HYPERTROPHIC CARDIOMYOPATHIES

WHICH MORPHOLOGICAL TEST TO PRESCRIBE AND WHY?

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DISCLOSURE STATEMENT OF FINANCIAL INTEREST

I currently have, or have had over the last two years, an affiliation or financial interests or interests of any order with a company or I receive compensation or fees or research grants with a commercial company:

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- Boehringer Ingelheim
- Edimark
- General Electric
- Novartis
- Shire
WHY PRESCRIBE A MORPHOLOGICAL TEST?

For **diagnostic** and **prognostic** assessment
LEFT VENTRICULAR HYPERTROPHY

Diagnosis

Prognosis

Elliott PM, Eur Heart J. 2014 Oct 14;35(39):2733-79
5.4 Echocardiography

Echocardiography is central to the diagnosis and monitoring of HCM.
ECHOCARDIOGRAPHY
Patterns and Significance of Distribution of Left Ventricular Hypertrophy in Hypertrophic Cardiomyopathy

A Wide Angle, Two Dimensional Echocardiographic Study of 125 Patients

Magnetic Resonance in Transthyretin Cardiac Amyloidosis

Asymmetric hypertrophy. Sigmoid septal contour (55%)

Asymmetric hypertrophy. Reverse septal contour (24%)

Martinez-Naharro A, J Am Coll Cardiol 2017;70:466–77
Mitral Valve Abnormalities Identified by Cardiovascular Magnetic Resonance Represent a Primary Phenotypic Expression of Hypertrophic Cardiomyopathy

Maron MS. Circulation. 2011;124:40-47
PROGNOSIS

HCM Risk-SCD Calculator

- Age: Years
- Maximum LV wall thickness: mm
- Left atrial size: mm
- Max LVOT gradient: mmHg

- Family History of SCD: No/Yes
- Non-sustained VT: No/Yes
- Unexplained syncope: No/Yes

Elliott PM, Eur Heart J. 2014 Oct 14;35(39):2733-79
PROGNOSIS

2D and Doppler echocardiography at rest, Valsalva and standing

- Maximum provoked peak LVOTO ≥50 mm Hg
  - see 9.1 Treatment of left ventricular outflow tract obstruction

- Maximum provoked peak LVOTO <50 mm Hg
  - Asymptomatic
    - Repeat echocardiography 1 year
  - Symptomatic
    - Exercise stress echocardiography
    - Maximum provoked peak LVOTO ≥50 mm Hg
    - Maximum provoked peak LVOTO <50 mm Hg

Elliott PM, Eur Heart J. 2014 Oct 14;35(39):2733-79
ECHOCARDIOGRAPHY

3
Relative apical sparing of longitudinal strain using two-dimensional speckle-tracking echocardiography is both sensitive and specific for the diagnosis of cardiac amyloidosis.

Phelan D. Heart 2012;98:1442e1448
Relative apical sparing of longitudinal strain using two-dimensional speckle-tracking echocardiography is both sensitive and specific for the diagnosis of cardiac amyloidosis.
ECHOCARDIOGRAPHY
Right ventricular longitudinal strain: a tool for diagnosis and prognosis in light-chain amyloidosis

RV wall thickness >5 mm

area under the curve = 0.744

Uzan C. Amyloid. 2017 Dec 20:1-8
PROGNOSIS

Right ventricular longitudinal strain: a tool for diagnosis and prognosis in light-chain amyloidosis

Uzan C. Amyloid. 2017 Dec 20:1-8
Diagnostic score for the detection of cardiac amyloidosis in patients with left ventricular hypertrophy and impact on prognosis

<table>
<thead>
<tr>
<th>Continuous variables associated with the diagnosis of cardiac amyloidosis and value selected for the diagnostic score.</th>
<th>AUC</th>
<th>IC95%</th>
<th>p-value</th>
<th>Best cutoff value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systolic arterial pressure</td>
<td>0.789</td>
<td>0.619–0.919</td>
<td>.001</td>
<td>&lt;130 mmHg</td>
</tr>
<tr>
<td>Troponin</td>
<td>0.667</td>
<td>0.515–0.819</td>
<td>.061</td>
<td></td>
</tr>
<tr>
<td>NT pro-BNP</td>
<td>0.667</td>
<td>0.515–0.817</td>
<td>.063</td>
<td></td>
</tr>
<tr>
<td>PR duration</td>
<td>0.740</td>
<td>0.600–0.881</td>
<td>.007</td>
<td>&gt;200 ms</td>
</tr>
<tr>
<td>Sokolow index</td>
<td>0.793</td>
<td>0.669–0.917</td>
<td>.001</td>
<td>&lt;12 mV</td>
</tr>
<tr>
<td>DI + DII + DIII</td>
<td>0.758</td>
<td>0.594–0.921</td>
<td>.004</td>
<td>&gt;13 mm</td>
</tr>
<tr>
<td>Diastolic LV posterior thickness</td>
<td>0.758</td>
<td>0.624–0.891</td>
<td>.004</td>
<td></td>
</tr>
<tr>
<td>Transmitral flow peak E velocity</td>
<td>0.751</td>
<td>0.612–0.890</td>
<td>.005</td>
<td></td>
</tr>
<tr>
<td>E wave deceleration time</td>
<td>0.756</td>
<td>0.608–0.904</td>
<td>.004</td>
<td></td>
</tr>
<tr>
<td>Transmitral flow E/A ratio</td>
<td>0.741</td>
<td>0.587–0.895</td>
<td>.007</td>
<td></td>
</tr>
<tr>
<td>E/Ea ratio</td>
<td>0.808</td>
<td>0.688–0.928</td>
<td>.001</td>
<td>&gt;10</td>
</tr>
<tr>
<td>Global longitudinal strain</td>
<td>0.775</td>
<td>0.644–0.907</td>
<td>.002</td>
<td>&gt;−12%</td>
</tr>
<tr>
<td>Sum of basal longitudinal strain</td>
<td>0.776</td>
<td>0.647–0.906</td>
<td>.002</td>
<td>&gt;−47%</td>
</tr>
</tbody>
</table>

AUC: area under the curve; LV: left ventricle; NT-ProBNP: N-terminal prohormone of brain natriuretic peptide.

Diagnostic score for the detection of cardiac amyloidosis in patients with left ventricular hypertrophy and impact on prognosis

Score >3:
- sensibilité 90%
- spécificité 81%

AUC = 0.933 (95%CI 0.889 - 0.978)

P value by log rank test = 0.002
Suspicion of cardiac amyloidosis

BONE SCINTIGRAPHY
Bone scintigraphy (particularly with 99mTc-DPD) should be considered in patients with symptoms, signs and non-invasive tests consistent with TTR-related amyloidosis.
13% de fixation cardiaque modérée à sévère à la scintigraphie au $^{99m}$Tc-DPD dans une population de patients ICFEp de plus de 60 ans avec HVG (SIV > 12 mm)
PROGNOSIS

Role of $^{99m}$Tc-DPD Scintigraphy in Diagnosis and Prognosis of Hereditary Transthyretin-Related Cardiac Amyloidosis

O’Hanlon R, J Am Coll Cardiol 2010;56:867–74
Suspicion of another diagnosis than cardiac amyloidosis

CARDIAC MAGNETIC RESONANCE
In the absence of contraindications, 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.
HCM  Fabry disease
Clinical recommendations for cardiovascular magnetic resonance mapping of T1, T2, T2* and extracellular volume: A consensus statement by the Society for Cardiovascular Magnetic Resonance (SCMR) endorsed by the European Association for Cardiovascular Imaging (EACVI)
PROGNOSIS

Prognostic Significance of Myocardial Fibrosis in Hypertrophic Cardiomyopathy

O’Hanlon R, J Am Coll Cardiol 2010;56:867–74
CONCLUSION

Imaging for diagnostic and prognostic assessment

**Echocardiography** plays an essential role in assessing left ventricular hypertrophy.

**Cardiac magnetic resonance** allows to specify the diagnosis when the echocardiography is nonspecific.

**Bone scintigraphy** diagnoses transthyretin related amyloidosis in case of suspicion of cardiac amyloidosis.
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