Exercise Guidelines in Cardiomyopathy

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Conflict of Interest Declared

None
Hypertrophic Cardiomyopathy
Causes of Sudden Cardiac Death in Athletes in Relation to Age

- HCM: 48%
- CAD: 10%
- Unexplained: 3%
- Ruptured Aorta: 7%
- Idiopathic LVH: 14%
- CAA: 18%
Exercise Related Deaths in HCM

90% occur during or immediately after exercise

Soccer and basketball

Males > Females 9:1

Blacks > Whites 7:1
Small Vessel Disease

Myocardial disarray

LVH with non-dilated LV cavity

Myocardial disarray

Small Vessel Disease
<table>
<thead>
<tr>
<th>Category</th>
<th>Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>General Population</td>
<td>0.2%</td>
</tr>
<tr>
<td>Competitive athletes</td>
<td>0.07%</td>
</tr>
<tr>
<td>Elite National athletes</td>
<td>0.03%</td>
</tr>
</tbody>
</table>
Abstinence from strenous exertion is essential to minimise the risk of sudden cardiac death.
Pertinent Issues

Strong association between exercise and SCD

Heterogeneous Disorder

Incomplete Penetrance

Gene testing may be inconclusive or misleading

Mild morphology

Risk stratification based on laboratory tests
Risk Pyramid in HCM

- **Low Risk**
  - No risk factors

- **Intermediate Risk**
  - 1 risk factor
  - Individualised therapy

- **High Risk**
  - Aborted sudden death
  - Sustained VT
  - Multiple risk factors

**ICD**
Potential triggers for Sudden Death

- Supraventricular arrhythmias
- Myocardial ischaemia
- LVOT obstruction
- Abnormal Vascular Responses
- Dehydration
- Adrenergic surges
- Electrolyte imbalance
- Acid/base disturbance

Unstable Electrophysiological Substrate
Exercise Guidelines in HCM

Conservative

Homogeneous

Encompass all preventable deaths
Guidelines

Bethesda (AHA)

European Society of Cardiology
Individuals with Unequivocal HCM or High Probability of HCM

Bethesda Guidelines (American)

Participation in class 1A sport (low intensity and low dynamic)

ESC Guidelines

No competitive sports if symptoms or any risk factors for sudden death. Recreational sport only.

Class IA sport (low intensity and low dynamic) in those with no symptoms or risk factors
## Sports Permitted in Most Individuals with Cardiomyopathy

<table>
<thead>
<tr>
<th>LEISURE ACTIVITY</th>
<th>COMPETITIVE ACTIVITY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stationary bicycle</td>
<td>Golf</td>
</tr>
<tr>
<td>Bowling</td>
<td>Archery</td>
</tr>
<tr>
<td>Brisk walking</td>
<td>Bowling</td>
</tr>
<tr>
<td>Golfing</td>
<td>Cricket</td>
</tr>
<tr>
<td>Moderate hiking</td>
<td></td>
</tr>
<tr>
<td>Skating</td>
<td></td>
</tr>
<tr>
<td>Tennis (doubles)</td>
<td></td>
</tr>
<tr>
<td>Treadmill</td>
<td></td>
</tr>
<tr>
<td>Low-intensity weights</td>
<td></td>
</tr>
</tbody>
</table>
Implications of the Guidelines in Athletic Individuals

1. Diagnosis of HCM in athletes
2. Isolated ECG abnormalities outside the context of a family history of HCM
3. Gene positive/phenotype negative individuals participating in regular sports
4. Individuals with ICD and individuals with therapeutic abolition of LV outflow gradient
Diagnosis of HCM is Based on Echocardiography

Left ventricular Hypertrophy ($\geq 13$ mm in adults and $> 11$ mm in adolescents) in association with a non-dilated LV cavity.
### Athlete’s Heart or HCM in Athlete with LVH ≥ 13 mm

<table>
<thead>
<tr>
<th>Feature</th>
<th>Athlete</th>
<th>HCM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bizarre patterns of LVH</td>
<td>_</td>
<td>+</td>
</tr>
<tr>
<td>LV cavity &gt; 54 mm</td>
<td>_</td>
<td>+</td>
</tr>
<tr>
<td>LA &gt; 50 mm</td>
<td>_</td>
<td>_</td>
</tr>
<tr>
<td>LV outflow obstruction</td>
<td>_</td>
<td>_</td>
</tr>
<tr>
<td>Impaired diastolic function</td>
<td>_</td>
<td>_</td>
</tr>
<tr>
<td>Isolated Sokolow-Lyon LVH</td>
<td>+</td>
<td>_</td>
</tr>
<tr>
<td>ST depression/Deep T wave inversion</td>
<td>_</td>
<td>_</td>
</tr>
<tr>
<td>Female gender</td>
<td>_</td>
<td>_</td>
</tr>
<tr>
<td>Absence of HCM in first degree rels</td>
<td>+</td>
<td>_</td>
</tr>
<tr>
<td>Peak VO2 &gt; 50 ml/kg/min</td>
<td>+</td>
<td>_</td>
</tr>
</tbody>
</table>
Gene Positive/Phenotype Negative Individuals

No symptoms
No risk factors for sudden death

Bethesda Guidelines
Can participate in all sports
Should have annual follow up to check for HCM phenotype

ESC Guidelines
Recreational, non competitive sports only

Annual Evaluation
Isolated ECG Abnormalities in the Absence of LVH
Role of ECG in Differentiating Physiological LVH from HCM

HCM

Athlete

+ Pathological Q waves
+ Deep T wave inversions
- Isolated Sokolow-Lyon LVH
+ Marked ST segment depression
+ Left bundle branch block
Isolated ECG Abnormalities in the Absence of LVH

Detailed Echocardiographic Assessment
Cardiac MR
Holter Monitor
Cardiopulmonary Exercise Test
Screen first degree relatives for HCM
?Detrain if diagnostic uncertainty persists

Annual review
Isolated ECG Abnormalities in the Absence of LVH

ECHO/Cardiac MR/24 hour ECG, Cardiopulmonary exercise test

- diastolic dysfunction, SAM, LV outflow obstruction, apical LVH
- Non sustained ventricular tachycardia
- Flat blood pressure response
- Peak VO2 < 50 ml/kg/min or > 120% predicted for age

No

Yes → HCM

First degree relatives with HCM

No

Yes → Continue to compete (annual follow up)
Exercise Guidelines for Athletes with Isolated ECG Abnormalities in the absence of other phenotypic features of HCM or Familial HCM

**Bethesda Guidelines**

Can participate in all sports

**ESC Guidelines**

Can participate in all sports
Implantation of an ICD does not change guidelines. Reliability of ICD during sport is unpredictable.

Contact sport inhibited and most non contact sport (except class 1A) not recommended.
<table>
<thead>
<tr>
<th>Scenario</th>
<th>36th Bethesda</th>
<th>ESC 2005</th>
</tr>
</thead>
<tbody>
<tr>
<td>Definite diagnosis of HCM plus risk factors</td>
<td>Low dynamic and low static sport (class 1A)</td>
<td>No competitive sport</td>
</tr>
<tr>
<td>Definite diagnosis of HCM, but with low risk profile or with probable diagnosis</td>
<td>Low dynamic and low static sport (class 1A)</td>
<td>Low dynamic and low static sport (class 1A)</td>
</tr>
<tr>
<td>Athlete with positive genotype and negative phenotype</td>
<td>Can participate in all competitive sport</td>
<td>Only recreational, non-competitive sports</td>
</tr>
</tbody>
</table>
Disparity Between AHA and ESC

Bethesda guidelines broader; don’t take into account risk stratification.

Bethesda more liberal with gene positive phenotype negative individuals

No clear guidelines relating to definition of recreational sport. Recreational non competitive sport can be more strenuous than some competitive class 1a sports
Arrhythmogenic Right Ventricular Cardiomyopathy
Problems with Risk Stratification

Diagnosis is difficult

Incomplete Penetrance

Natural history not fully understood
Individuals with Unequivocal ARVC or High Probability of ARVC

ESC Guidelines

Participation in Class IA sport (low intensity and low dynamic) in most athletes

Bethesda Guidelines (American)

Participation in class 1A sport in all athletes