Evaluate for a monoclonal gammopathy

- a) Bone marrow aspirate & biopsy
- b) SPEP & IFE; UPEP & IFE
- c) Quantitative Ig measurement
- d) Serum FLC assessment

Evaluate for Systemic Disease and Organ Involvement

- Renal: Creatinine; 24 hour urine total protein
- Cardiac: EKG; Echocardiogram; Troponin; BNP (NT-ProBNP)
- GI/Hepatic: Alkaline phosphatase; Liver Ultrasound*; Endoscopy*
- Nerve: Orthostatics; Nerve conduction studies*; Gastric emptying*

Tissue Diagnosis by Congo red or EM

Monoclonal Gammopathy Present

- Likely AL amyloidosis
- Treatment of AL amyloidosis

Monoclonal Gammopathy Absent

- Consider other type: Proteomic Studies
- Immunogold Electron microscopy

Localized amyloidosis

Secondary amyloidosis

Familial amyloidosis

Senile Systemic amyloidosis

* If clinically indicated
† Localized amyloidosis
‡ Secondary amyloidosis
§ Familial amyloidosis
¥ Senile Systemic amyloidosis

†: kappa or lambda precursor protein; refer to system specific specialist
‡: Consider when clinically indicated i.e. chronic inflammatory condition
§: Most likely transthyretin (TTR) mutation but consider other hereditary forms
¥: Wild type TTR; often isolated cardiac involvement

†† Indications for familial testing with or w/out a monoclonal gammopathy
1. African Americans for mutant TTR
2. Dominant peripheral nervous system involvement
3. Isolated renal involvement and no amyloid in bone marrow; screen for fibrinogen Aα
4. Family history concerning for familial amyloidosis