Left Ventricular Hypertrophy
OR
Hypertrophic Cardiomyopathy

Dr Michael Papadakis
Lecturer in Cardiology, St George’s university of London
Member of the Sport’s Cardiology Nucleus of the EACPR
michael.papadakis@sgul.ac.uk
@MichaelPapadak2
Steady trickle of deaths
SCD is the beginning NOT the end
Aetiology of SCD in 118 athletic individuals in the UK

de Noronha SV. et al. Heart 2009;95:1409-1414
Hypertrophic Cardiomyopathy
HCM

LVOT obstruction and SAM

Diastolic Dysfunction

Mitral Regurgitation

Myocardial Ischaemia

Autonomic Dysfunction
How do we detect HCM?

12-lead ECG

- Up to 95% of HCM patients have an abnormal ECGs

Echocardiogram
What creates the overlap?

- Different patterns of hypertrophy
- 30% symmetric/concentric hypertrophy
Cardiac Adaptation to Exercise - The Athlete’s Heart

- Age
- Gender
- Size
- Sporting Discipline
- Ethnicity

Structural

Functional

Electrical
Meta-analysis of athlete’s heart
59 studies, 1451 athletes 1975-1998

<table>
<thead>
<tr>
<th></th>
<th>Athletes</th>
<th>Controls</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>LVPWd (mm)</td>
<td>10.8</td>
<td>8.8</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>IVSd (mm)</td>
<td>11.2</td>
<td>8.8</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>LVM (g)</td>
<td>268</td>
<td>174</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>LVIDd (mm)</td>
<td>54.0</td>
<td>49.6</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

B.M. Pluim et al. Circulation 1999;100:336-344
Athlete’s heart vs. Cardiac pathology

ECG abnormalities in the athlete

(Group 1) common (up to 80%)
- Sinus bradycardia
- First degree AV block
- Notched QRS in V1 or incomplete RBBB
- Early repolarization
- Isolated QRS voltage criteria for left ventricular hypertrophy

(Group 2) Uncommon (< 5%)
- T-wave inversion
- ST-segment depression
- Pathological Q waves
- Left atrial enlargement
- Left axis deviation/left anterior hemiblock
- Right axis deviation/left posterior hemiblock
- Right ventricular hypertrophy
- Complete LBBB or RBBB
- Long or short QT interval
- Brugada-like early repolarization
- Ventricular arrhythmias
The challenge

Left ventricular hypertrophy 13-16 mm
Athlete’s repolarisation anomalies
How to differentiate athlete’s heart from HCM

- History
- Physical examination
- 12-lead ECG
- Echocardiography
- Cardiopulmonary exercise test
- Prolonged ECG monitoring
- Cardiac MRI
- Familial evaluation
- Genetic testing
- Detraining
The role of the 12-lead ECG

- Lateral T-wave inversions
- ST-segment depression
- Pathological Q-waves
- Left bundle branch block

Raise suspicion of HCM
Black athlete’s ECG

Lateral T-wave inversions suggest HCM (particularly in female athletes)

Papadakis M et al. Eur Heart J 2011;32:2304-2313
Refining the ECG criteria for screening athletes

Refined Criteria Training Related Normal Variants
- Sinus bradycardia
- First-degree AV block
- Incomplete RBBB
- Early repolarisation
- Isolated QRS voltage criteria for LVH

Refined Criteria Borderline Variants
- Left atrial enlargement
- Right atrial enlargement
- Left axis deviation
- Right axis deviation
- Right ventricular hypertrophy
- TWI up to V4 in BAs

Refined Criteria Training Unrelated Changes
- ST-segment depression
- Pathological Q-waves
- Ventricular pre-excitation
- TWI beyond V1 in WAs beyond V4 in BAs
- Complete LBBB or RBBB
- QTc ≥470 ms in males ≥480 ms in females
- Brugada-like ER
- Atrial or vent. arrhythmias
- ≥2 PVCs per 10 sec tracing

If present in ISOLATION*
If TWO OR MORE present
<table>
<thead>
<tr>
<th>Spec</th>
<th>BLACK ATHLETES (n=805)</th>
<th>WHITE ATHLETES (n=2282)</th>
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</thead>
<tbody>
<tr>
<td>ESC</td>
<td>40%</td>
<td>74%</td>
</tr>
<tr>
<td>Seattle Criteria</td>
<td>79%</td>
<td>92%</td>
</tr>
<tr>
<td>Refined Criteria</td>
<td>84%</td>
<td>94%</td>
</tr>
</tbody>
</table>

Percentage of Positive ECGs

- **ESC Recommendations**
  - **WA**: 16.2%
  - **BA**: 40.4%

- **Seattle Criteria**
  - **WA**: 8.5%
  - **BA**: 23.1%

- **Refined Criteria**
  - **WA**: 5.3%
  - **BA**: 11.5%
Role of ECG in differentiating athlete’s heart from HCM

- Sokolow-Lyon voltage criterion for LVH +
+ ST segment depression -
+ Pathological q waves -
+ Deep T wave inversion in any lead in Caucasians (females) -
+ Deep T wave inversion in lateral leads in blacks -
+ Deep T wave inversion in inferior leads ?
+ Left bundle branch block -
- Isolated atrial enlargement, axis deviation, RVH +
The role of echocardiography

- Determine the magnitude of LVH
- Assess the pattern of LVH
- Compare LVH relative to LV cavity size
- Identify left ventricular outflow obstruction
- Measure indices of diastolic function
- Measure indices of longitudinal systolic function
- Risk stratification
Determinants of left ventricular hypertrophy

- Male sex
- Large BSA
- Endurance Sports
- Adult athletes
- Black Ethnicity

LVH
Effect of black ethnicity on LVH in athletes

LVWT >16mm in males & >13mm in females suggests HCM

Papadakis M et al. Eur Heart J 2011;32:2304-2313
Pattern of LVH

- Athletes exhibit concentric LVH

Non-concentric pattern of LVH suggests HCM
Left ventricular wall thickness relative to cavity size

- Athlete’s heart
- h/R ratio (h = IVSd + LVPWd, R=LVIDd)

\[
\text{h/R} = \frac{13 + 13}{68} = 0.38
\]

h/R ≥ 0.5 suggests HCM
Echocardiography – SAM

- Septum
- Mitral valve
## Indices of diastolic function

<table>
<thead>
<tr>
<th></th>
<th>Physiological LVH</th>
<th>HCM</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>E wave</strong></td>
<td>Increased/Normal</td>
<td>Decreased</td>
</tr>
<tr>
<td><strong>A wave</strong></td>
<td>Normal</td>
<td>Increased</td>
</tr>
<tr>
<td><strong>E/A ratio</strong></td>
<td>&gt; 1</td>
<td>≤ 1</td>
</tr>
<tr>
<td><strong>Deceleration time</strong></td>
<td>Normal</td>
<td>Increased</td>
</tr>
<tr>
<td><strong>IVRT</strong></td>
<td>Normal</td>
<td>Increased</td>
</tr>
<tr>
<td><strong>E’</strong></td>
<td>&gt; 9</td>
<td>≤ 9</td>
</tr>
<tr>
<td><strong>E/E’</strong></td>
<td>&lt; 12</td>
<td>≥ 12</td>
</tr>
<tr>
<td><strong>Pulmonary vein</strong></td>
<td>S/D &gt; 1</td>
<td>S/D &lt; 1</td>
</tr>
</tbody>
</table>
## Indices of systolic function

<table>
<thead>
<tr>
<th></th>
<th>Physiological LVH</th>
<th>HCM</th>
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<tbody>
<tr>
<td>EF</td>
<td>Decreased/Normal</td>
<td>Increased/Normal</td>
</tr>
<tr>
<td>FS</td>
<td>Decreased/Normal</td>
<td>Increased/Normal</td>
</tr>
<tr>
<td>S’</td>
<td>&gt; 9 cm/sec</td>
<td>≤ 9 cm/sec</td>
</tr>
<tr>
<td>TDI L strain</td>
<td>Normal/Homogeneous</td>
<td>Low/Heterogeneous</td>
</tr>
<tr>
<td>STI</td>
<td>Normal</td>
<td>Attenuated</td>
</tr>
</tbody>
</table>

TDI derived strain in HCM

Attenuated systolic & diastolic velocities

Heterogeneous systolic velocities & paradoxical loops
## Markers to identify athletes with HCM

19 athletes with LVH vs. 37 athletes with mild LVH

<table>
<thead>
<tr>
<th>Echo parameter</th>
<th>Sensitivity (%)</th>
<th>Specificity (%)</th>
<th>PPV (%)</th>
<th>NPV (%)</th>
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</thead>
<tbody>
<tr>
<td>S' ≤ 9cm/s</td>
<td>43</td>
<td>84</td>
<td>84</td>
<td>43</td>
</tr>
<tr>
<td>E/A ratio ≤ 1</td>
<td>5</td>
<td>95</td>
<td>67</td>
<td>34</td>
</tr>
<tr>
<td>E' ≤ 9cm/s</td>
<td>35</td>
<td>100</td>
<td>100</td>
<td>44</td>
</tr>
<tr>
<td>E/ E' ≥ 12</td>
<td>14</td>
<td>100</td>
<td>100</td>
<td>37</td>
</tr>
</tbody>
</table>
The role of CPET

- Peak VO2 >50ml/kg/min or >120% predicted

Role of CPET

- Evidence of malignant arrhythmias
- BP response to exercise (systolic rise ≥ 25mmHg)

VT/NSVT
Abnormal BP response
pVO2 < 50 ml/kg/min OR
< 120% predicted

suggest HCM
The role of cardiac MRI

- Presence and pattern of fibrosis
  - Late gadolinium enhancement (LGE)
  - Found in up to 84% of patients with HCM
  - Marker of scar / fibrosis

- Pattern of LVH

- Accurate assessment of wall thickness

- Other features
  - Clefts, trabeculations, papillary muscle architecture

- Assessment of LVOT gradient

- Perfusion defects (stress MRI)
Apical HCM
Cardiac MRI – Risk and prognosis
Role of genetic testing

- Heterogeneous disease
- Sarcomeric gene mutations account for 40%-75%
  - 12 different genes
  - More than 450 mutations

<table>
<thead>
<tr>
<th>GENE</th>
<th>SARCOMERIC PROTEIN</th>
<th>FREQUENCY (%)</th>
</tr>
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<tbody>
<tr>
<td>MYH7</td>
<td>Beta myosin heavy chain</td>
<td>12-20</td>
</tr>
<tr>
<td>MYBPC3</td>
<td>Myosin binding protein C</td>
<td>20-28</td>
</tr>
<tr>
<td>TNNT2</td>
<td>Troponin T</td>
<td>2-5</td>
</tr>
<tr>
<td>TNNI3</td>
<td>Troponin I</td>
<td>1-3</td>
</tr>
<tr>
<td>MYL2</td>
<td>Regulatory myosin light chain 2</td>
<td>&lt;1</td>
</tr>
<tr>
<td>ACTC1</td>
<td>Apha cardiac actin</td>
<td>&lt;1</td>
</tr>
<tr>
<td>MYL3</td>
<td>Essential myosin light chain 3</td>
<td>&lt;1</td>
</tr>
<tr>
<td>TMP1</td>
<td>Alpha tropomyosin</td>
<td>&lt;1</td>
</tr>
<tr>
<td>TTN</td>
<td>Titin</td>
<td>&lt;1</td>
</tr>
<tr>
<td>TNNC</td>
<td>Troponin C</td>
<td>&lt;1</td>
</tr>
<tr>
<td>MYH6</td>
<td>Alpha myosin heavy chain</td>
<td>&lt;1</td>
</tr>
</tbody>
</table>
When to test in cardiomyopathies?

- Where it makes a difference – Clinical impact
- Always after detailed clinical & family assessment
- Always think of the potential consequences
- Multidisciplinary approach

Geneticists

Cardiologist with expertise in cardiomyopathy

Genetic councillors

www.sportscardiology.eu
Atypical phenotypic features

- Pompe’s disease
  - Acid α-1,4 glucosidase
- Familial Amyloid TTR
- Mitochondrial cytopathies
- LAMP2 Cardiomyopathy (Danon disease)
- Fabry’s disease
  - α-galactosidase A
- PRKAG cardiomyopathy
Cascade Screening

Known genetic mutation in an affected family member

Screen first degree relatives

Gene positive

Life-long follow up

Gene negative

Reassure & discharge
Role of detraining

Detrain for 4-6 weeks

- Regression of LVH (Physiological)
- No regression of LVH (Pathological)
17-year-old asymptomatic swimmer with abnormal ECG and LVH of 14 mm

ECG Before and After Detraining for 8 Weeks
Conclusions

- Athletic training is associated with electrical and structural changes that may mimic morphologically mild HCM.
- The differentiation between HCM and ‘Athlete’s Heart’ is possible using a methodical approach.
  - Can be challenging particularly in black athletes.
Indicators of pathology

- Lateral deep T-wave inversions
- ST-segment depression
- Non-concentric LVH
- LVWT > 16mm (males), >13mm (females)
- Impaired diastolic function
- Peak VO₂ <50 ml/kg/min
- Myocardial fibrosis
- No regressions with detraining
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