# 2014 ESC Guidelines for the Diagnosis & Management of Hypertrophic Cardiomyopathy

**The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC)**

Authors/Task Force members:

**Perry M. Elliott (Chairperson)**

# 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)**

**Authors/Task Force Members: Silvia G. Priori* (Chairperson)**
**Carina Blomström-Lundqvist* (Co-chairperson)**

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# HCM: Definition

*In an adult, HCM is defined by a wall thickness ≥ 15 mm in one or more LV myocardial segments—as measured by any imaging technique (echocardiography, cardiac magnetic resonance imaging (CMR) or computed tomography (CT))—that is not explained solely by loading conditions.*

Genetic and non-genetic disorders can present with lesser degrees of wall thickening (13–14 mm); in these cases, the diagnosis of HCM requires evaluation of other features including family history, non-cardiac symptoms and signs, electrocardiogram (ECG) abnormalities, laboratory tests and multi-modality cardiac imaging.

European Heart Journal (2014) **35**, 2733–2779

# Transthoracic echocardiography evaluation in hypertrophic cardiomyopathy

| Recommendations | Class | Level |
|---|---|---|
| In all patients with HCM at initial evaluation, transthoracic 2-D and Doppler echocardiography are recommended, at rest and during Valsalva manœuvre in the sitting and semi-supine positions—and then on standing if no gradient is provoked. | I | B |
| Measurement of maximum diastolic wall thickness is recommended, using 2-D short-axis views in all LV segments, from base to apex. | I | C |
| A comprehensive evaluation of LV diastolic function is recommended, including pulsed Doppler of mitral valve inflow, tissue Doppler velocities at the mitral annulus, pulmonary vein flow velocities, pulmonary artery systolic pressure, and measurement of LA size and volume. | I | C |
| In *symptomatic* patients with a resting or provoked peak instantaneous LV outflow tract gradient <50 mm Hg, 2-D and Doppler echocardiography *during exercise* in the standing, sitting or semi-supine position is recommended to detect provocable LVOTO and exercise induced mitral regurgitation. | I | B |


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# Cardiovascular magnetic resonance evaluation in hypertrophic cardiomyopathy

| Recommendations | Class | Level |
|---|---|---|
| It is recommended that CMR studies be performed and interpreted by teams experienced in cardiac imaging and in the evaluation of heart muscle disease. | I | C |
| In the absence of contra-indications, CMR with LGE is recommended in patients with suspected HCM who have inadequate echocardiographic windows, in order to confirm the diagnosis. | I | B |
| In the absence of contra-indications, CMR with LGE should be considered in patients fulfilling diagnostic criteria for HCM, to assess cardiac anatomy, ventricular function, and the presence and extent of myocardial fibrosis. | IIa | B |
| CMR with LGE imaging should be considered in patients with suspected apical hypertrophy or aneurysm. | IIa | C |
| CMR with LGE imaging should be considered in patients with suspected cardiac amyloidosis. | IIa | C |

**Fibrose myocardique non-prise en compte dans ESC-SCD score**


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# Non-compaction VG

## Définition: besoin de consensus

- Trabéculations volumineuses, multiples et profondes du sous-endocarde
- Recessus circulants inter/intra-trabéculaires
- Ratio épaisseur NC/C > 2 (2.3 en IRM!)

- Quelle étendue/Nb segments?

JA Towbin et al. Lancet 2015; 386 :813-825

# Non-compaction VG

Nombreuses variantes selon les anomalies anatomiques et fonctionnelles VG associées

- **Bénigne (NC isolée)** : épaisseurs pariétales, diamètres cavitaires, fonction systolique et diastolique normaux
- **Dilatée** : épaisseurs pariétales normales, cavité dilatée, dysfonction systolique

  Variante des CMD ?
- **Hypertrophique** : HVG septale, hyperkinésie

  Variante des CMH?
- **Hypertrophique et dilatée** : mauvais pronostic

  Evolution terminale d'une CMH mais survenue à un âge précoce
- **Restrictive**
- **Biventriculaire**

**Incidence pronostique propre ???**

JA Towbin et al. Lancet 2015; 386 :813-825

# Non-compaction VG

Many patients with LV non-compaction are completely asymptomatic, but some present with HF, thromboembolism, arrhythmias or SCD. Increased age, LV end diastolic diameter at presentation, symptomatic HF, permanent or persistent AF, bundle branch block and associated neuromuscular disease are reported predictors for increased mortality, but there are few data to suggest that LV non-compaction by itself is an indication for an ICD.[422–425] The need for an ICD should be guided by the severity of LV systolic dysfunction and the presence of sustained VA using the same criteria for DCM

**2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death**

European Heart Journal
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# Discussion

- **Diagnostic de la cardiopathie**
  - CMH
  - NCVG
- **Stratification du risque de MS:**
  - Investigations utiles à la stratification
  - ESC score
- **Choix thérapeutiques**
  - Traitement médical
  - DAI: oui/non
  - Type d'appareillage
- **Suivi**

# Investigations guidées par les symptomes

## Investigation of syncope

| Recommendations | Class | Level |
|---|---|---|
| 12–lead ECG, upright exercise test, resting and exercise 2-D and Doppler echocardiography, and 48–hour ambulatory ECG monitoring are recommended in patients with unexplained syncope, to identify the cause of their symptoms. | I | C |
| An ILR should be considered in patients with recurrent episodes of unexplained syncope, who are at low risk of SCD. | IIa | C |

## Palpitations

| Recommendations | Class | Level |
|---|---|---|
| For patients with frequent or sustained palpitations,48–hour ambulatory ECG monitoring is recommended, to identify the likely cause. | I | C |
| An ILR may be considered in patients with frequent palpitations, in whom no cause is identified following prolonged ECG monitoring. | IIb | C |

# Cardiopulmonary exercise testing

| Recommendations | Class | Level |
|---|---|---|
| Cardiopulmonary exercise testing, with simultaneous measurement of respiratory gases, is recommended in severely symptomatic patients with systolic and/or diastolic LV dysfunction being evaluated for heart transplantation or mechanical support. | I | B |
| Irrespective of symptoms, cardiopulmonary exercise testing with simultaneous measurement of respiratory gases (or standard treadmill or bicycle ergometry when unavailable) should be considered to assess the severity and mechanism of exercise intolerance and change in systolic blood pressure. | IIa | B |
| Cardiopulmonary exercise testing, with simultaneous measurement of respiratory gases (or standard treadmill or bicycle ergometry when unavailable), should be considered in symptomatic patients undergoing septal alcohol ablation and septal myectomy to determine the severity of exercise limitation. | IIa | C |

**Non-systématique: la réponse tensionnelle à l'effort n'est plus utilisée pour la stratification du risque**


European Heart Journal (2014);35:2733–2779 - doi:10.1093/eurheartj/ehu284

# Electrophysiologic testing

| Recommendations | Class | Level |
|---|---|---|
| Invasive electrophysiological study is recommended in patients with documented persistent or recurrent supraventricular tachycardia (atrial flutter, atrial tachycardia, atrioventricular nodal re-entry tachycardia, accessory atrioventricular pathway mediated tachycardias) and in patients with ventricular pre-excitation, in order to identify and treat an ablatable substrate. | I | C |
| Invasive electrophysiological study may be considered in selected patients with documented, symptomatic, monomorphic, sustained (>30 s) ventricular tachycardia in order to identify and treat an ablatable arrhythmia substrate. | IIb | C |
| Invasive electrophysiological study with programmed ventricular stimulation is not recommended for sudden cardiac death risk stratification. | III | C |


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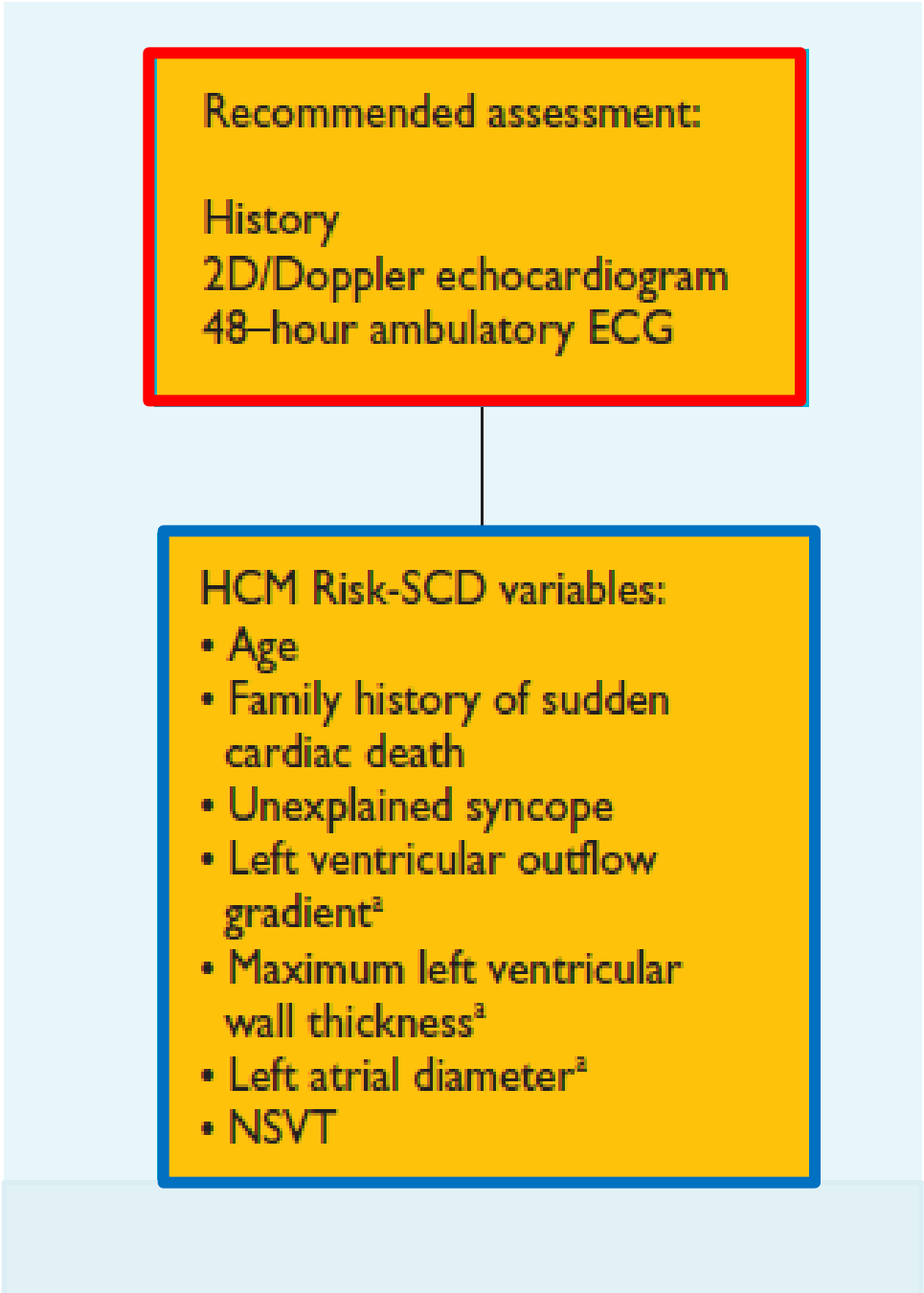
Recommended assessment:
History
2D/Doppler echocardiogram
48–hour ambulatory ECG
HCM Risk-SCD variables:
• Age
• Family history of sudden cardiac death
• Unexplained syncope
• Left ventricular outflow gradient[a]
• Maximum left ventricular wall thickness[a]
• Left atrial diameter[a]
• NSVT

# 5-year risk of SCD using the HCM Risk-SCD model

**Probability** $_{\text{SCD at 5 years}}$ **= 1-0.998** $^{\exp(\text{Progostic index})}$

where Prognostic index = [0.15939858 x maximal wall thickness (mm)] – [0.00294271 x maximal wall thickness$^2$ (mm$^2$)] + [0.0259082 x left atrial diameter (mm)] + [0.00446131 x maximal (rest/Valsalva) left ventricular outflow tract gradient (mm Hg)] + [0.4583082 x family history SCD] + [0.82639195 x NSVT] + [0.71650361 x unexplained syncope] – [0.01799934 x age at clinical evaluation (years)].


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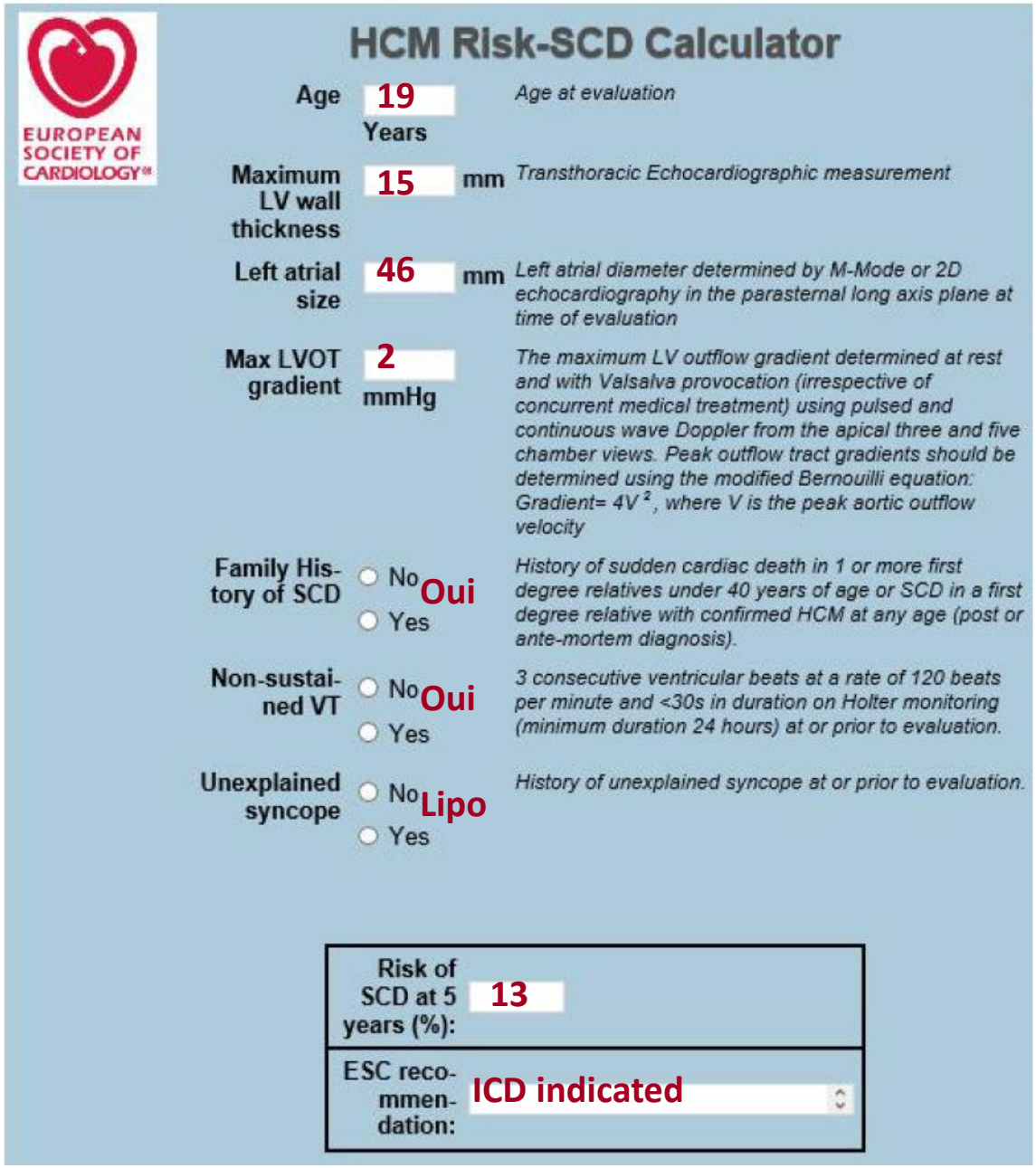
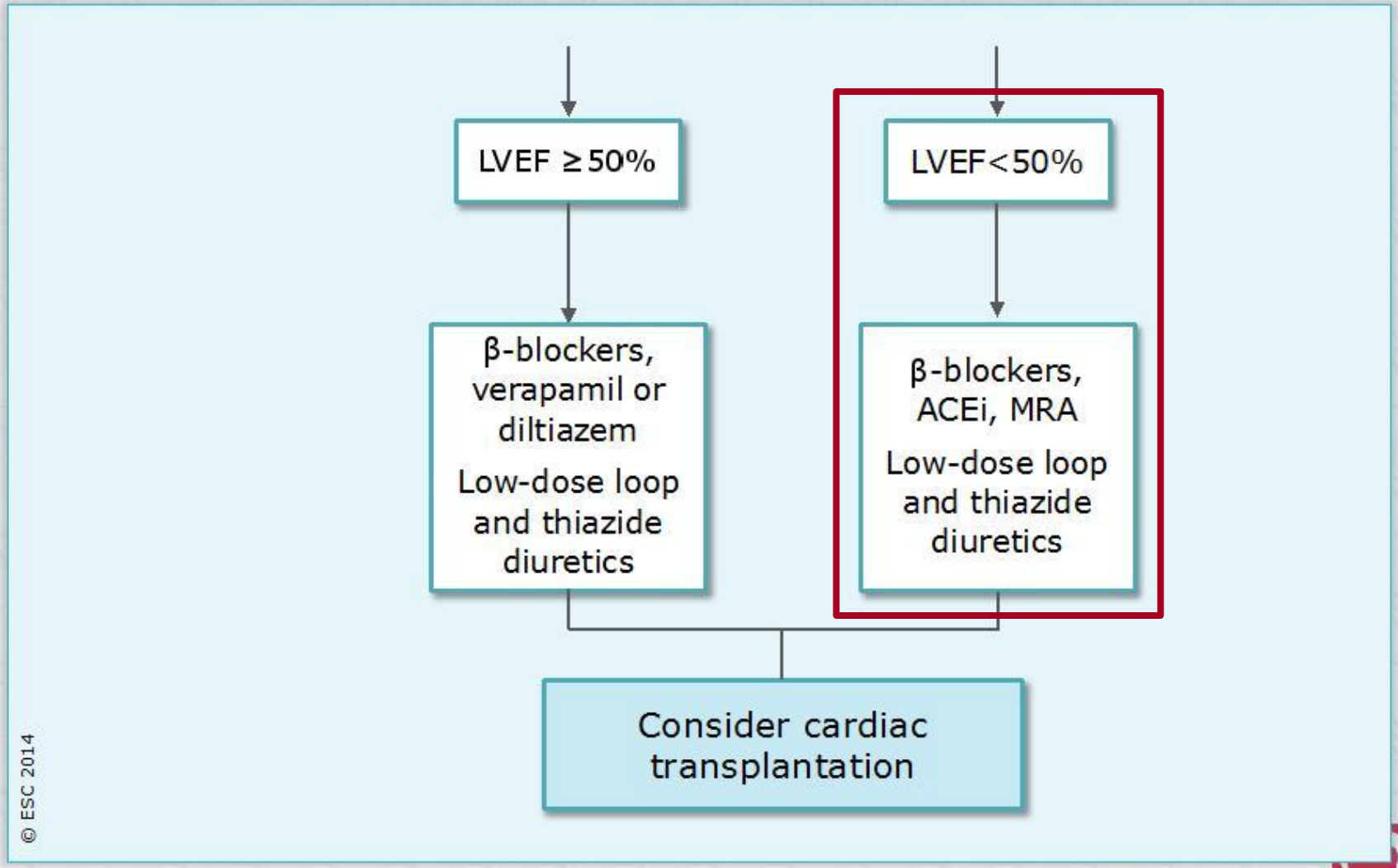
# Estimation du risque de mort subite à 5 ans



# Discussion

- **Diagnostic de la cardiopathie**
  - CMH
  - NCVG
- **Stratification du risque de MS:**
  - Instruments de la stratification
  - ESC score
- **Choix de traitement**
  - Traitement médical: FEVG=40%; pas d'obst.
  - DAI: oui/non
  - Type d'appareillage
- **Suivi**

# Algorithm for the treatment of heart failure in hypertrophic cardiomyopathy *(Cont.)*







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# Patients with heart failure and reduced LV ejection fraction (<50%)

| Recommendations | Class | Level |
|---|---|---|
| An ACE-inhibitor (or ARB if ACE inhibitor not tolerated) should be considered, in addition to a ß-blocker, for patients without LVOTO who have an LVEF <50%, to reduce the risk of HF hospitalization and risk of premature death[a]. | IIa | C |
| A ß-blocker should be considered, in addition to an ACE-inhibitor (or ARB if ACE-inhibitor not tolerated), for patients without LVOTO who have an LVEF <50% to improve symptoms and reduce the risks of HF hospitalization and premature death[a]. | IIa | C |
| Low-dose loop diuretics should be considered for symptomatic patients in NYHA functional Class II–IV with an LVEF <50%, to improve symptoms and reduce the risk of HF hospitalization[a]. | IIa | C |
| For all patients with persisting symptoms (NYHA functional Class II–IV) and an LVEF <50%—despite treatment with an ACE-inhibitor (or an ARB if an ACE-inhibitor is not tolerated) and a ß-blocker—a mineralocorticoid receptor antagonist (MRA) should be considered, to reduce the risks of HF hospitalization and premature death[a]. | IIa | C |

[a]In the absence of randomized trials in HCM, the benefit on hospitalization, symptoms and mortality is assumed but unproven.


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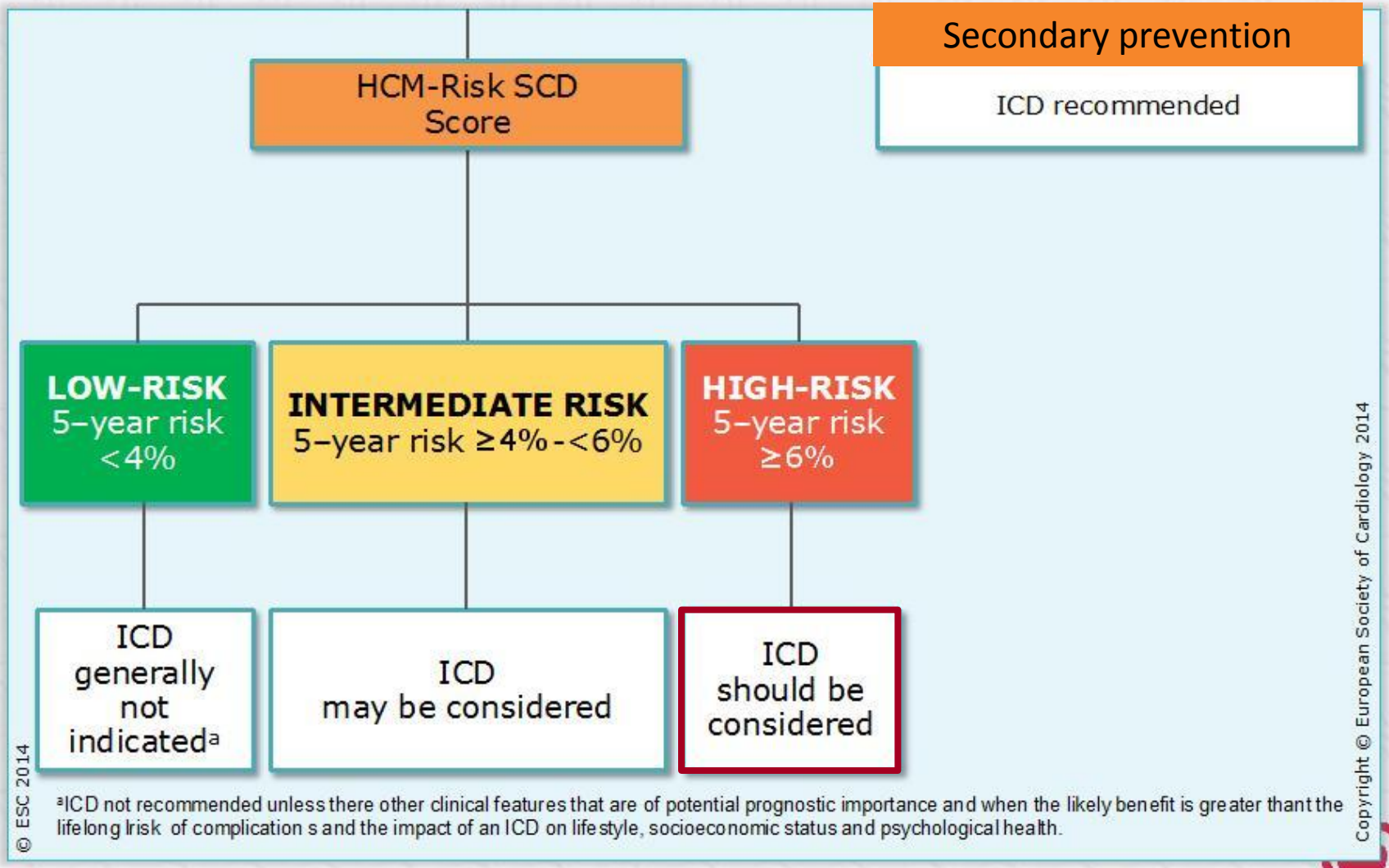
# Flow chart for ICD implantation





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# Prevention of sudden cardiac death

| Recommendations | Class | Level |
|---|---|---|
| Avoidance of competitive sports is recommended in patients with HCM. | I | C |
| ICD implantation is recommended in patients who have survived a cardiac arrest due to VT or VF, or who have spontaneous sustained VT causing syncope or haemodynamic compromise, and have a life expectancy of >1 year. | I | B |
| HCM Risk-SCD is recommended as a method of estimating risk of sudden death at 5 years in patients aged ≥16 years without a history of resuscitated VT/VF or spontaneous sustained VT causing syncope or haemodynamic compromise. | I | B |
| It is recommended that the 5-year risk of SCD be assessed at first evaluation and re-evaluated at 1–2 yearly intervals or whenever there is a change in clinical status. | I | B |
| ICD implantation should be considered in patients with an estimated 5-year risk of sudden death of ≥6% and a life expectancy of >1 year, following detailed clinical assessment that takes into account the lifelong risk of complications and the impact of an ICD on lifestyle, socio-economic status and psychological health. | IIa | B |


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# Prevention of sudden cardiac death *(Cont.)*

| Recommendations | Class | Level |
|---|---|---|
| ICD implantation may be considered in individual patients with an estimated 5–year risk of SCD of between ≥4% and <6% and a life expectancy of >1 year following detailed clinical assessment that takes into account the lifelong risk of complications and the impact of an ICD on lifestyle, socio-economic status and psychological health. | **IIb** | **B** |
| ICD implantation may be considered in individual patients with an estimated 5–year risk of SCD of <4% only when they have clinical features that are of proven prognostic importance, and when an assessment of the lifelong risk of complications and the impact of an ICD on lifestyle, socio-economic status and psychological health suggests a net benefit from ICD therapy. | **IIb** | **B** |
| ICD implantation is not recommended in patients with an estimated 5–year risk of SCD of <4% and no other clinical features that are of proven prognostic importance. | **III** | **B** |


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# Practical aspects of implantable cardioverter defibrillator therapy

| Recommendations | Class | Level |
|---|---|---|
| Prior to ICD implantation, patients should be counselled on the risk of inappropriate shocks, implant complications and the social, occupational, and driving implications of the device. | I | C |
| ß-Blockers and/or amiodarone are recommended in patients with an ICD, who have symptomatic ventricular arrhythmias or recurrent shocks despite optimal treatment and device re-programming. | I | C |
| Electrophysiological study is recommended in patients with ICD, and inappropriate shocks due to regular supraventricular tachycardias, to identify and treat any ablatable arrhythmia substrate. | I | C |
| A subcutaneous ICD lead system (S-ICD™) may be considered in HCM patients who do not have an indication for pacing. | IIb | C |

**Simple ou double-chambre? Tt associé de l'obstruction**
**Endocavitaire ou sous-cutané?**


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# DAI double-chambre: Stimulation DDD pour traiter l'obstruction

| | | |
|---|---|---|
| In patients with resting or provocable LVOTO ≥50 mm Hg, sinus rhythm and drug-refractory symptoms, in whom there is an indication for an ICD, a dual-chamber ICD (instead of a single-lead device) may be considered, to reduce the LV outflow tract gradient or to facilitate medical treatment with ß-blockers and/or verapamil . | IIb | C |

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# Recommandations:

- Définition et diagnostic

- Stratification du risque

- Traitement

- Cardiopathies ischémiques, incluant SCA
- Dysfonction VG/Insuffisance cardiaque
- Cardiomyopathies:
  - CMD
  - CMH
  - DAVD…
- Canalopathies
  - QT long
  - QT court
  - Brugada
  - TV catécholergiques
  - Repolarisation précoce …
- TV/FV sur « cœur sain »
- Enfant et cardiopathies congénitales

…

# Diagnostic workup in patients presenting with sustained ventricular tachycardia or ventricular fibrillation.