



## Diagnosis of Light Chain Amyloidosis Audit Report

Use this set of metrics to assess your compliance with the American Society of Hematology (ASH) Clinical Practice Guidelines on Diagnosis of Light Chain Amyloidosis. Results may be used to identify quality gaps and target quality improvement efforts at your institution.

### 1. Time to Diagnosis

Quality Priority	Minimize time between symptom onset and diagnosis of amyloidosis
Population	Patients diagnosed with amyloidosis
Metric	Time between first symptom onset to diagnosis
Audit Metric 1	Median time (in days) between initial symptom onset and confirmed diagnosis
Implementation Comments	Symptoms can be vague at initial presentation and missed across disciplines (e.g., cardiology, nephrology, neurology, general internal medicine and hematology)
Suggested Areas of Focus	<ol style="list-style-type: none"><li>1. Patients with heart failure with a preserved ejection fraction (HFPEF) – investigations are done to rule out/in light chain amyloidosis – complete paraprotein assessments (SIFE, UIFE, sFLC) to start with - suggest &gt;90% of patients have this testing done</li><li>2. Patients with nephrotic range proteinuria – investigations are done to rule out/in light chain amyloidosis – complete paraprotein assessments to start with (SIFE, UIFE, sFLC) – suggest &gt;90% of patients have this testing done</li></ol>

### 2. Paraprotein Testing

Recommendation	ASH recommends serum and urine immunofixation and serum free light chain assay (SIFE, UIFE, sFLC) to increase clinical suspicion of cardiac AL amyloidosis
Population	Individuals with suspected cardiac amyloidosis defined (see “Red Flag Signs and Symptoms for Cardiac Amyloidosis”)
Metric	Test patients with suspected cardiac amyloidosis with serum and urine immunofixation and serum free light chain assay (SIFE, UIFE, sFLC)
Audit Metric 2	% of patients with suspected cardiac amyloidosis tested with all three assays (SIFE, UIFE, sFLC)

## Red Flag Signs and Symptoms for Cardiac Amyloidosis

- HFpEF (heart failure with preserved ejection fraction)
- Moderate or Severe LVH in absence of a significant history of untreated hypertension on imaging
- Echocardiogram: Severe left ventricular wall thickening, advanced diastolic dysfunction, reduced left ventricular global longitudinal strain with an apical sparing pattern
- EKG/Arrhythmia: Low voltage and/or discordance between voltage on EKG and left ventricular wall thickness on imaging, pseudo-infarct pattern, and arrhythmias including atrial fibrillation, heart block, and ventricular tachycardia/ventricular fibrillation
- Elevated biomarkers (Troponin and NT-Pro BNP) in absence of CAD
- Constellation of symptoms suggesting cardiac, renal, and peripheral nervous system disease
- Low Flow, Low Gradient Aortic Stenosis

## 3. Cardiac Magnetic Resonance

Recommendation	ASH <b>suggests</b> performing cardiac magnetic resonance (CMR) rather than not performing CMR to increase clinical suspicion of cardiac amyloidosis
Population	Individuals with suspected cardiac amyloidosis and positivity in any of the following studies: SIFE, UIFE, or sFLC + abnormal cardiac biomarkers, + non-diagnostic echocardiography
Metric	Perform CMR in patients with suspected cardiac amyloidosis and positivity in any of the following studies: SIFE, UIFE, or sFLC + abnormal cardiac biomarkers, + non-diagnostic echocardiography
Audit Metric 3	<b>% of patients with positive SIFE, UIFE, or sFLC + abnormal cardiac biomarkers, + non-diagnostic echocardiography who have a CMR</b>

### Definitions

- **Abnormal cardiac biomarker:** Any one or combination of the NT pro-BNP, BNP, troponin (hs, I, or T) above the reference range for that laboratory test
- **Diagnostic echocardiography:** Diagnostic echocardiographic findings that increase clinical suspicion for cardiac amyloidosis include unexplained left ventricular wall thickening, reduced left ventricular global longitudinal strain (especially when accompanied by an apical sparing pattern), myocardium with a sparkling appearance, worsening degrees of diastolic dysfunction, elevation in left ventricular filling pressures, elevations in right heart pressures, and presence of varying degrees of pericardial effusion

## 4. Bone Scintigraphy for AL Amyloidosis

Recommendation	ASH <b>recommends against</b> the use of bone scintigraphy (PYP, DPD, HMDP) for the diagnosis of AL cardiac amyloidosis
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<b>Population</b>	Individuals with a suspicion of light chain amyloidosis cardiomyopathy and serum or urine monoclonal protein or abnormal serum free light chains)
<b>Metric</b>	Not testing with bone scintigraphy in individuals with suspicion of light chain amyloidosis
<b>Audit Metric 4</b>	<b>% of patients with concern for light chain amyloidosis who are tested with bone scintigraphy</b>
<b>Remarks</b>	Immunological assays (SIFE, UIFE, sFLC) must be completed and confirmed negative before a bone scintigraphy test is done for cardiac amyloidosis

## 5. Bone Scintigraphy for ATTR Amyloidosis

<b>Recommendation</b>	ASH <b>recommends</b> the use of bone scintigraphy (PYP, DPD, HMDP) for the diagnosis of ATTR cardiac amyloidosis
<b>Population</b>	Individuals with suspicion of cardiac amyloidosis without evidence of a plasma cell disorder (normal serum free light chain levels and absence of monoclonal protein on serum and urine immunofixation electrophoresis
<b>Metric</b>	Testing with bone scintigraphy in patients with suspicion of cardiac amyloidosis and no evidence of plasma cell disorder
<b>Audit Metric 5</b>	<b>% of patients tested with bone scintigraphy in patients with suspicion of cardiac amyloidosis and no evidence of plasma cell disorder</b>
<b>Remarks</b>	Immunological assays (SIFE, UIFE, sFLC) should be done and confirmed as negative before a bone scintigraphy test is done for cardiac amyloidosis If bone scintigraphy is positive (Grade 2–3 uptake) in the absence of monoclonal protein, the diagnosis of ATTR cardiac amyloidosis is confirmed

## 6. Time to Bone Marrow Biopsy

<b>Guideline consensus</b>	Limit the time between abdominal fat pad sampling and bone marrow biopsy
<b>Population</b>	Patients with abdominal fat pad sampling
<b>Metric</b>	Time (in days) between abdominal fat pad and bone marrow biopsy
<b>Audit Metric 6</b>	<b>Median time interval between abdominal fat pad sampling and bone marrow biopsy</b>
<b>Remarks</b>	Both procedures for surrogate sites biopsy should be performed promptly to ensure adequate diagnostic sensitivity for AL amyloidosis – <b>suggest within 14 days</b>



## AL Amyloidosis Audit Feedback Report

Institution:

Date Range:

Metric #	Audit Metric	Denominator	Numerator	Compliance Rate
Audit Metric 1	Median time (in days) between initial symptom onset and confirmed diagnosis			
Audit Metric 2	% of patients with suspected cardiac amyloidosis tested with all three assays (SIFE, UIFE, sFLC)			
Audit Metric 3	% of patients with positive SIFE, UIFE, or sFLC + abnormal cardiac biomarkers, + non-diagnostic echocardiography who have a CMR			
Audit Metric 4	% of patients with concern for light chain amyloidosis who are tested with bone scintigraphy			
Audit Metric 5	% of patients tested with bone scintigraphy in patients with suspicion of cardiac amyloidosis and no evidence of plasma cell disorder			
Audit Metric 6	Median time interval (in days) between abdominal fat pad sampling and bone marrow biopsy			

Identified areas for improvement:

Completed by: