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Evaluation of Perugini score and myocardial mass and voltage abnormalities in transthyretine cardiac amyloidosis

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Background: Transthyretine cardiac amyloidosis (TTR-CA) is an emerging treatable disease in cardiology. Severity of TTR infiltration can be assessed by bone scintigraphy with Perugini score, depending on HPD-Tc99M myocardial intensity uptake. Less is known on how Perugini score interplays with routine cardiac structural parameters, especially cardiac mass and voltage.

Aim: To evaluate correlation between Perugini score and myocardial mass and voltage abnormalities in patients with TTR-CA.

Methods: Patients diagnosed with ATTR-CA and addressed to our center for treatment underwent complete evaluation including electrocardiogram (ECG) and transthoracic echocardiography (TTE). Routine electrical and echographic parameters were obtained such as: Sokolow indice (SV1+RV5) in millivolt (mV), PR delay (ms), QRS width (in ms), left ventricular mass (LVM, in grams) and diastolic interventricular septum (dIVS) thickness (in mm). Patients were classified in three categories according to Perugini score: I (cardiac uptake < chest bones), II (cardiac uptake = chest bones) and III (cardiac uptake > bones). Main end-point were LVM and Voltage/Mass ratio (VMR) variations, depending on Perugini score.

Results: Among the 17 patients included, 14 were male (82.4%) and mean age was 82±8 years old. On ECG, 6 patients (35.3%) had permanent atrial fibrillation (AF). Mean voltage according to Sokolow index, mean PR delay and mean QRS width were respectively 1.5±0.5 mV, 212±54 ms and 113±19 ms. On TTE, mean LVM and mean dIVS width were respectively 333±98 g (188±55 g/m^2) and 19±4 mm. Mean voltage/mass ratio (VMR) was 0.48±0.26 uV/g (see table). On bone scintigraphy, 3 patients were scored with Perugini I (17.6%), 4 with Perugini II (23.5%) and 10 with Perugini III (58.8%). According to Pearson test, there was a strong correlation between Perrugini score and LVM (r=+0.56, p=0.03) (graph A), and a strong negative correlation between Perrugini score and VMR (r=−0.70, p=0.003) (graph B). There were a trend for correlation between Perugini score and Voltage according to Sokolow (r=−0.46, p=0.07) and between Perugini score and dIVS width (r=0.49, p=0.07).

Conclusion: In TTR-CA patients, Perugini score appears to strongly correlate with left ventricle mass and voltage. Larger scale studies are needed to confirm these results.

	All patients	Perugini I	Perugini II	Perugini III
Patients, n (%)	17 (100)	3 (17.6)	4 (23.5)	10 (58.8)
Age, mean (SD)	82 ±8	80 ±12	86 ±1	81 ±8
Electrocardiographic parameters :				
– Atrial fibrillation	6 (35.3)	0 (0)	2 (50)	4 (40)
– PR, *in ms (SD)*	212 ±54	173 ±29	295 ±33	202 ±40
– QRS width, *in ms (SD)*	112 ±18	98 ±7	122 ±21	113 ±19
– Sokolow (SV1+RV5) voltage, *in mV (SD)*	1.5 ± 0.5	1.7 ±0.3	1.95 ±0.3	1.2 ±0.6
Echocardiographic parameters :				
– Left ventricle mass (g)	333 ±98	213 ±39	309 ±62	333 ±98
– Diastolic interventricular septum thickness, *in mm*	19 ±4	14 ±2	19 ± 4	20 ±4
Voltage/Mass ratio, in microV/g	0.48 ±0.26	0.78 ± 0.28	0.66 ± 0.17	0.34 ±0.18

ECG and TEE trends according to Perugini
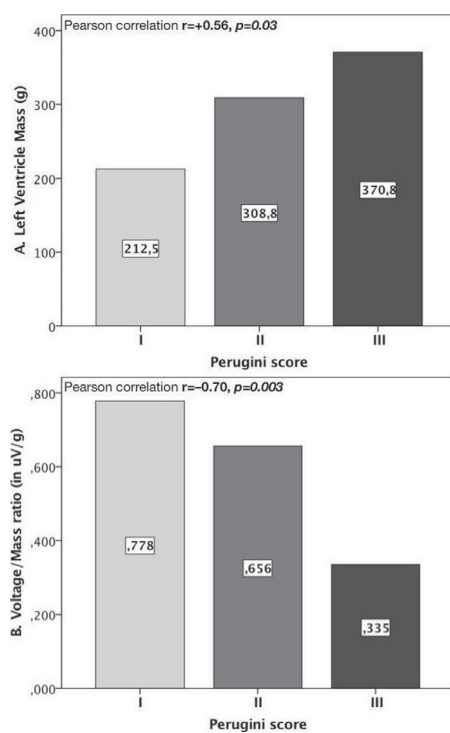
Cardiac mass and voltage with Perugini