A DIAGNOSTIC APPROACH
FOR PATIENTS WITH SUSPECTED CARDIAC AMYLOIDOSIS
THAT INCLUDES TESTING FOR MONOCLONAL PROTEIN
FOLLOWED BY SCINTIGRAPHY AND/OR BIOPSY

Symptoms, ECG, Echo, MRI, or Biomarkers suggestive of cardiac amyloidosis

Screen for the presence of a monoclonal protein
Order the following three tests:
- Serum kappa/lambda free light chain ratio (abnormal if ratio is <0.26 or >1.65)
- Serum protein immunofixation (abnormal if monoclonal protein is detected)
- Urine protein immunofixation (abnormal if monoclonal protein is detected)

1 or more abnormal

Bone Scintigraphy Available?

- yes
  - Positive Bone Scintigraphy
  - Referral for Bone Scintigraphy or invasive evaluation with Heart Biopsy
    - Positive Congo Red
    - Tissue typing by mass spectrometry or immunostaining

- no
  - Negative or indeterminate

Referral to Hematology

Biopsy of clinically involved organ (cardiac or renal) or fat pad*
- Positive Congo Red
- Tissue typing by mass spectrometry or immunostaining

- AL, ATTR, other amyloidosis and/or MGUS
  - Cardiac Amyloidosis Unlikely

Genetic Testing

- ATTR Amyloidosis
  - ATTRwt
  - ATTRm

- Cardiac Amyloidosis Unlikely

Consider Heart Biopsy if suspicion is high

*If fat pad is negative, biopsy of involved organ is required.

**99m**-Technetium-pyrophosphate (**99m**Tc-PYP) is a noninvasive radioactive tracer utilized as an adjunct in the diagnosis of ATTR-CM, though not FDA approved for that use.

Both planar and SPECT imaging should be reviewed and interpreted using visual and quantitative approaches.


ATTRm, mutant transthyretin amyloidosis; MGUS, monoclonal gammopathy of undetermined significance; MRI, magnetic resonance imaging; SPECT, single-photon emission computed tomography.