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Cardiac 99mTc-DPD uptake in transthyretin V30M amyloidosis depends on the age of disease onset

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Background : Transthyretin (TTR) V30M familial amyloid polyneuropathy (FAP) is a progressive and fatal disease for which several promising therapies are in development. There is a growing need of a simple non-invasive test for the early diagnosis of patients with TTR cardiac amyloidosis, who may benefit from emerging treatments.

Purpose: To evaluate the utility and limitations of 99mTc-3,3-diphosphono-1,2-propanodicarboxylic acid (DPD) scintigraphy for the diagnosis of cardiac involvement in patients with TTR-V30M amyloidosis.

Methods and Results: A cohort of 179 TTR-V30M mutation carriers underwent DPD scintigraphy, which was compared with the results of echocardiogram, ambulatory blood pressure monitoring, 24h-Holter, myocardial 123-iodine metaiodobenzylguanidine imaging and NT-proBNP. Amyloid cardiomyopathy, defined as septal thickness =13mm, was present in 32 patients (17.9%) and was more prevalent in patients with late-onset disease (OR: 3.68, P=0.003). Abnormal cardiac DPD uptake was present in 22 individuals (12.3%). Overall, DPD uptake ratio increased with septal thickness (Pearson R: 0.51; P <0.001) and correlated with other parameters indicative of cardiac amyloidosis. However, DPD imaging was strongly influenced by the age of the disease onset. Among patients with myocardial thickening, cardiac DPD retention was present in 11 of 15 (73.3%) with late-onset disease, in contrast to only 4 of 17 (26.7%) with early-onset disease (P=0.005). Two of the 13 early-onset disease patients with myocardial thickening and no DPD uptake underwent endomyocardial biopsy that confirmed the presence of TTR amyloidosis.

Conclusions: Cardiac DPD imaging is less accurate in TTR-V30M FAP than it is in other forms of TTR amyloidosis. Particularly, it cannot be used to rule out cardiac involvement in symptomatic patients with early-onset disease.