Technetium-99m-3,3-diphosphono-1,2-2 propanodicarboxylic acid (DPD) in AL cardiac amyloidosis

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In this issue of the *Journal of Nuclear Cardiology*, Persia Paulino et al. present a case of a 40 year old man with a positive Technetium-99m-3,3-diphosphono-1,2-2 propanodicarboxylic acid (DPD) scan who was found to have AL cardiac amyloidosis by cardiac biopsy.¹ The patient had no prior medical history and was not taking any medications when he presented to the emergency room with symptomatic atrial flutter. Laboratory tests revealed elevated serum troponin, NT-pro B-type brain natriuretic peptide, and creatinine. Echocardiography showed mild global left ventricular hypokinesis with longitudinal strain pattern demonstrating preserved apical function with progressive dysfunction towards the base and Doppler imaging consistent with restrictive physiology. Cardiac magnetic resonance imaging showed late gadolinium enhancement with diffuse subendocardial pattern and difficulty in complete suppression. Since these findings were highly suggestive of cardiac amyloidosis, a DPD scan was done which demonstrated significant cardiac uptake (Perugini visual score Grade 3, Heart-to-Contralateral ratio 1.67). Serum

analysis demonstrated a monoclonal gammopathy with a high Lambda light chain. Endomyocardial biopsy was performed with histology showing amyloid deposition in the heart with positive Kappa and Lambda light chains on immunohistochemistry.

A recent expert consensus statement led by the American Society of Nuclear Cardiology (ASNC) in collaboration with other societies including the American Heart Association and American Society of Echocardiography performed a comprehensive review of existing evidence on the utility of imaging in the management of patients with cardiac amyloidosis.^{2,3} These consensus recommendations establish a clinical pathway for the diagnosis of transthyretin cardiac amyloidosis (ATTR) using bone scan imaging such as DPD or Technetium-99m-pyrophosphate (PYP) that does not require the use of histology to confirm the diagnosis. This statement is a testament to the exceptionally high sensitivity and specificity of PYP/DPD scans for ATTR cardiac amyloid. However, it is important to note that the statement requires the absence of a clonal plasma cell process by free light chains and serum and urine immunofixation as an essential component of this clinical pathway (in addition to typical cardiac imaging features). As evidenced in this case, AL cardiac amyloid can demonstrate radionuclide cardiac uptake in the absence of cardiac ATTR. It is therefore extremely important to actively rule out AL amyloid any time that a radionuclide bone scan is contemplated for evaluation of cardiac amyloid since missing the diagnosis of AL amyloid can be devastating to the patient. Finally, using single photon emission computed tomography (SPECT) can be very useful for the interpretation of these scans since it may allow the differentiation of myocardial uptake from blood pool activity.

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Disclosures

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