



# Cardiac amyloidosis with a cherry on top



John Stacy, MD MS<sup>1</sup>; Natalie Longino, MD, PhD<sup>1</sup>; Jennifer Doran, MD<sup>1</sup>; Jeffrey Spence, MD<sup>2</sup>; Noelle Northcutt, MD<sup>2</sup>

1. University of Colorado Internal Medicine Residency Program; 2. Denver Health Medical Center

## Patient Presentation

A 66-year-old African American male with a history of chronic obstructive pulmonary disease (COPD) on 2 liters of oxygen at baseline, left ventricular hypertrophy (LVH), heart failure with preserved ejection fraction (HFpEF) status-post cardiac resynchronization therapy for prior heart failure with reduced ejection fraction (HFrEF) and chronic kidney disease stage III (baseline creatinine 1.9) was admitted for altered mental status and failure to thrive.

- Vitals: T 36.9; HR 64; RR 16; BP 143/88; SpO2 94% on home 2 L
- Exam: Disheveled, minimal bibasilar crackles, no LE edema JVP not elevated
- Labs: WBC 5.8, Hgb 10.8, BUN 36, Cr 1.82; HS troponin I 41; NT-proBNP 1,417
- ECG: A sensed, V paced, e/o LVH
- Deemed not to have capacity, patient is entering his fifth month of hospitalization in pursuance of guardianship

### Blood pressure lability:

Systolic pressures frequently fluctuated from 190's to 90's after treatment with home Coreg

### Acute on chronic respiratory insufficiency (AHRI) and acute kidney injury (AKI):

- Progressive increase in O2 requirement, maximum 6 L
- Progressively rising Cr raised suspicion for cardiorenal syndrome
- Diuresis was initiated without relief, subsequently an echo was ordered
  - Echo findings: LV longitudinal strain pattern with apical sparing, so called "cherry on top" finding 93% sensitive and 82% specific for cardiac amyloidosis (CA)
- Serum light chain (AL) studies subsequently obtained:
  - Elevated kappa level (31 mg/dl)
  - Elevated kappa:lambda ratio (2.54)
- Presumed diagnosis of AL CA made, diuresis and beta blockade were discontinued
- Further workup and treatment deferred after being deemed outside the scope of the patient's goals of care

## Imaging

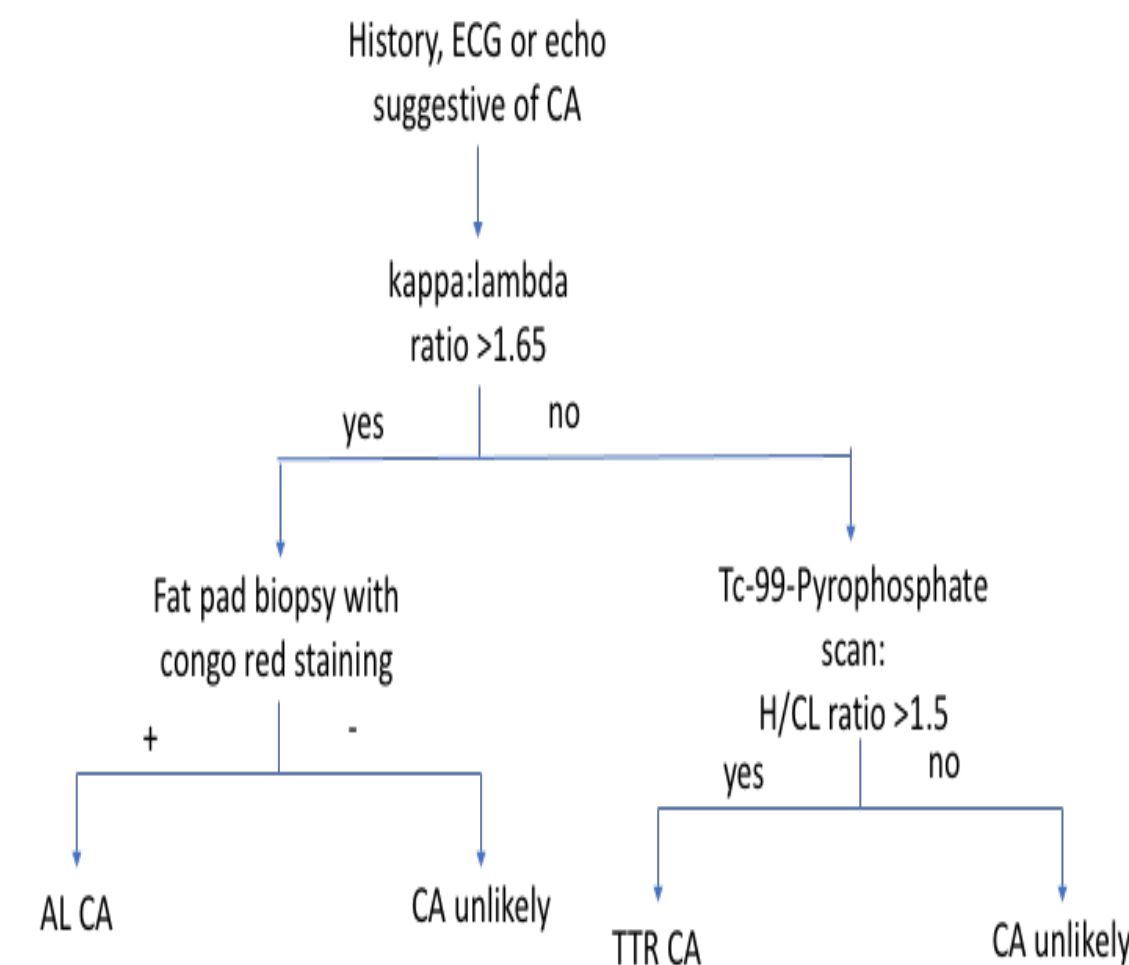


Figure 1: CA diagnosis algorithm

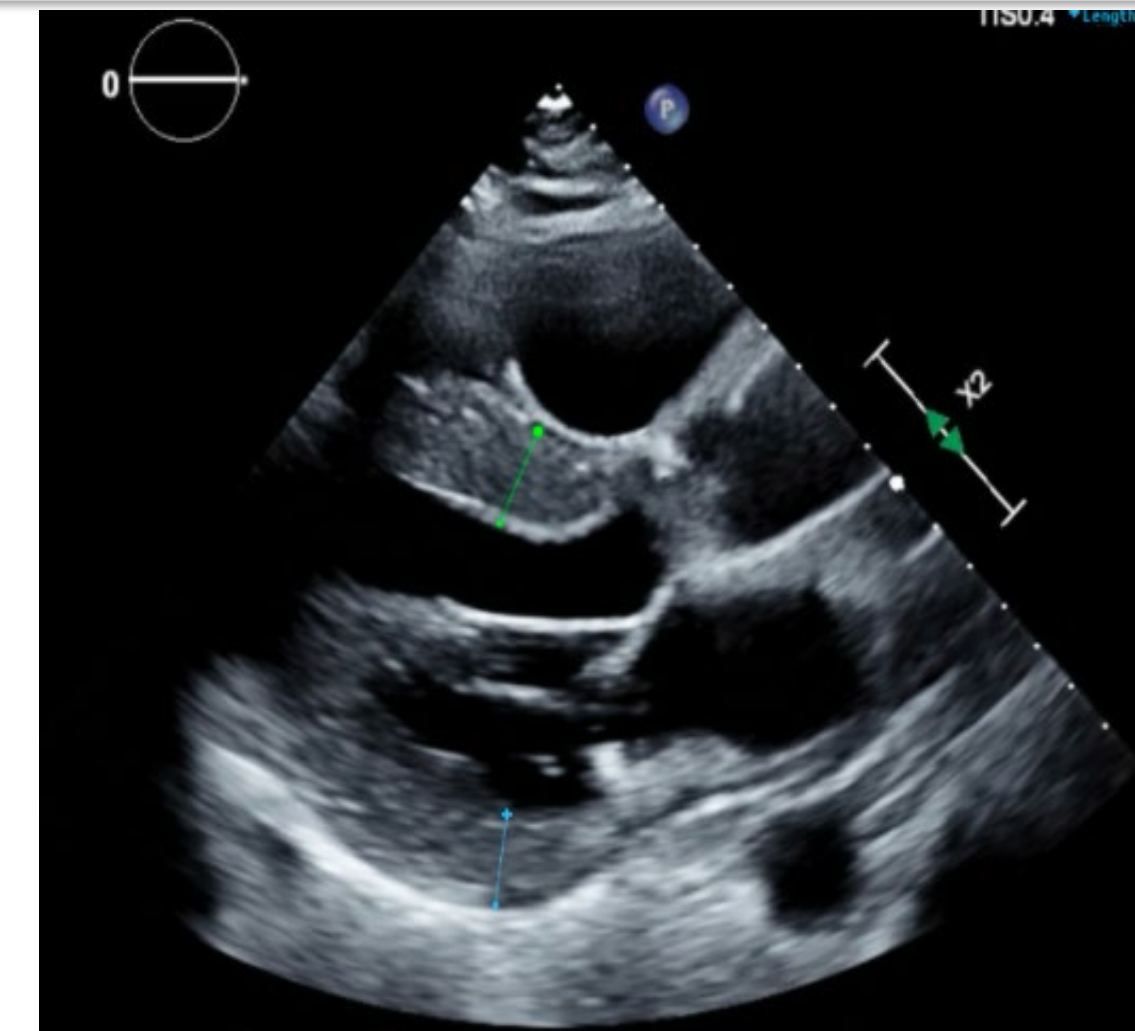


Figure 2: Echocardiogram showing diffuse LV wall thickening

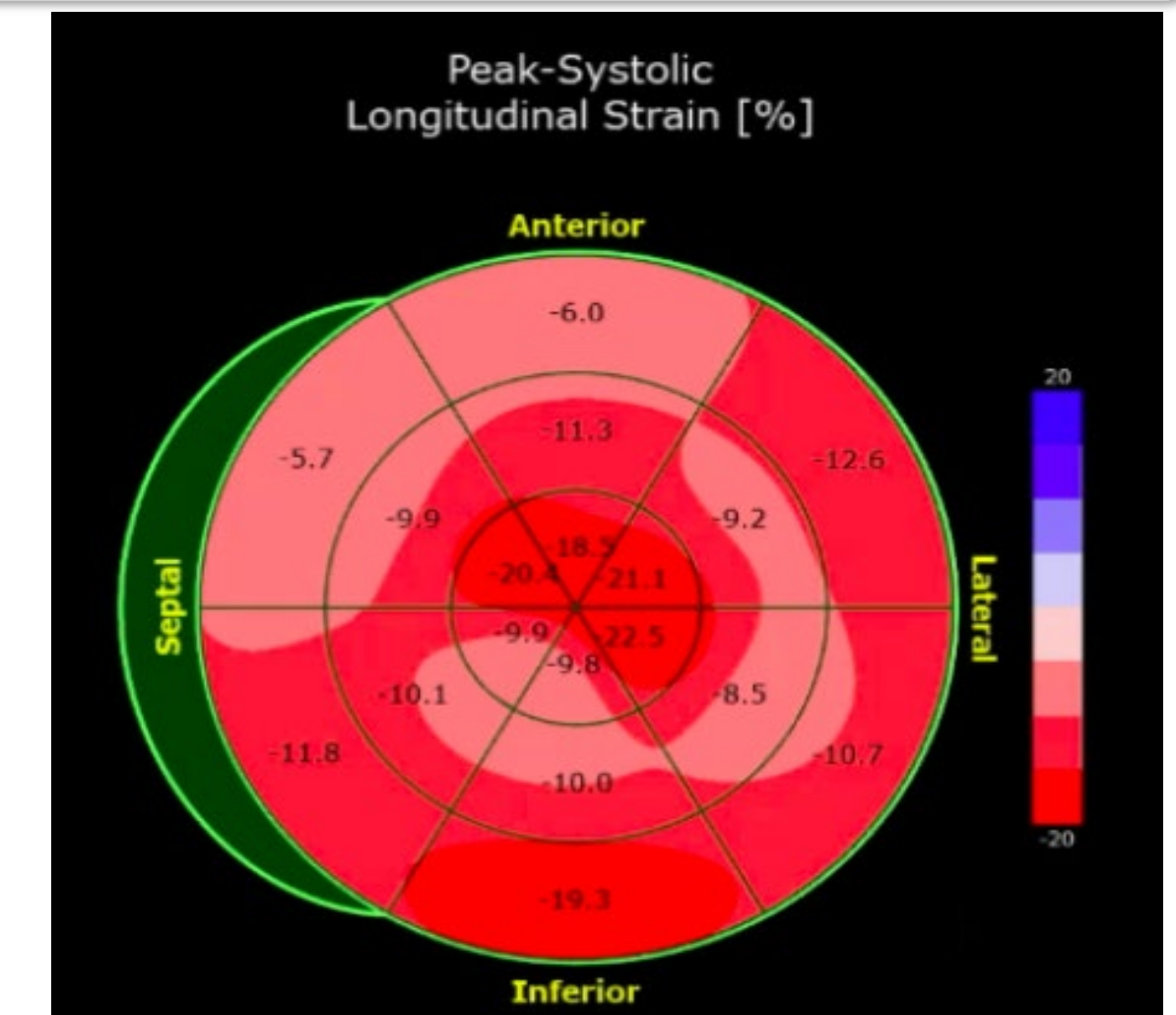


Figure 3: Longitudinal strain pattern showing preserved apical longitudinal shortening and impaired basilar shortening, the "cherry on top" pattern

## Discussion

### What is CA?

Deposition of misfolded proteins in the myocardium leading to LV wall thickening and a restrictive cardiomyopathy

- Monoclonal immunoglobulin light chains (ALs)
- Liver-synthesized transthyretin (TTR)

### When to consider CA as HF etiology?

Despite its perceived rarity, amyloidosis is a relatively common cause of HFpEF, especially in certain demographics:

- Up to 30% of HFpEF hospitalizations in those >75 may be due to ATTR amyloidosis
- The prevalence in African Americans is 3-4 times that in Caucasians
- Individuals with LV wall thickness >14 mm should be considered for a CA diagnosis

### Why Identification of CA as HF Etiology is Important:

Treatment of CA induced HF differs from other etiologies:

- ACEi/ARBs and are poorly tolerated in CA inducing hypotension secondary to a small LV cavity and inability to compensate for vasodilation with increased stroke volume
- Beta blockade further inhibits stroke volume compensation and can lead to development or exacerbation of bradyarrhythmias
- Digoxin has been shown to bind amyloid fibrils, increasing the incidence of toxicity even in the setting of normal circulating levels

Early diagnosis and treatment with novel drugs leads to better outcomes:

- AL: cardiac involvement w/out chemotherapy → median survival of 6 mo
- TTR: 75 mo median survival, 2 biologics now approved for treatment of TTR associated neuropathy

## References

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