Υπερτροφική μυοκαρδιοπάθεια: ποιος κινδυνεύει?

Δέσποινα Παρχαρίδου
Καρδιολόγος
Επιστημονικός Συνεργάτης
Α Καρδιολογική Κλινική ΑΧΕΠΑ
Ιατρείο Μυοκαρδιοπαθειών
1st Case

- Female, 41 yo
- HCM first diagnosis → 2008
- Reason for diagnosis → check up
- Lost to follow up
- New consultation at outpatient clinic → 11/2018
- NYHA II
- No syncope
- No angina
- Palpitations

- Medical treatment → b-blocker (bisoprolol)

- Holter → **NSVT** (4 QRS)
- Stable increase in hs-cTnT
Family tree
Fibrosis 29%

<table>
<thead>
<tr>
<th>Description</th>
<th>Measurement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Μεσοκοιλιακό διάφραγμα</td>
<td>30mm</td>
</tr>
<tr>
<td>Πρόσθιο τοίχωμα</td>
<td>24mm</td>
</tr>
<tr>
<td>Πλάγιο τοίχωμα</td>
<td>17mm</td>
</tr>
<tr>
<td>Κατώτερο τοίχωμα</td>
<td>18mm</td>
</tr>
</tbody>
</table>
MAGNITUDE OF LEFT VENTRICULAR HYPERTROPHY AND RISK OF SUDDEN DEATH IN HYPERTROPHIC CARDIOMYOPATHY

PAOLO SPIRITO, M.D., PIETRO BELLONE, M.D., KEVIN M. HARRIS, M.D., PAOLA BERNABÒ, M.D., PAOLO BRUZZI, M.D., PH.D., AND BARRY J. MARON, M.D.

480 consecutive patients
# Prognostic Value of Quantitative Contrast-Enhanced Cardiovascular Magnetic Resonance for the Evaluation of Sudden Death Risk in Patients With Hypertrophic Cardiomyopathy

Raymond H. Chan, MD, MPH; Barry J. Maron, MD; Iacopo Olivotto, MD;
Significance of High-Sensitivity Cardiac Troponin T in Hypertrophic Cardiomyopathy

183 HCM patients/ 99 abnormal troponine
CV deaths, HF, VT, embolic, NYHA progression

Table 4: Cardiovascular Events in 183 Patients With Normal hs-cTnT Values and With Abnormal hs-cTnT Values (Univariate Analysis)

<table>
<thead>
<tr>
<th>Event Type</th>
<th>Normal hs-cTnT (n = 84)</th>
<th>Abnormal hs-cTnT (n = 99)</th>
<th>Hazard Ratio</th>
<th>95% CI</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>All cardiovascular events</td>
<td>6</td>
<td>32</td>
<td>5.05</td>
<td>2.11–12.09</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Cardiac deaths</td>
<td>0</td>
<td>9</td>
<td>NA</td>
<td>NA</td>
<td>—</td>
</tr>
<tr>
<td>Cardiac events</td>
<td>2</td>
<td>27</td>
<td>12.56</td>
<td>2.99–52.82</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Heart failure events</td>
<td>2</td>
<td>24</td>
<td>11.16</td>
<td>2.64–47.21</td>
<td>0.001</td>
</tr>
<tr>
<td>Arrhythmic events</td>
<td>1</td>
<td>7</td>
<td>5.81</td>
<td>0.72–47.30</td>
<td>0.100</td>
</tr>
</tbody>
</table>
## Table 2. Risk Factors for Sudden Death in HCM*

<table>
<thead>
<tr>
<th>Major</th>
<th>Possible in Individual Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac arrest (ventricular fibrillation)</td>
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</tr>
<tr>
<td>Unexplained syncope</td>
<td>High-risk mutation</td>
</tr>
<tr>
<td><strong>LV thickness greater than or equal to 30 mm</strong></td>
<td>Intense (competitive) physical exertion</td>
</tr>
<tr>
<td>Abnormal exercise blood pressure</td>
<td></td>
</tr>
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<td>Nonsustained ventricular tachycardia (Holter)</td>
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</tbody>
</table>
2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy: Executive Summary
A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines

LVOTO
Late Gad
Apical Aneurysm
Compound mutations
A validation study of the 2003 American College of Cardiology/European Society of Cardiology and 2011 American College of Cardiology Foundation/American Heart Association risk stratification and treatment algorithms for sudden cardiac death in patients with hypertrophic cardiomyopathy

Constantinos O’Mahony,¹,² Maite Tome-Esteban,¹ Pier D Lambiase,¹ Antonios Pantazis,¹ Shaughan Dickie,¹ William J McKenna,¹ Perry M Elliott¹

1606 patients retrospective
A validation study of the 2003 American College of Cardiology/European Society of Cardiology and 2011 American College of Cardiology Foundation/American Heart Association risk stratification and treatment algorithms for sudden cardiac death in patients with hypertrophic cardiomyopathy.

Constantinos O'Mahony,¹,² Maite Tome-Esteban,¹ Pier D Lambiase,¹ Antonios Pantazis,¹ Shaughan Dickie,¹ William J McKenna,¹ Perry M Elliott¹
Flow chart for ICD implantation

HCM-Risk SCD Score

LOW-RISK
5-year risk <4%
- ICD generally not indicated

INTERMEDIATE RISK
5-year risk ≥4% - <6%
- ICD may be considered

HIGH-RISK
5-year risk ≥6%
- ICD should be considered

ICD recommended

*ICD not recommended unless there other clinical features that are of potential prognostic importance and when the likely benefit is greater than the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.
# HCM Risk-SCD calculator

## HCM Risk-SCD Calculator

<table>
<thead>
<tr>
<th><strong>Parameter</strong></th>
<th><strong>Value</strong></th>
<th><strong>Description</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>41 Years</td>
<td>Age at evaluation</td>
</tr>
<tr>
<td>Maximum LV wall thickness</td>
<td>30 mm</td>
<td>Transthoracic Echocardiographic measurement</td>
</tr>
<tr>
<td>Left atrial size</td>
<td>30 mm</td>
<td>Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation</td>
</tr>
<tr>
<td>Max LVOT gradient</td>
<td>27 mmHg</td>
<td>The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernoulli equation: ( \text{Gradient} = 4V^2 ), where ( V ) is the peak aortic outflow velocity</td>
</tr>
<tr>
<td>Family History of SCD</td>
<td>No / Yes</td>
<td>History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).</td>
</tr>
<tr>
<td>Non-sustained VT</td>
<td>No / Yes</td>
<td>3 consecutive ventricular beats at a rate of 120 beats per minute and &lt;30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.</td>
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<tr>
<td>Unexplained syncope</td>
<td>No / Yes</td>
<td>History of unexplained syncope at or prior to evaluation.</td>
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## Risk of SCD at 5 years (%): 4.43

**ESC recommendation:** ICD may be considered
Effectiveness of the 2014 European Society of Cardiology guideline on sudden cardiac death in hypertrophic cardiomyopathy: a systematic review and meta-analysis

Constantinos O’Mahony,1,2,3 Mohammed Majid Akhtar,1,3,2 Zacharias Anastasiou,4 Oliver P Guttmann,1,3,2 Pieter A Vriesendorp,5 Michelle Michels,5 Damiano Magri,6 Camillo Autore,6 Adrián Fernández,7 Juan Pablo Ochoa,7,8,9 Kevin M W Leong,10 Amanda M Varnava,10 Lorenzo Monserrat,9,11 Aristides Anastasakis,12 Pablo Garcia-Pavia,3,13,14,15 Claudio Rapezzi,16 Elena Biagini,16 Juan Ramon Gimeno,3,17 Giuseppe Limongelli,3,18 Rumana Z Omar,4 Perry M Elliott1,3,2

six retrospective observational studies  653 articles

<table>
<thead>
<tr>
<th></th>
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</tr>
</thead>
<tbody>
<tr>
<td>Patients (n)</td>
<td>3675</td>
<td>706</td>
<td>623*</td>
<td>502</td>
<td>288+</td>
<td>3703</td>
</tr>
<tr>
<td>Centres (n)</td>
<td>6</td>
<td>2</td>
<td>5</td>
<td>1</td>
<td>1</td>
<td>14</td>
</tr>
</tbody>
</table>

Heart 2018
High-risk patients contributed 51% of SCD endpoints; 68% of all SCD endpoints occurred in patients with an estimated 5-year risk of ≥4% (intermediate and high risk group).

The intermediate risk group had a relatively low prevalence of SCD endpoints and the meta-analysis suggests that the risk in these patients is overestimated.
Major Risk Markers
- Family history of HCM-related sudden death
- Unexplained syncope
- Multiple, repetitive NSVT
- Massive LVH (≥30 mm)
- LV apical aneurysm
- Extensive LGE
- End stage (ejection fraction <50%)

Potential Risk Mediators
- Hypotensive response to exercise
- Marked LV outflow obstruction at rest
- Alcohol septal ablation (?)
- Reduced risk: age ≥60 yr

Increased Risk
(≥1 major marker alone or with mediator)

Consideration of primary prevention with ICD

Appropriate intervention (VT or VF), 4% per yr

Low risk of heart failure after ICD intervention (<1% per yr)
• Consultation for ICD implantation
2\textsuperscript{nd} Case

- Man 52 yo

- Date of first diagnosis 1985 (19 yo)

- Reason for diagnosis → check up before joining the army
  no symptoms before diagnosis

- Played football in a team after the diagnosis of HCM
  no syncope or other symptoms during exercise
• **Syncope** → 5 episodes
  1\textsuperscript{st} episode 2000, last episode 2004
during rest, never in exertion
feeling of tachycardia after the episodes
Holter → no NSVT

• Loop recorder → **NSVT** (presyncope)
• NYHA II
• Angina like
• Palpitations
Family tree
ECG
Patients with a recent unexplained syncope (within 6 months before initial evaluation) had a relative risk of sudden death 5-fold higher than patients without syncope.

- 1500 HCM patients
- 153 patients with unexplained syncope

*Circulation* 2009
not all NSVT runs have the same predictive power

160 ICD patients

Circ Arrhythm Electrophysiol. 2017
Table 2. Risk Factors for Sudden Death in HCM*

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Risk assessment ACCF/ AHA 2011

Flowchart:
- **Prior cardiac arrest or Sustained VT**
  - Yes: ICD recommended
  - No:
    - **Family history-SD in first-degree relative or LV wall thickness >20 mm or Recent unexplained syncope**
      - Yes: ICD reasonable
      - No: Nonsustained VT
    - Nonsustained VT Abnormal BP response
      - Yes: Other SCD Risk Modifiers Present?
        - Yes: ICD can be useful
        - No: Role of ICD uncertain
      - No: ICD not recommended

Legend:
- Class I
- Class IIa
- Class IIb
- Class III

LVOTO: Late Gad Apical Aneurysm Compound mutations
HCM Risk-SCD Calculator

**Age** 52 Years  
*Age at evaluation*

**Maximum LV wall thickness** 18 mm  
*Transthoracic Echocardiographic measurement*

**Left atrial size** 35 mm  
*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*

**Max LVOT gradient** 15 mmHg  
*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernoulli equation: Gradient = 4V^2, where V is the peak aortic outflow velocity*

**Family History of SCD**  
- No
- Yes  
*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis)*

**Non-sustained VT**  
- No
- Yes  
*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation*

**Unexplained syncope**  
- No
- Yes  
*History of unexplained syncope at or prior to evaluation*

---

**Risk of SCD at 5 years (%):** 6.39

**ESC recommendation:** ICD should be considered
Barry J. Maron, NEJM 2018
• ICD 2004
• 2 appropriate interventions many years after ICD implantation (1st episode 2012)
3rd Case

- Man, 52 yo

- Date of first diagnosis → 2005 (39 yo)

- Reason for diagnosis → check up

- **History of sudden death** → sister 16 yo (while sleeping) father 48 yo (while working)

- Played football (16-21 yo) → no symptoms
• NYHA II
• No syncope
• No angina
• No palpitations
• Holter → no NSVT

• Exercise test → abnormal arterial pressure response (40 yo)
  failure to increase by at least 20 mm Hg or a drop of at least 20 mm Hg during effort (1/3 patients)
Family tree
ECG
It has been recognized that SCD events can cluster in families. Notably, some studies have not demonstrated an independent link between family history of SCD and risk for individual patients on multivariate analysis, whereas others have suggested that family history is an independent predictor. These differences may be explained in part by the relative infrequency of events but also likely reflect variability in the definition of a family history of SCD.
LVOTO had a low positive predictive accuracy for SCD; however there was an interaction between outflow tract obstruction and other risk markers, suggesting that LVOTO modifies the expression of the underlying arrhythmogenic substrate in patients already predisposed to sudden death.
SCD Risk after myectomy

Single tertiary centre
1809 patients
64% myectomy

Primary composite endpoint
SCD and appropriate ICD discharge

Desai MY J Thor and Cardiovasc Surgery 2018
Why?

• Severe obstruction increases LV wall stress and reduce myocardial perfusion with resultant myocardial ischemia, which could potentially trigger malignant ventricular arrhythmias in already predisposed patients

• long-standing pressure overload could lead to deleterious additional myocardial hypertrophy and fibrosis

• An extensive surgical myectomy removes a substantial amount of hypertrophied muscle. There is an association between LV hypertrophy and myocardial fibrosis

Desai MY, *J Thor and Cardiovasc Surgery* 2018
<table>
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<td>Nonsustained ventricular tachycardia (Holter)</td>
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</tr>
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</table>
Risk assessment ACCF/ AHA 2011

- Prior cardiac arrest or Sustained VT
  - Yes → ICD recommended
  - No → Family history-SD in first-degree relative or LV wall thickness ≥30 mm or Recent unexplained syncope
    - Yes → ICD reasonable
    - No → Nonsustained VT
      - Abnormal BP response
        - Yes → Other SCD Risk Modifiers* Present?
          - Yes → ICD can be useful
          - No → Role of ICD uncertain
        - No → ICD not recommended
      - No → ICD not recommended

Legend:
- Class I
- Class IIa
- Class IIb
- Class III

LVOTO
- Late Gad
- Apical Aneurysm
- Compound mutations
## HCM Risk- SCD calculator

<table>
<thead>
<tr>
<th></th>
<th>Value</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>39</td>
<td>Years</td>
</tr>
<tr>
<td>Maximum LV wall thickness</td>
<td>16</td>
<td>mm</td>
</tr>
<tr>
<td>Left atrial size</td>
<td>46</td>
<td>mm</td>
</tr>
<tr>
<td>Max LVOT gradient</td>
<td>64</td>
<td>mmHg</td>
</tr>
<tr>
<td>Family History of SCD</td>
<td>Yes</td>
<td>History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).</td>
</tr>
<tr>
<td>Non-sustained VT</td>
<td>Yes</td>
<td>3 consecutive ventricular beats at a rate of 120 beats per minute and &lt;30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.</td>
</tr>
<tr>
<td>Unexplained syncope</td>
<td>Yes</td>
<td>History of unexplained syncope at or prior to evaluation.</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Risk of SCD at 5 years (%)</th>
<th>4.06</th>
</tr>
</thead>
<tbody>
<tr>
<td>ESC recommendation</td>
<td>ICD may be considered</td>
</tr>
</tbody>
</table>
• ICD 2006 (40 yo)
• No appropriate discharges
• 2 inappropriate discharges due to AF

• ICD infection 2010
• ICD and leads removal/ antibiotics
• New ICD
4th case

- Female, 51 yo
- HCM first diagnosis → 2013
- Reason for diagnosis → palpitations

Medication: bisoprolol

- 2016 AHEPA Cardiomyopathy outpatient clinic
• NYHA II
• Palpitations
• No FH of sudden death
• Holter → NSVT (12 QRS)
• No syncope
HKГ
## Table 2. Risk Factors for Sudden Death in HCM*

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<td></td>
</tr>
<tr>
<td>(Holter)</td>
<td></td>
</tr>
</tbody>
</table>
Risk assessment ACCF/ AHA 2011
# ESC risk score

## HCM Risk-SCD Calculator

<table>
<thead>
<tr>
<th>Factor</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>51 Years</td>
</tr>
<tr>
<td>Maximum LV wall thickness</td>
<td>17 mm</td>
</tr>
<tr>
<td>Left atrial size</td>
<td>44 mm</td>
</tr>
<tr>
<td>Max LVOT gradient</td>
<td>17 mmHg</td>
</tr>
<tr>
<td>Family History of SCD</td>
<td>No</td>
</tr>
<tr>
<td>Non-sustained VT</td>
<td>No</td>
</tr>
<tr>
<td>Unexplained syncope</td>
<td>No</td>
</tr>
</tbody>
</table>

**Age at evaluation**

**Transesophageal Echocardiographic measurement**

**Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation**

The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernoulli equation: \( \text{Gradient} = 4V^2 \), where \( V \) is the peak aortic outflow velocity.

**Family History of SCD**

History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).

**Non-sustained VT**

3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.

**Unexplained syncope**

History of unexplained syncope at or prior to evaluation.

<table>
<thead>
<tr>
<th>Risk of SCD at 5 years (%)</th>
<th>3.88</th>
</tr>
</thead>
<tbody>
<tr>
<td>ESC recommendation</td>
<td>ICD generally not indicated</td>
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</table>

*ICD not recommended unless there other clinical features that are of potential prognostic importance and when the likely benefit is greater than the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.*
Hypertrophic Cardiomyopathy With Left Ventricular Apical Aneurysm
Implications for Risk Stratification and Management

1.940 patients
4.8% apical aneurysm

SD event rate of almost 5%/year

20% life-saving ICD interventions for VT/VF

J Am Coll Cardiol 2017
• ICD implantation
• Electrical storm → ICD discharges
• VT ablation
5th case

- Male, 40 yo

- First presentation → Cardiac arrest (VF)

- No symptoms
- NYHA class I
- No FH for cardiomyopathy/ SD
- Normal coronary arteries
- Holter during hospitalization → normal
ECG (after cardioversion)
Table 2. Risk Factors for Sudden Death in HCM

**Major**
- Cardiac arrest (ventricular fibrillation)
- Spontaneous sustained ventricular tachycardia
- Family history of premature sudden death
- Unexplained syncope
- LV thickness greater than or equal to 30 mm
- Abnormal exercise blood pressure
- Nonsustained ventricular tachycardia (Holter)

**Possible in Individual Patients**
- Atrial fibrillation
- Myocardial ischemia
- LV outflow obstruction
- High-risk mutation
- Intense (competitive) physical exertion

**Major Risk Markers**
- Family history of HCM-related sudden death
- Unexplained syncope
- Multiple, repetitive NSVT
- Massive LVH (≥30 mm)
- LV apical aneurysm
- Extensive LGE
- End stage (ejection fraction <50%)

**Potential Risk Mediators**
- Hypotensive response to exercise
- Marked LV outflow obstruction at rest
- Alcohol septal ablation (?)
- Reduced risk: age ≥60 yr

**Increased Risk**
(≥1 major marker alone or with mediator)

**Consideration of Primary Prevention with ICD**

**Appropriate Intervention (VT or VF), 4% per yr**

**Low Risk of Heart Failure after ICD Intervention (<1% per yr)**
ESC risk score

**HCM Risk-SCD Calculator**

- **Age**: 40 Years
- **Maximum LV wall thickness**: 15 mm
- **Left atrial size**: 48 mm
- **Max LVOT gradient**: 16 mmHg
- **Family History of SCD**: No
- **Non-sustained VT**: No
- **Unexplained syncope**: No

**Risk of SCD at 5 years (%)**: 2.02

**ESC recommendation**: ICD generally not indicated **

**Note**: ICD not recommended unless there are other clinical features that are of potential prognostic importance and when the likely benefit is greater than the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.
>650 patients
- none of the conventional HC risk factors for sudden death
- no or mild symptoms (NYHA class I or II)
- no history of AF
- no previous surgical myectomy, alcohol septal ablation, or ICD
- no clinical features compatible with evolution to the end-stage phase of HC

At the time of SD
- SD 0.6% per year
- no/ mild symptoms
- 50% were aged <40 y
5.100 consecutive cases of SCD
196, (4%) HCM deaths
1994-2017

Sudden Death Can Be the First Manifestation of Hypertrophic Cardiomyopathy

Data From a United Kingdom Pathology Registry

JACC: CLINICAL ELECTROPHYSIOLOGY
FEBRUARY 2019:252-5

78% diagnoses made at autopsy

Cardiac symptoms in 26%

Death during exercise or emotional stress in 23%

Average age: 43±18
10% > 60 yrs old

73% males

Myocardial fibrosis in 59%

CAD in 16%

Heart weight 558 ± 186 g
MWT 21.6 ± 6.3 mm
Take home messages

• risk of sudden death remains the most feared consequence
• it is now possible to achieve significantly improved survival with a low HCM-related mortality of 0.5% per year across all ages
• Many ‘grey areas’ of ambiguity
• Difficulty in translating guideline language into real-world clinical decisions for individual patients
Many thanks