due to VA, an ICD can be useful if meaningful survival greater than 1 year is expected (10, 11, 13).</td>
</tr>
<tr>
<td>Ila</td>
<td>B-NR</td>
<td>8. In patients with clinical evidence of arrhythmogenic right ventricular cardiomyopathy but not VA, a beta blocker can be useful (14, 15).</td>
</tr>
<tr>
<td>Ila</td>
<td>B-NR</td>
<td>9. In patients with arrhythmogenic right ventricular cardiomyopathy and recurrent symptomatic sustained VT in whom a beta blocker is ineffective or not tolerated, catheter ablation with availability of a combined endocardial/epicardial approach can be beneficial (27-33).</td>
</tr>
<tr>
<td>Ila</td>
<td>B-NR</td>
<td>10. In patients with suspected arrhythmogenic right ventricular cardiomyopathy, a signal averaged ECG can be useful for diagnosis and risk stratification (14, 34, 35).</td>
</tr>
<tr>
<td>Iib</td>
<td>B-NR</td>
<td>11. In asymptomatic patients with clinical evidence of arrhythmogenic right ventricular cardiomyopathy, an electrophysiological study may be considered for risk stratification (5, 36).</td>
</tr>
</tbody>
</table>
ΥΠΕΡΤΡΟΦΙΚΗ ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΑ
Al-Khatib SM, et al.
2017 VA/SCD Guideline

Table 8. Major Clinical Features Associated With Increased Risk of SCD in Patients With HCM

<table>
<thead>
<tr>
<th>Established risk factors*</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Survival from a cardiac arrest due to VT or VF (1, 5, 6)</td>
</tr>
<tr>
<td>- Spontaneous sustained VT causing syncope or hemodynamic compromise (1, 5, 6)</td>
</tr>
<tr>
<td>- Family history of SCD associated with HCM (25, 26)</td>
</tr>
<tr>
<td>- LV wall thickness ≥30 mm (2, 3, 23, 24)</td>
</tr>
<tr>
<td>- Unexplained syncope within 6 mo (8, 26)</td>
</tr>
<tr>
<td>- NSVT ≥ beats (2, 26, 27)</td>
</tr>
<tr>
<td>- Abnormal blood pressure response during exercise† (5, 28, 29)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Potential risk modifiers‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>- &lt;30 y (5, 26)</td>
</tr>
<tr>
<td>- Delayed hyperenhancement on cardiac MRI (37-39, 54)</td>
</tr>
<tr>
<td>- LVOT obstruction (2, 4)</td>
</tr>
<tr>
<td>- Syncope &gt;5 y ago (8, 26)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>High-risk subsets§</th>
</tr>
</thead>
<tbody>
<tr>
<td>- LV aneurysm (40, 55, 56)</td>
</tr>
<tr>
<td>- LVEF &lt;50% (52)</td>
</tr>
</tbody>
</table>

*There is general agreement in the literature that these factors independently convey an increased risk for SCD in patients with HCM.
†Decrease in blood pressure of 20 mm Hg or failure to increase systolic blood pressure >20 mm Hg during exercise.
‡There is a lack of agreement in the literature that these modifiers independently convey an increased risk of SCD in patients with HCM; however, a risk modifier when combined with a risk factor often identifies a patient with HCM at increased risk for SCD beyond the risk conveyed by the risk factor alone.
§A small subset of patients with an LVEF <50% (end-stage disease) or an LV aneurysm warrant consideration for ICD implantation (52).

HCM indicates hypertrophic cardiomyopathy; ICD, implantable cardioverter-defibrillator; LV, left ventricular; LVEF, left ventricular ejection fraction; LVOT, left ventricular outflow tract; NSVT, nonsustained ventricular tachycardia; SCD, sudden cardiac death; VT, ventricular tachycardia; and VF, ventricular fibrillation.
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From: Left ventricular outflow tract obstruction and sudden death risk in patients with hypertrophic cardiomyopathy
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### 7.4. Hypertrophic Cardiomyopathy

**Recommendations for HCM**

References that support the recommendations are summarized in Online Data Supplement 31.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-NR</td>
<td>1. In patients with HCM, SCD risk stratification should be performed at the time of initial evaluation and periodically thereafter (1-8).</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>2. In patients with HCM who have survived an SCA due to VT or VF, or have spontaneous sustained VT causing syncope or hemodynamic compromise, an ICD is recommended if meaningful survival greater than 1 year is expected (1, 6, 9, 10).</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>3. In first-degree relatives of patients with HCM, an ECG and echocardiogram should be performed (11-17).</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>4. In first-degree relatives of patients with HCM due to a known causative mutation, genetic counseling and mutation-specific genetic testing are recommended (13-15, 18, 19).</td>
</tr>
<tr>
<td>IIa</td>
<td>B-NR</td>
<td>5. In patients with clinically suspected or diagnosed HCM, genetic counseling and genetic testing are reasonable (13-15, 18-22).</td>
</tr>
<tr>
<td>IIa</td>
<td>C-LD</td>
<td>6. In patients with HCM and 1 or more of the following risk factors, an ICD is reasonable if meaningful survival of greater than 1 year is expected:</td>
</tr>
<tr>
<td></td>
<td>C-LD</td>
<td>a. Maximum LV wall thickness ≥30 mm (LOE: B-NR) (2, 3, 23, 24).</td>
</tr>
<tr>
<td></td>
<td>C-LD</td>
<td>b. SCD in 1 or more first-degree relatives presumably caused by HCM (LOE: C-LD) (25, 26).</td>
</tr>
<tr>
<td></td>
<td>C-LD</td>
<td>c. 1 or more episodes of unexplained syncope within the preceding 6 months (LOE: C-LD) (8, 26).</td>
</tr>
<tr>
<td>IIa</td>
<td>B-NR</td>
<td>7. In patients with HCM who have spontaneous NSVT (LOE: C-LD) (2, 26, 27) or an abnormal blood pressure response with exercise (LOE: B-NR) (5, 28, 29), who also have additional SCD risk modifiers or high risk features, an ICD is reasonable if meaningful survival greater than 1 year is expected.</td>
</tr>
<tr>
<td>IIa</td>
<td>C-LD</td>
<td>8. In patients with HCM who have NSVT (LOE: B-NR) (2, 26, 27) or an abnormal blood pressure response with exercise (LOE: B-NR) (5, 28, 29) but do not have any other SCD risk modifiers, an ICD may be considered, but its benefit is uncertain.</td>
</tr>
<tr>
<td>IIb</td>
<td>B-NR</td>
<td>9. In patients with HCM and a history of sustained VT or VF, amiodarone may be considered when an ICD is not feasible or not preferred by the patient (30, 31).</td>
</tr>
<tr>
<td>IIb</td>
<td>C-LD</td>
<td>10. In patients with HCM, an invasive electrophysiological study with programmed ventricular stimulation should not be performed for risk stratification (32, 33).</td>
</tr>
<tr>
<td>III: No Benefit</td>
<td>B-NR</td>
<td>11. In patients with an identified HCM genotype in the absence of SCD risk factors, an ICD should not be implanted (7, 34, 35).</td>
</tr>
</tbody>
</table>
Figure 7. Prevention of SCD in Patients With HCM

Patients with HCM

SCD survivor; sustained VT No

Family Hx SCD; LVWT >30 mm; syncope <6 mo No

NSVT; abnormal BP response to exercise No

Yes

ICD candidate*

No

ICD (Class IIa)

Yes

ICD (Class III: No Benefit)

Amiodarone (Class IIb)

ICD (Class IIb)

SCD risk modifier† present

No

ICD (Class I)

Colors correspond to Class of Recommendation in Table 1.
See Section 7.4 for discussion.

*ICD candidacy as determined by functional status, life expectancy, or patient preference.
†Risk modifiers: Age <30 y, late gadolinium enhancement on cardiac MRI, LVOT obstruction, LV aneurysm, syncope >5 y.
BP indicates blood pressure; HCM, hypertrophic cardiomyopathy; Hx, history; ICD, implantable cardioverter-defibrillator;
LVOT, left ventricular outflow tract; LVWT, left ventricular wall thickness; MRI, magnetic resonance imaging; NSVT, nonsustained ventricular tachycardia; SCD, sudden cardiac death; and VT, ventricular tachycardia.
### 7.5. Myocarditis

**Recommendations for Myocarditis**

References that support the recommendations are summarized in Online Data Supplement 32.

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>C-LD</td>
<td>1. In patients with life-threatening VT or VF associated with confirmed or clinically suspected myocarditis, referral to centers with mechanical hemodynamic support and advanced arrhythmia management is recommended (1).</td>
</tr>
<tr>
<td>IIb</td>
<td>C-LD</td>
<td>2. In patients with giant cell myocarditis with VF or hemodynamically unstable VT treated according to GDMT, an ICD and/or an antiarrhythmic medication may be considered if meaningful survival of greater than 1 year is expected (2-4).</td>
</tr>
</tbody>
</table>
Figure 2. Survival without heart transplantation starting 1 year after initial GCM diagnosis.

Figure 3. Survival free of death, transplantation, GCM recurrence, heart failure, and ventricular arrhythmias starting 1 year after initial GCM diagnosis.
The cost-effectiveness of primary prophylactic implantable defibrillator therapy in patients with ischaemic or non-ischaemic heart disease: a European analysis

Tim Smith¹,², Luc Jordaens¹,³, Dominic A.M.J. Theuns¹, Pascal F. van Dessel³, Arthur A. Wilde³, and M.G. Myriam Hunink²,⁴,⁵

Conclusion

Our results suggest that primary prophylactic ICD therapy in patients with a left ventricular ejection fraction <40% and ischaemic or non-ischaemic heart disease is cost-effective in the European setting.
Ευχαριστώ