Declaration of conflict of interest
Longitudinal Base-to-Apex Strain Gradient is a Good Diagnostic Marker for Differentiating Patients with Cardiac Amyloidosis from Other Hypertrophic Cardiomyopathies

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Cardiac amyloidosis (CA) need to be distinguished from other causes of left ventricular hypertrophy in clinical practice.

As described by our previous study, longitudinal base-to-apex strain gradient in the septum detected by speckle tracking imaging is frequently documented in patients with CA.

Whether this typical myocardial deformation gradient could be clinically useful as a specific diagnostic marker?
Study Aims

The aims of this study were...

- to compare the longitudinal strain characteristics between cardiac amyloidosis and other concentric LV hypertrophy diseases.
  - Fabry disease (FD)
  - Friedreich ataxia (FA)
  - isolated arterial hypertension (HP)

- to evaluate the diagnostic value of the typical longitudinal strain pattern in concentric LV hypertrophic cardiomyopathies.
Methods

study population

- cardiac amyloidosis (CA, n = 25)
- Fabry disease (FD, n = 25)
- Friedreich ataxia (FA, n = 25)
- isolated arterial hypertension (HP, n = 25)
- normal control (n = 25)

standard echocardiography

speckle tracking imaging

- longitudinal peak systolic strain (LS) in the septum
Methods

inclusive criteria for cardiac amyloidosis

- biopsy-proven systemic amyloidosis
- typical findings of echocardiography and electrocardiography

Echocardiology
1) LV wall thickness $\geq 12$ mm
2) “granular sparkling” texture of myocardium
3) enlarged left and right atria
4) pericardial effusion
5) diastolic pseudonormal or restrictive filling pattern

Electrocardiology
1) unexplained low voltage in peripheral leads
2) QRS and T-wave pseudo-infarct changes
3) conduction abnormalities
Methods

inclusive criteria for cardiac amyloidosis
  • biopsy-proven systemic amyloidosis
  • typical findings of echocardiography and electrocardiography

exclusive criteria
  • coronary artery disease, moderate to severe cardiac valve stenosis, and other endocrine or systemic disease
  • septum or left ventricular posterior wall thickness < 12 mm
# Results

Clinical and standard echocardiographic characteristics

<table>
<thead>
<tr>
<th></th>
<th>Control</th>
<th>Cardiac amyloidosis</th>
<th>Friedreich’s ataxia</th>
<th>Fabry disease</th>
<th>Arterial hypertension</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>63 ± 7</td>
<td>66 ± 10</td>
<td>25 ± 10*†</td>
<td>62 ± 9‡</td>
<td>67 ± 13‡</td>
</tr>
<tr>
<td>Male (%)</td>
<td>60</td>
<td>56</td>
<td>72</td>
<td>40</td>
<td>48</td>
</tr>
<tr>
<td>Mean LV thickness (mm)</td>
<td>9 ± 1</td>
<td>14 ± 1*</td>
<td>13 ± 1*</td>
<td>14 ± 2*</td>
<td>14 ± 2*</td>
</tr>
<tr>
<td>LVEDD (mm)</td>
<td>50 ± 4</td>
<td>42 ± 6*</td>
<td>42 ± 7*</td>
<td>46 ± 7†‡</td>
<td>47 ± 7†‡</td>
</tr>
<tr>
<td>EF (%)</td>
<td>66 ± 6</td>
<td>58 ± 12*</td>
<td>65 ± 7</td>
<td>64 ± 8</td>
<td>60 ± 12</td>
</tr>
<tr>
<td>E/A</td>
<td>0.99 ± 0.22</td>
<td>1.46 ± 0.71</td>
<td>1.54 ± 0.45*</td>
<td>1.15 ± 0.52</td>
<td>0.80 ± 0.23*†‡ §</td>
</tr>
<tr>
<td>DT (ms)</td>
<td>221 ± 52</td>
<td>147 ± 46*</td>
<td>183 ± 52</td>
<td>255 ± 85†‡</td>
<td>251 ± 60†‡</td>
</tr>
<tr>
<td>E/E´</td>
<td>10 ± 4</td>
<td>23 ± 10*</td>
<td>8 ± 2†</td>
<td>19 ± 7*‡</td>
<td>14 ± 5*†‡</td>
</tr>
</tbody>
</table>

* P < 0.05 vs. Control; † P < 0.05 vs. CA; ‡ P < 0.05 vs. FA; § P < 0.05 vs FD.
Cardiac amyloidosis

Friedreich’s ataxia

Arterial hypertension

Fabry Disease
Cardiac amyloidosis

IVSd: 13 mm
EF: 53%

Friedreich’s ataxia

IVSd: 13 mm
EF: 54%

Arterial hypertension

IVSd: 13 mm
EF: 50%

Fabry Disease

IVSd: 15 mm
EF: 63%
**Longitudinal Base-to-Apex Strain Gradient**

### Table: Longitudinal Strain Curves

<table>
<thead>
<tr>
<th></th>
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<th>Friedreich’s ataxia</th>
<th>Fabry disease</th>
<th>Arterial hypertension</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>N</strong></td>
<td>25</td>
<td>25</td>
<td>25</td>
<td>25</td>
<td>25</td>
</tr>
<tr>
<td><strong>LS_septum (%)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>basal</td>
<td>-17 ± 3</td>
<td>-6 ± 2*</td>
<td>-16 ± 2†</td>
<td>-13 ± 6*†</td>
<td>-14 ± 6†</td>
</tr>
<tr>
<td>mid</td>
<td>-18 ± 4</td>
<td>-10 ± 4*</td>
<td>-15 ± 3*†</td>
<td>-15 ± 5†</td>
<td>-15 ± 6†</td>
</tr>
<tr>
<td>apical</td>
<td>-20 ± 5</td>
<td>-20 ± 7</td>
<td>-21 ± 4</td>
<td>-17 ± 7</td>
<td>-21 ± 8</td>
</tr>
<tr>
<td><strong>LS_api/bas</strong></td>
<td>1.2 ± 0.3</td>
<td>3.3 ± 1.6*</td>
<td>1.3 ± 0.3†</td>
<td>1.7 ± 1.8†</td>
<td>1.8 ± 0.9†</td>
</tr>
</tbody>
</table>
LS_api/bas > 2.1 as a cut-off value for differentiating CA from other hypertrophic cardiomyopathies (FA, FD and HP)

Sensitivity: 88%
Specificity: 85%
LS_api/bas > 2.1 + DT < 200 ms

Sensitivity: 88%
Specificity: 100%
Conclusions

The longitudinal base-to-apex strain gradient within the septum (LS_api/bas > 2.1) in combination with the short deceleration time of early filling (DT < 200ms) could be used as a diagnostic marker for differentiating CA from other hypertrophic cardiomyopathies.
Thanks for your attention!