Etiology of Amyloidosis Determines Myocardial ^{99m}Tc-DPD Uptake in Amyloidotic Cardiomyopathy

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Abstract: ^{99m}Tc-DPD (^{99m}Tc-3,3-diphosphono-1,2-propanodicarboxylic acid) has a high affinity for transthyretin (TTR)-infiltrated myocardium, allowing a differential diagnosis with light chain cardiac amyloidosis and other nonamyloidotic cardiomyopathies with a hypertrophic phenotype, in which myocardial tracer uptake is low or absent. Myocardial bone tracer uptake in the rarer forms of amyloidosis (eg, apolipoprotein-related) has been rarely studied. We present 4 cases of cardiac amyloidosis that underwent ^{99m}Tc-DPD scintigraphy; myocardial DPD uptake was present in patients with ATTR, wtTTR and apolipoprotein AI and negative in cases with AL and apolipoprotein AII-related disease.

Key Words: amyloidosis, scintigraphy, DPD, transthyretin, apolipoprotein

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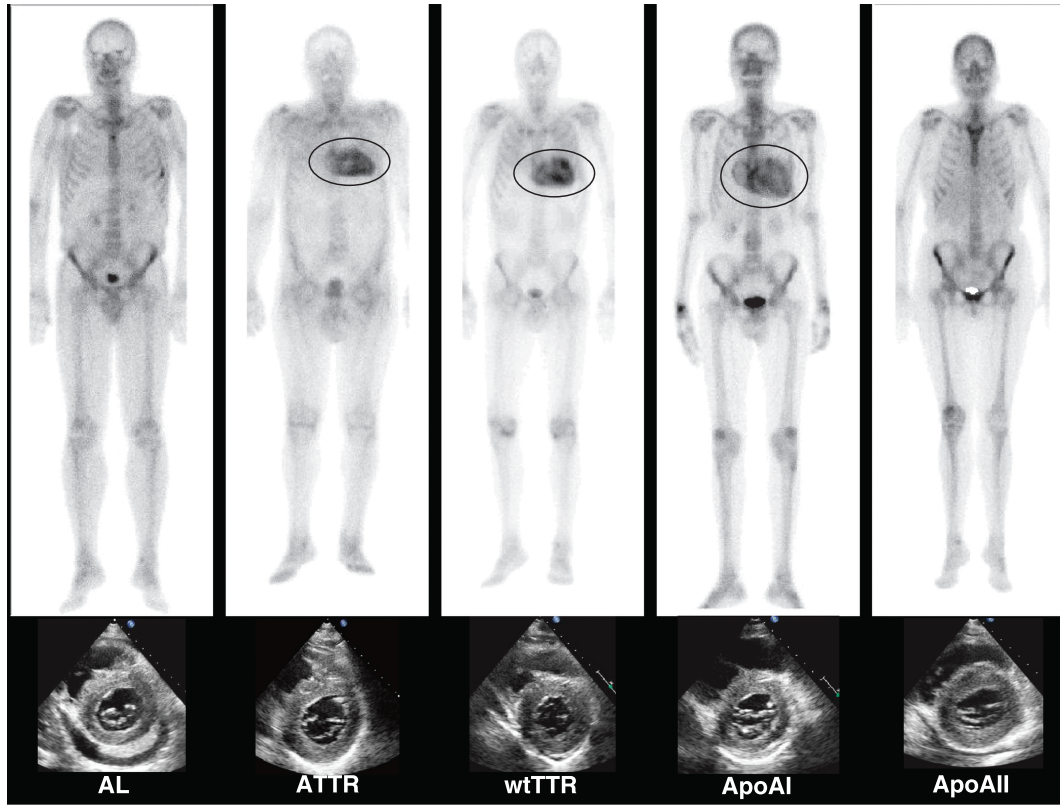

FIGURE 1. The role of ^{99m}Tc-DPD (^{99m}Tc-3,3-diphosphono-1,2-propanodicarboxylic acid) scintigraphy in the diagnosis of transthyretin (TTR)-related amyloidotic cardiomyopathy has been well documented in recent years.[1,2] The affinity of DPD for TTR-infiltrated myocardium has proven useful in differentiating TTR-related amyloidosis from the primary light chain (AL) form and from other nonamyloidotic cardiomyopathies with a hypertrophic phenotype.[3,4] Knowledge regarding myocardial bone tracer uptake in the rarer forms of amyloidosis (eg, apolipoprotein related[5–7]) is limited.[8] In 2 cases of biopsy-proven cardiac amyloidosis that recently underwent ^{99m}Tc-DPD scintigraphy at our center, myocardial uptake was present in apolipoprotein AI (ApoAI)[8] and negative in apolipoprotein AII (ApoAII). These findings therefore configure an extreme heterogeneity of myocardial uptake in amyloidosis that seems to depend on etiology rather than on ventricular wall thickness (expression of the degree of infiltration). Our image shows late DPD sequences (180 minutes after DPD administration, previously described methods[1]) in 5 different cases of biopsy-proven amyloidotic cardiomyopathy; myocardial tracer uptake is intense in the mutated TTR-related form (ATTR), in wild-type TTR-related (wtTTR), and in ApoAI; on the contrary, no uptake can be appreciated in AL or ApoAII despite similar wall thickness (maximum, 18 mm). Our images therefore strengthen the concept that DPD uptake is mainly determined by the specific etiology of amyloidosis and not by the severity of myocardial amyloidotic infiltration.