Valvular, Myocardial, Pericardial, Pulmonary, Congenital Heart Disease – Myocardial Disease, Clinical, Infiltrative Myocardial Disease

Myocardial Stiffness evaluation by shear wave elastography in transthyretin amyloidosis with and without cardiac involvement

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Introduction: Cardiac involvement (CI) in hereditary transthyretin amyloidosis (ATTRh) occurs by deposition of amyloid fibrils in the heart, resulting in stiffening and diastolic dysfunction. Shear wave elastography, non-invasively and without the emission of ionizing radiation, quantitatively assesses tissue stiffness, a passive component of diastole, complementing the complex echocardiographic assessment.

Objective: To evaluate myocardial stiffness, through diastolic myocardial elasticity (DME), in ATTRh patients with and without CI and a healthy control group (CG).

Methodology: Prospective, cross-sectional study, 60 patients divided into 3 groups: ATTRh with CI (n=20), ATTRh without CI (n=20) and GC (n=20). DME was evaluated in the septal wall of the left ventricle (LV) (basal, middle, and apical segments) and in the free wall of the right ventricle (RV). They were also submitted to 2D-ECO, EKG, troponin, BNP, 6-minutes-walking test and pyrophosphate-labeled scintigraphy (Tc-99m-PYP). Data were analyzed using a one-way ANOVA, verifying myocardial stiffness, measured by DME, among the 3 groups. Then, pairwise comparisons were adjusted using Tukey’s technique.

Results: DME was a significant factor for distinguishing between groups when analyzed in the parasternal long axis (PLAX) basal septum (p=0.013), parasternal short axis (PSAX) basal septum (p=0.03) and in the PLAX RV free wall (p=0.004). Was observed in the apical septum segment similar stiffness between groups, suggesting a pattern of apical preservation. In the post hoc evaluation, DME in the basal septum region was significantly higher among the ATTRh with CI group than in the control group, but without significant difference between ATTRh with CI and ATTRh without CI, suggesting that the latter group has intermediate values between the sick patient and the healthy patient. Also, right ventricle DME in ATTRh with CI was greater than in the other two groups (Table 1 and Figure 1). In a regression model, the results indicate that 17.2% of the DME variance can be attributed to 3-hour scan uptake on the Tc-99m-PYP (R²=0.172, F (1-36) =7.2, p< 0.011), estimated by the equation: 3-hour scan uptake = 0.91 +0.078 x DME.

Conclusion: Myocardial stiffness is increased in ATTRh with CI, showing a decreasing pattern of stiffness in the apex direction, suggesting a pattern of apical preservation. Cardiac elastography has the potential to be an important tool in the assessment of cardiac involvement in systemic amyloidosis.
### Table 1 Cardiac elastography

<table>
<thead>
<tr>
<th></th>
<th>ATTRh with CI (n=20)</th>
<th>ATTRh without CI (n=19)</th>
<th>Control (n=20)</th>
<th>P-value</th>
<th>η²</th>
</tr>
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<tbody>
<tr>
<td><strong>Elastography</strong></td>
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<tr>
<td>Basal Septum (PLAX) (kPa)</td>
<td>6.6 ± 1.4</td>
<td>5.6 ± 1.4</td>
<td>5.4 ± 1.1</td>
<td>P=0.013*</td>
<td>0.14</td>
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<tr>
<td>Basal Septum (PSAX) (kPa)</td>
<td>6.7 ± 1.4</td>
<td>5.8 ± 1.4</td>
<td>5.6 ± 1.2</td>
<td>P=0.03*</td>
<td>0.11</td>
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<tr>
<td>Middle Septum (PSAX) (kPa)</td>
<td>8.1 ± 1.7</td>
<td>5.3 ± 1.4</td>
<td>5 ± 0.8</td>
<td>P=0.065</td>
<td>-</td>
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<tr>
<td>Apical Septum (PSAX) (kPa)</td>
<td>4.5 ± 0.9</td>
<td>4.3 ± 0.8</td>
<td>4.5 ± 0.8</td>
<td>P=0.6</td>
<td>-</td>
</tr>
<tr>
<td>RV (PLAX) (kPa)</td>
<td>5.4 (3.8 – 7)</td>
<td>3.7 (3.1-4.1)</td>
<td>4 (3.5 – 4.6)</td>
<td>P=0.004**</td>
<td>0.23</td>
</tr>
</tbody>
</table>

kPa = Kilopascal; RV = right ventricle
PLAX = Parasternal short-axis; PSAX = Parasternal long-axis

*In patient EAS1-20006, of the ATTR group without CI, there was not possibility acquire septum hardness (PLAX) due to acoustic window limitation.

*In patient EAS1-20007, of the ATTR group without CI, there was not possibility acquire chamber thickness at all 3 points due to technical difficulty due to acoustic window limitation.

**In this comparison with three independent groups, there was statistically significant difference only between the ATTR and control group (ATTRACCONTROL) and ATTRAC and ATTR without CI (ATTRACATTR without CI).

**In this comparison with three independent groups, there was statistically significant difference between the ATTRAC and control group (ATTRACCONTROL) and ATTRAC and ATTR without CI (ATTRACATTR without CI).

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**Figure 1** DME variation (mean) in basal-middle-apical segments in the ATTRh without CI, ATTRh with CI and control group.