85-year-old male presenting with worsening fatigue, dyspnea on exertion, and weight gain

Mohaned Serdah, MS3

Edward Gillis, DO

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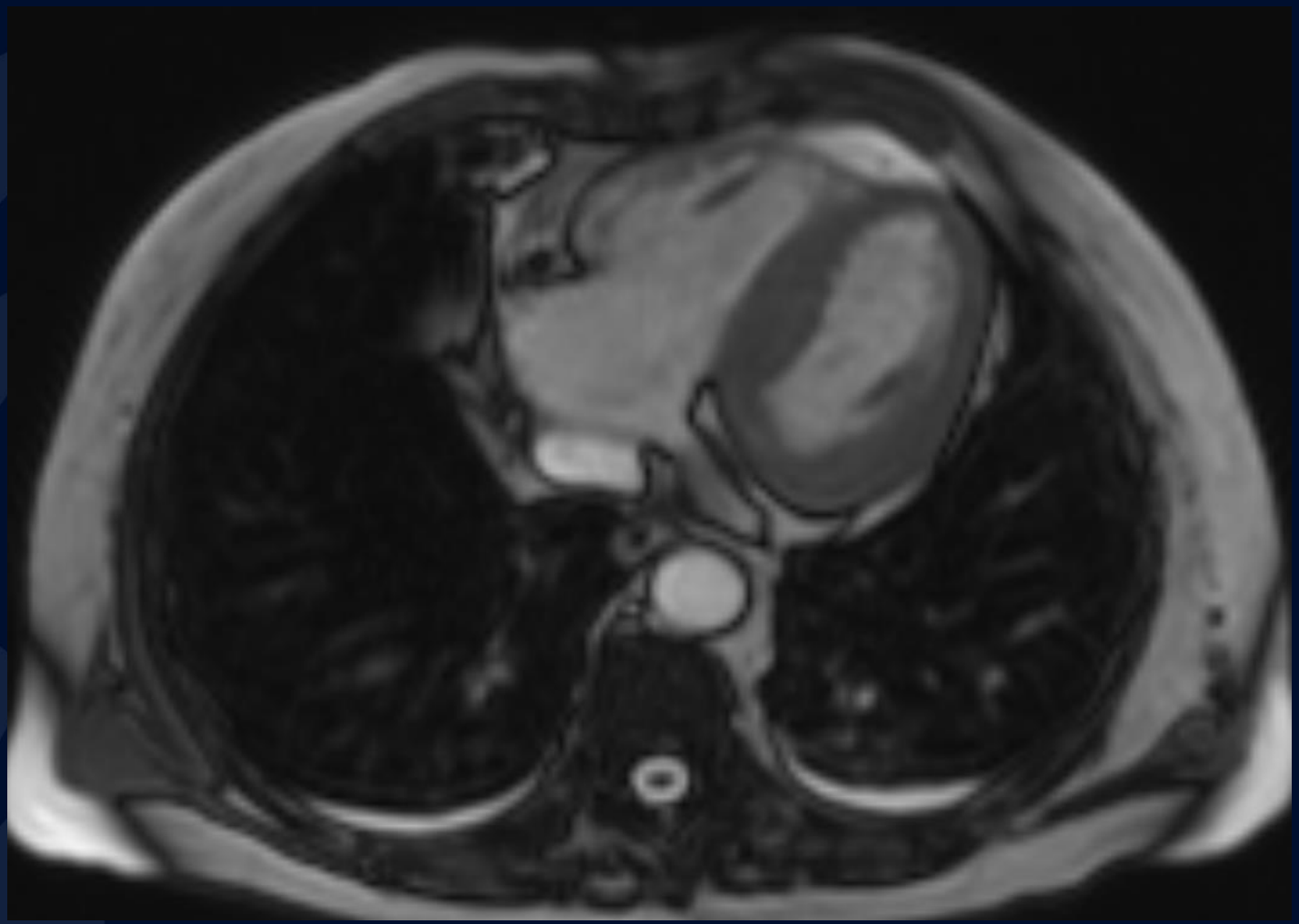
Cardiac MR- Axial Scout



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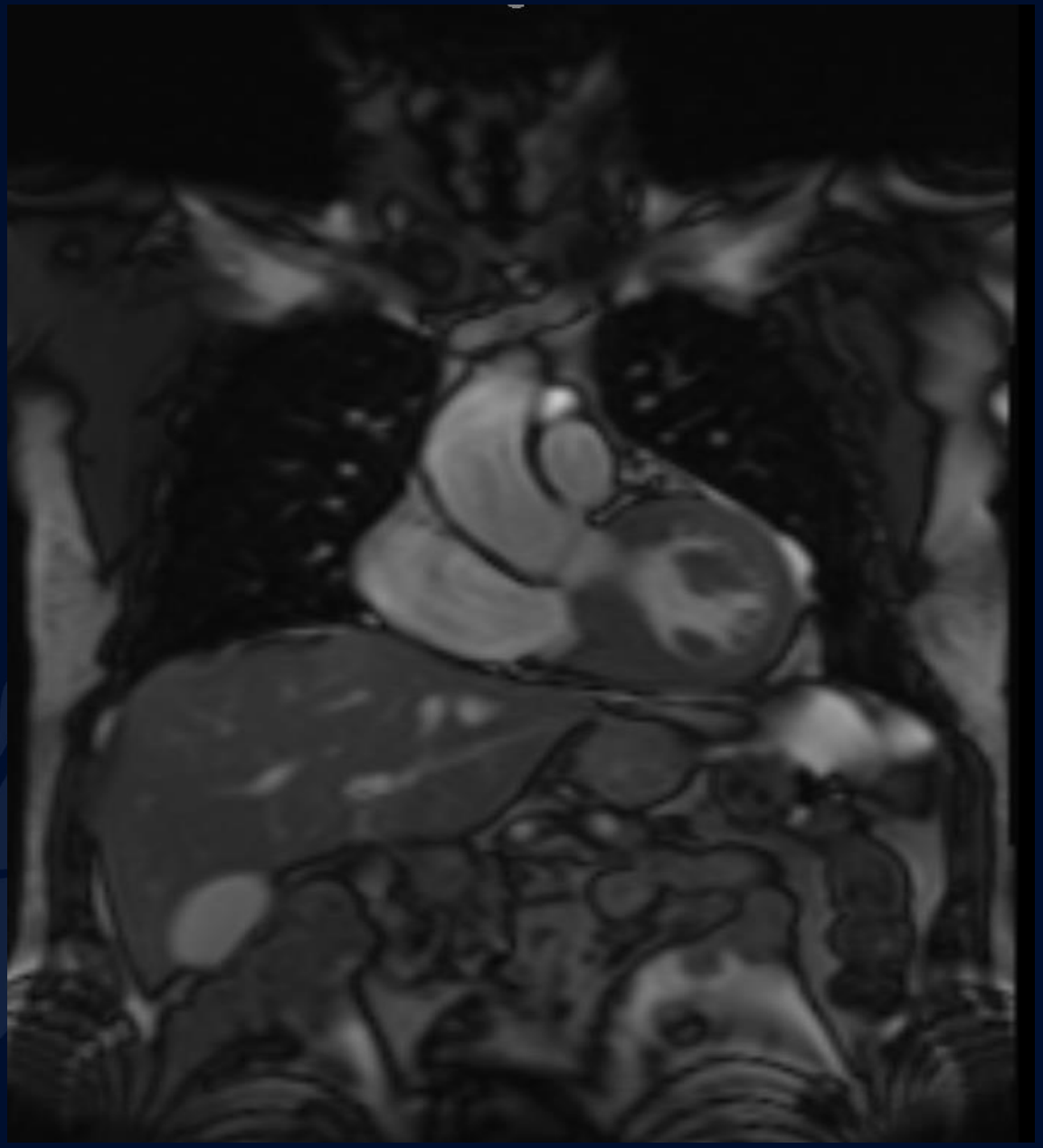
Cardiac MR- Coronal Scout



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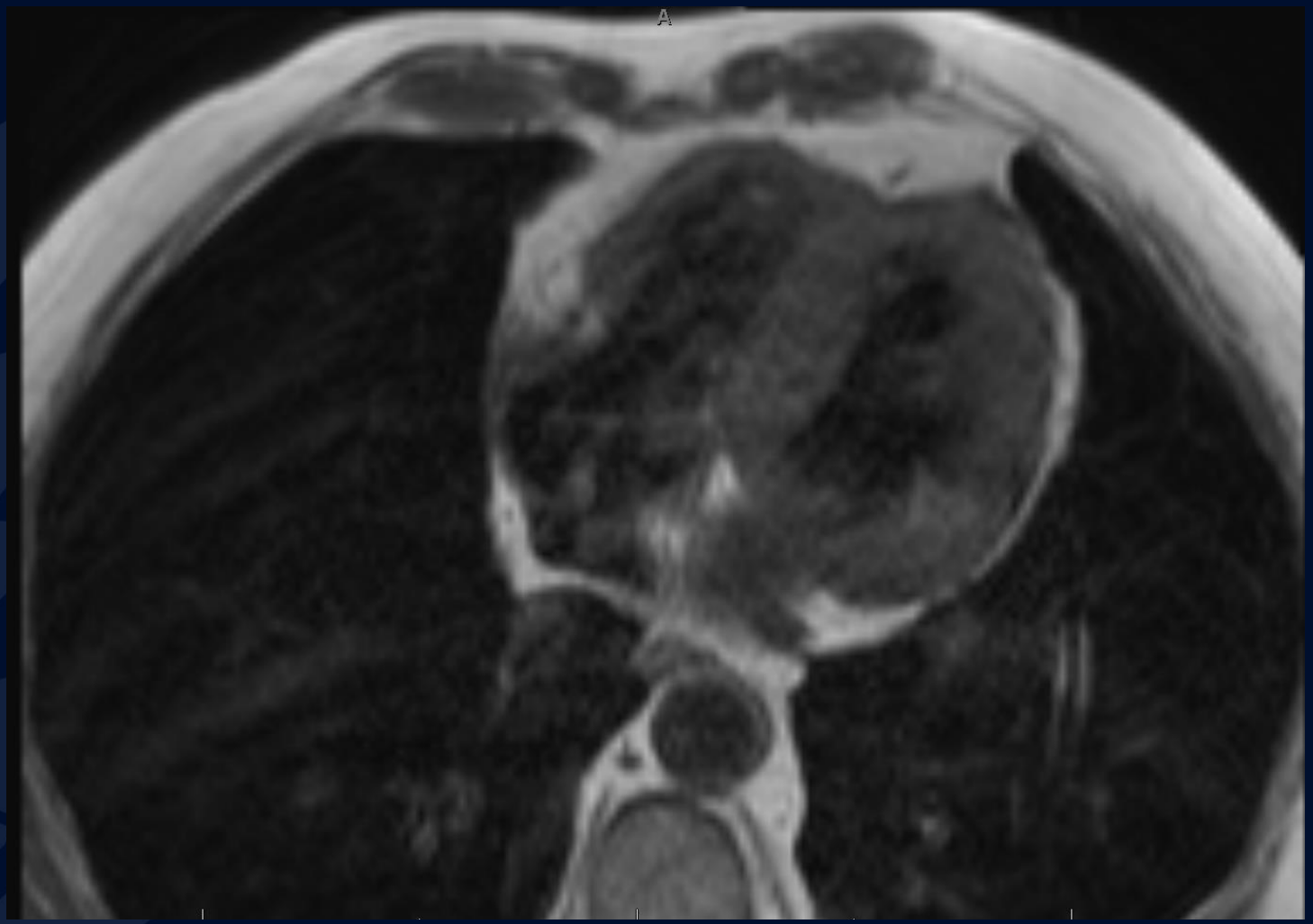
T1- Axial TSE



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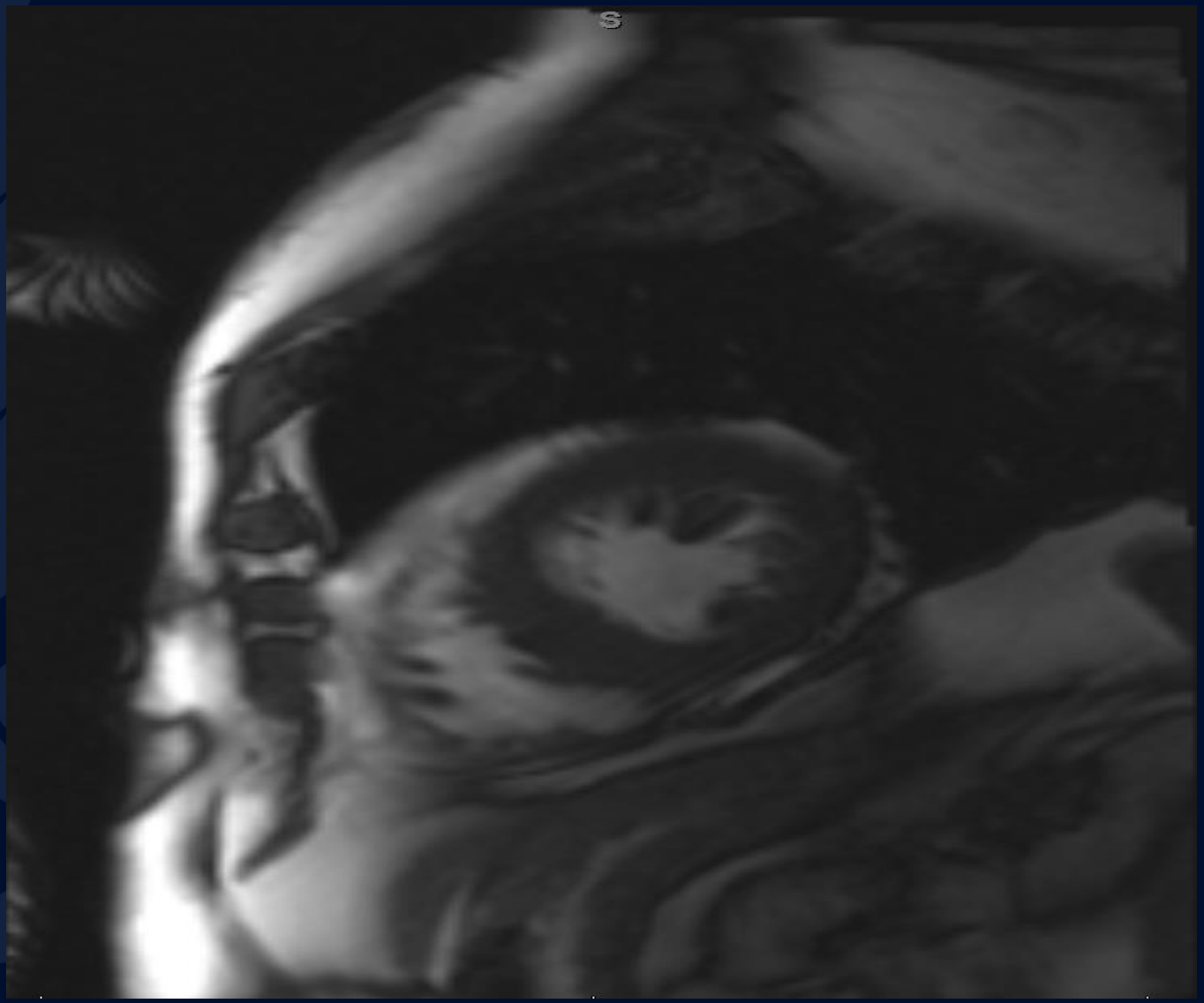
Short Axis



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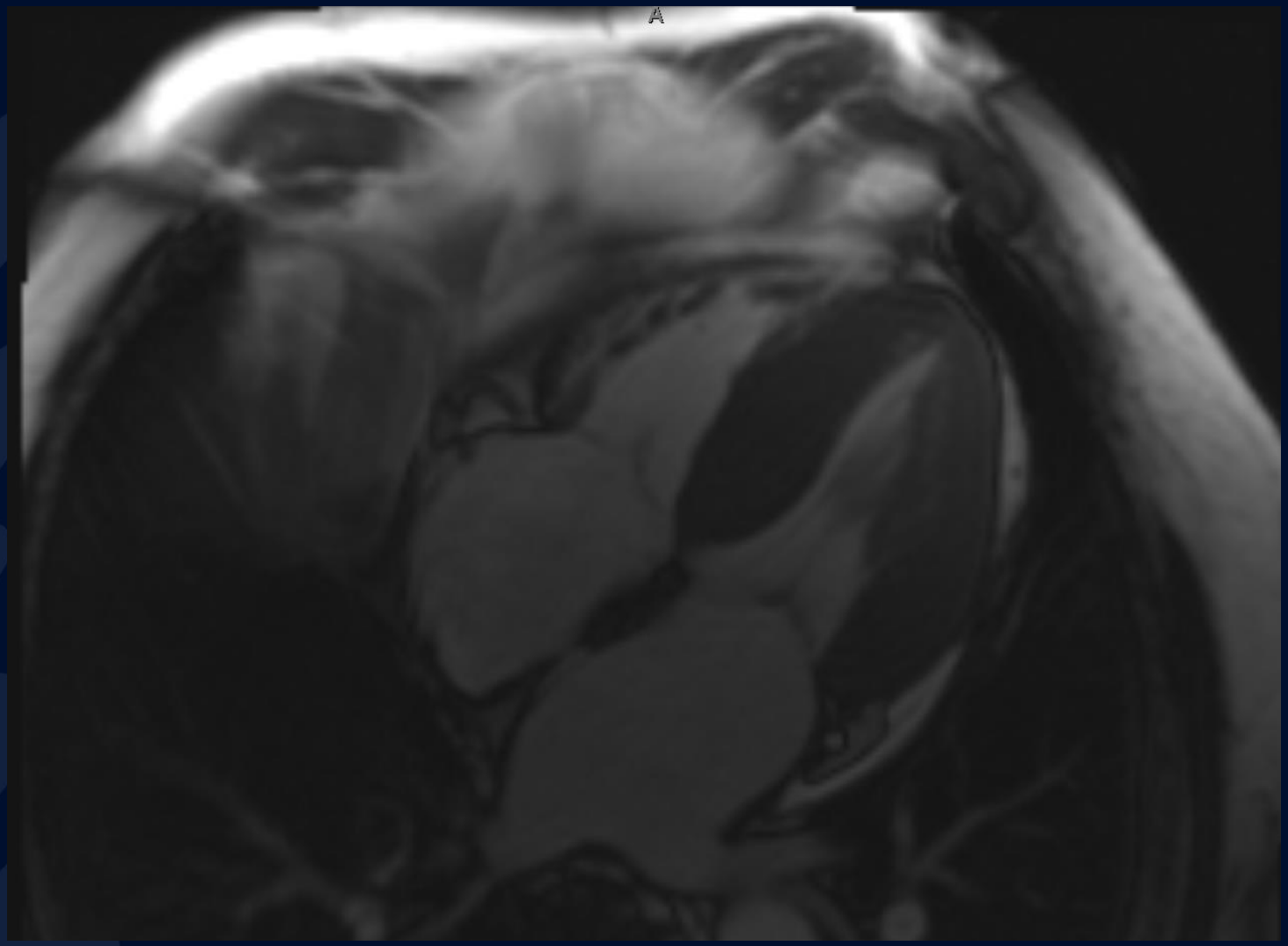
4 Chamber View



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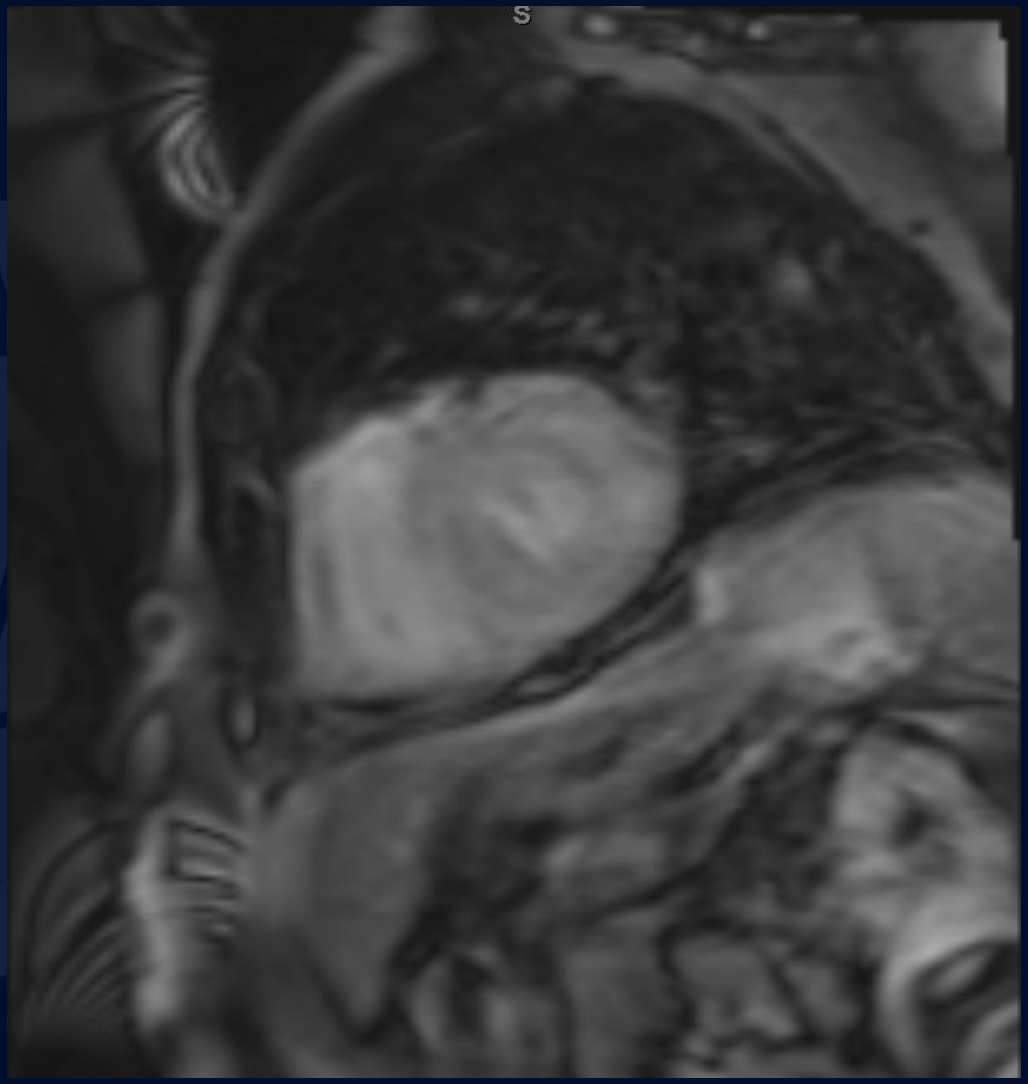
T1 Scout- Pre-Gadolinium



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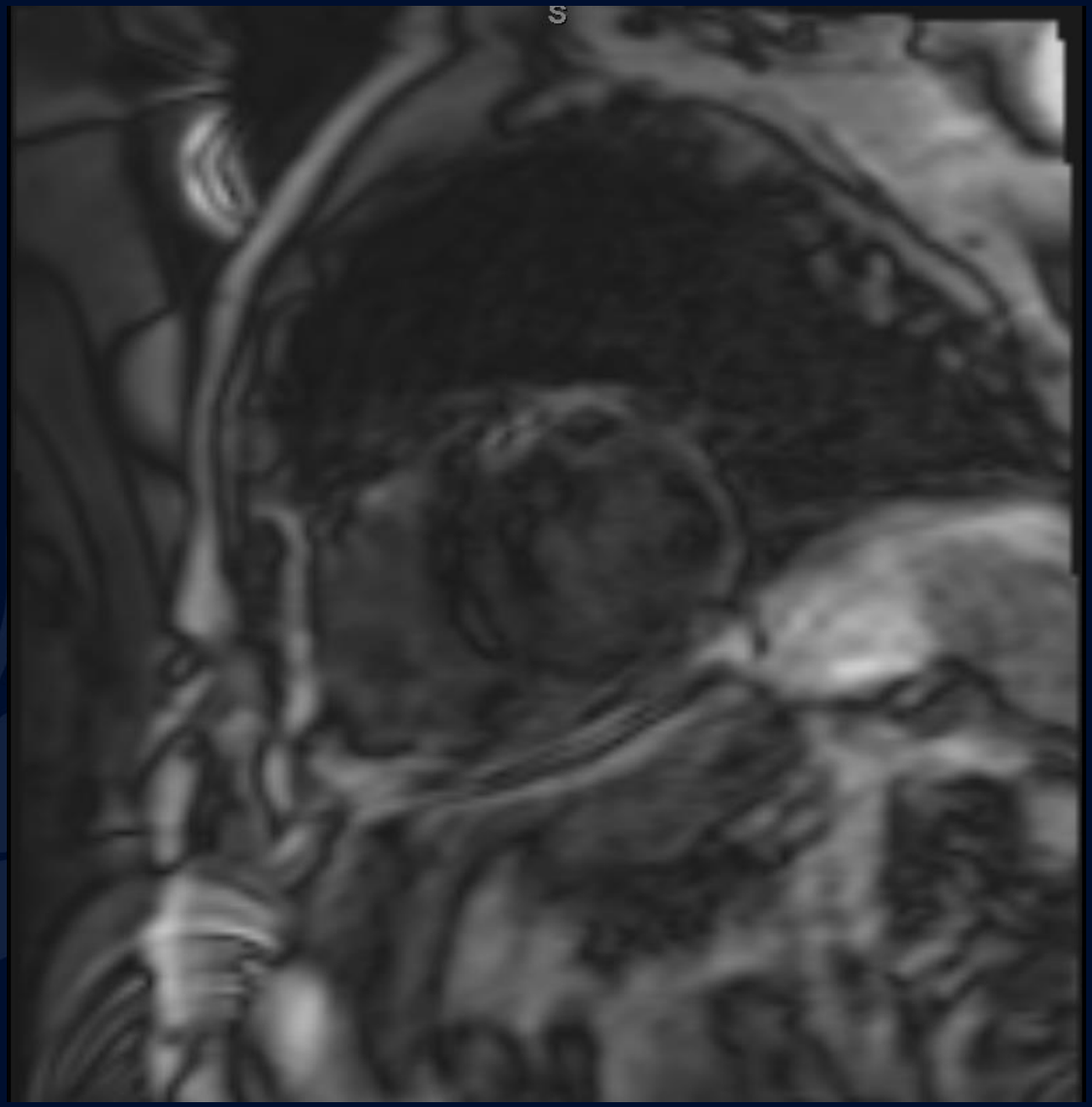
T1 Scout- Post Gadolinium



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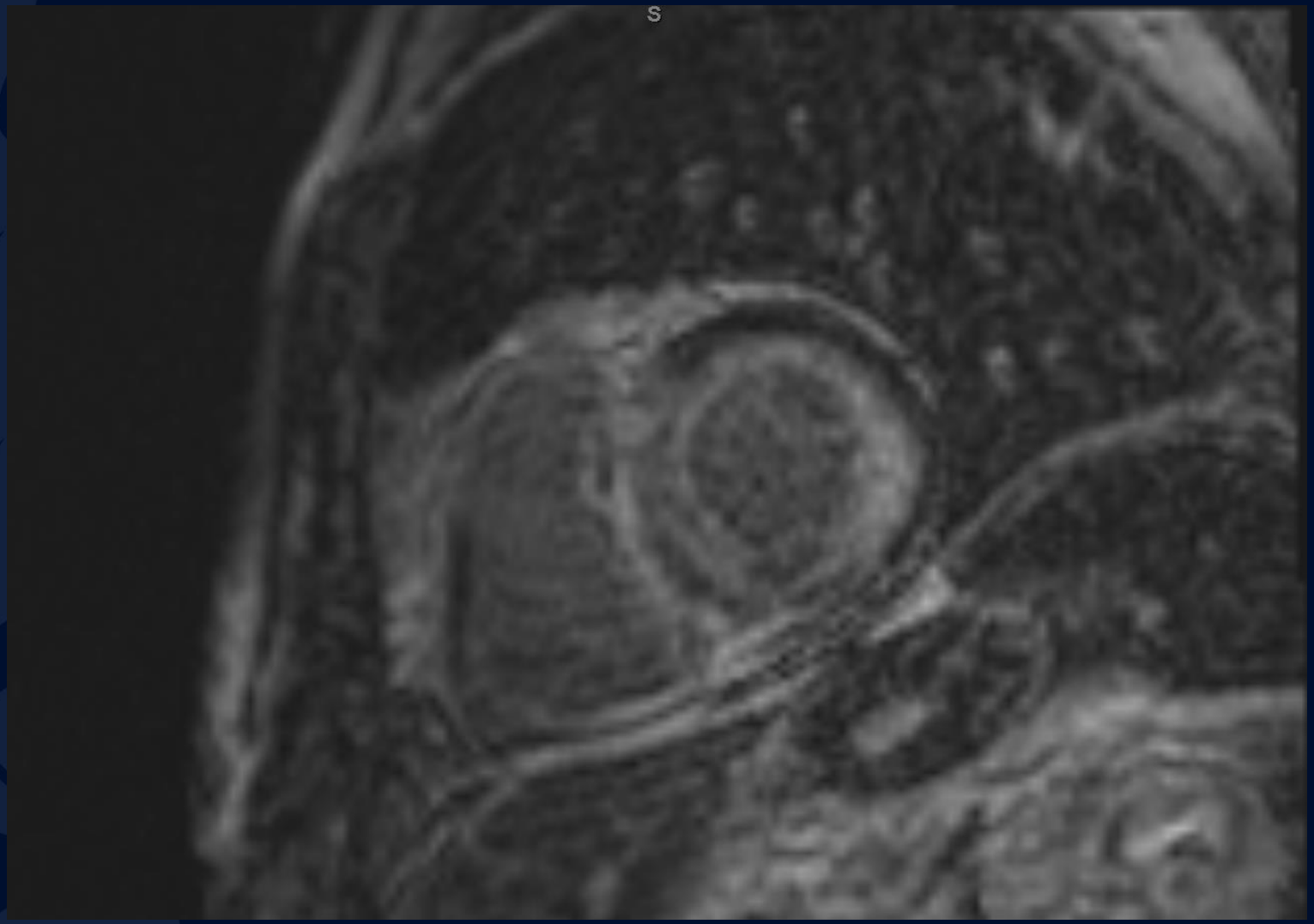
Short Axis Late Gadolinium Enhancement



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Cardiac Amyloidosis

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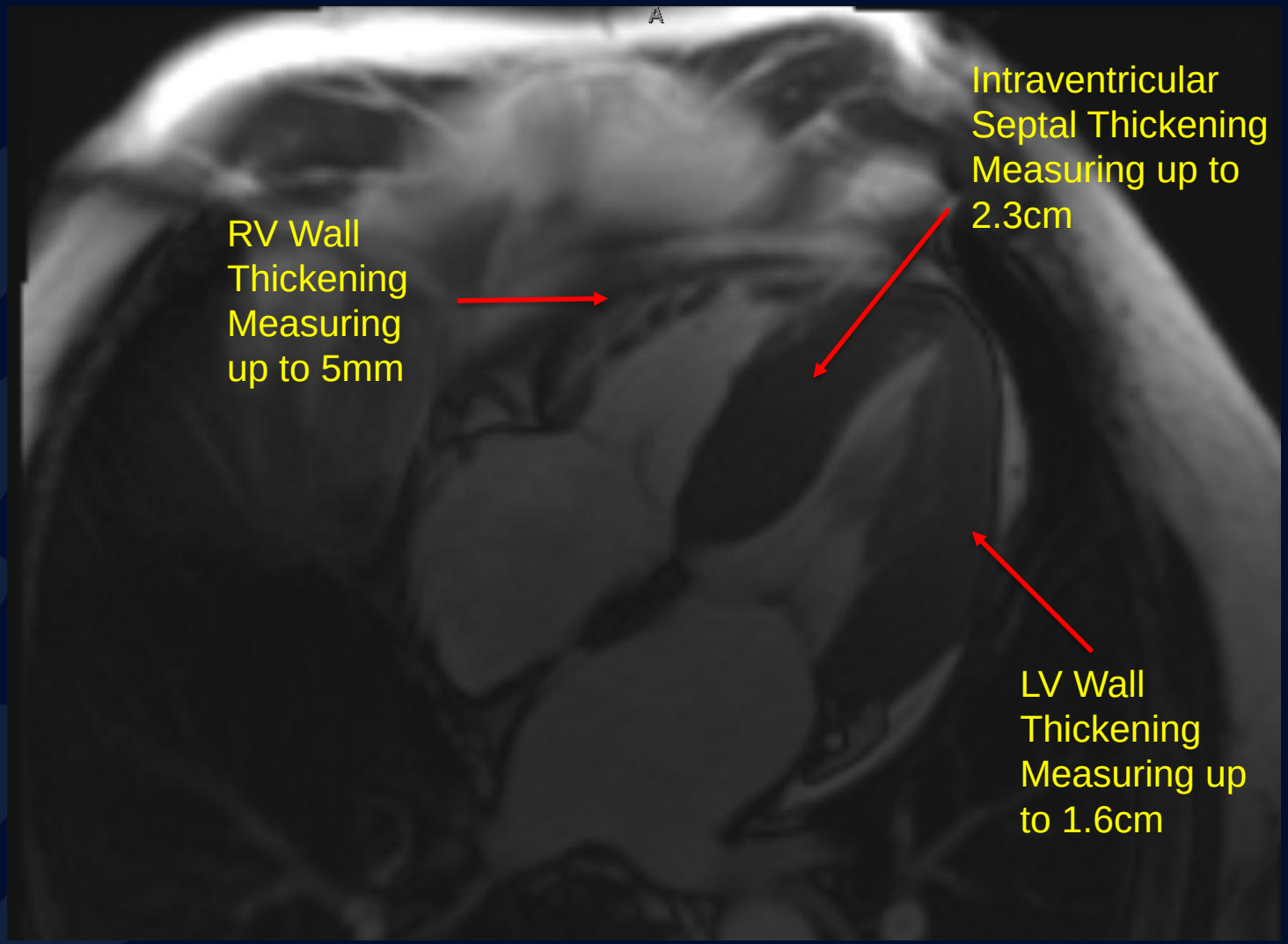
4-Chamber View



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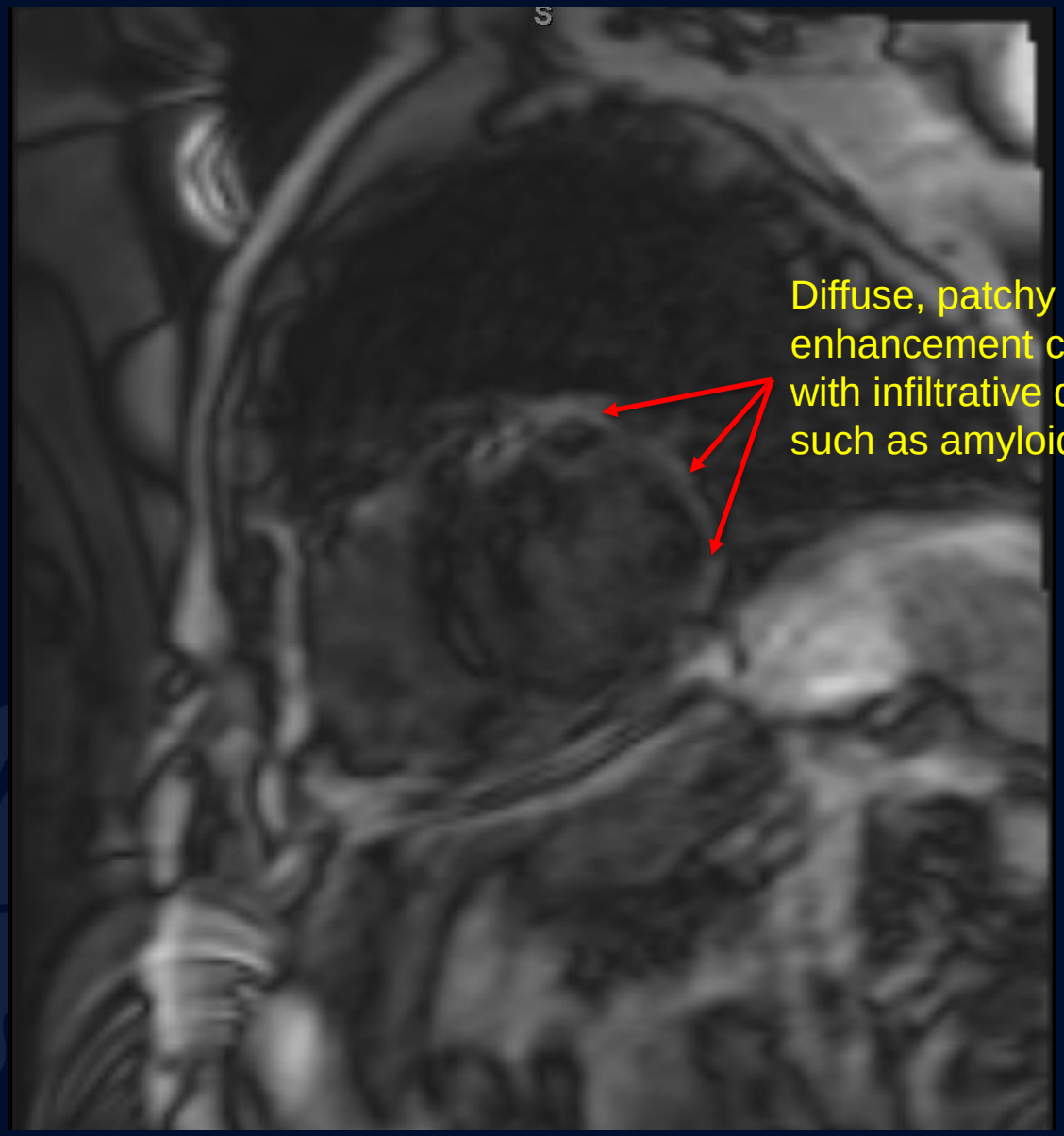
T1 Scout- Post Gadolinium



Diffuse, patchy enhancement consistent with infiltrative disease such as amyloidosis.

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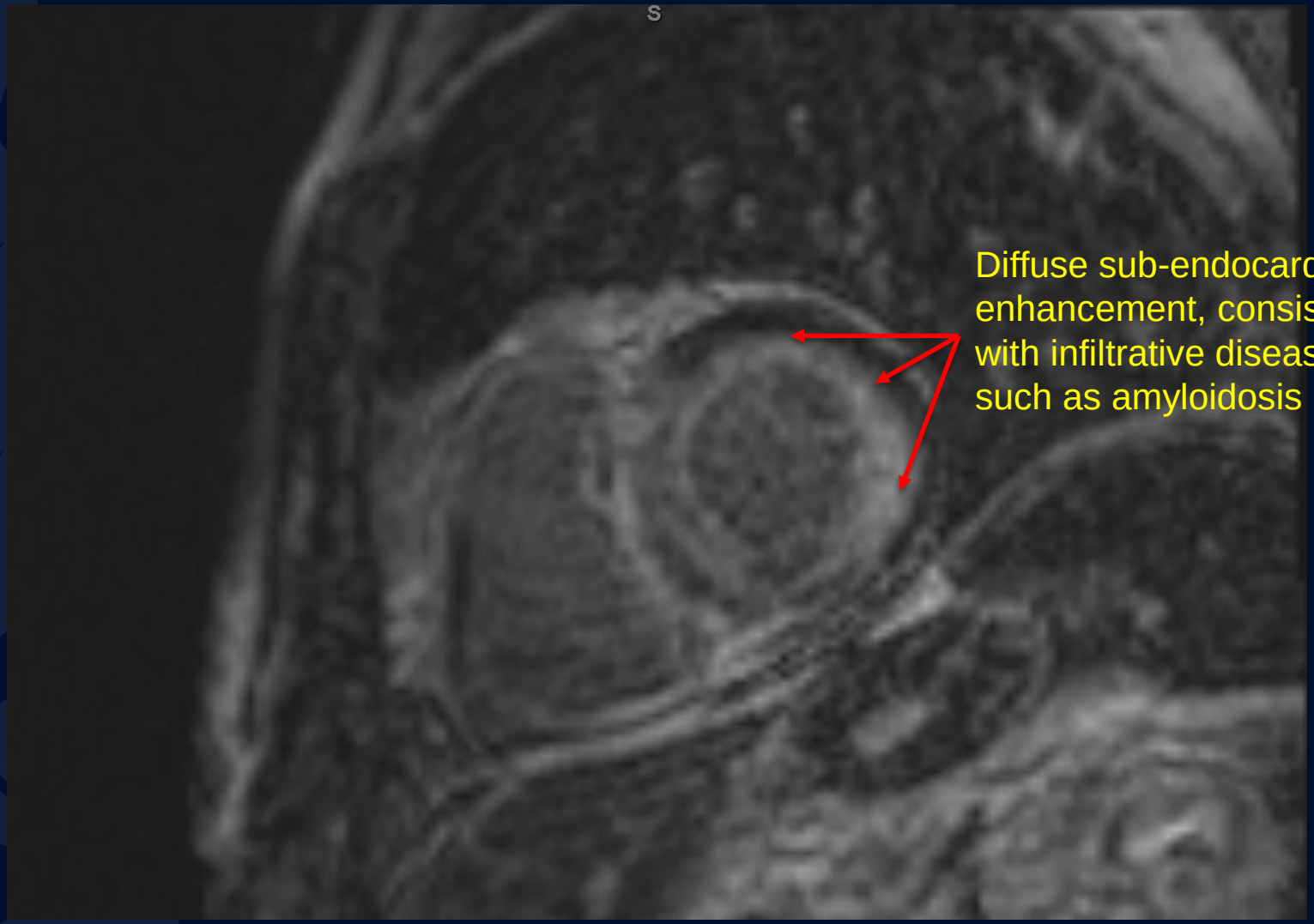
Short Axis Late Gadolinium Enhancement



Diffuse sub-endocardial enhancement, consistent with infiltrative disease such as amyloidosis

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Cardiac Amyloidosis

- Cardiac amyloidosis is protein deposition and aggregation of amyloid within the myocardium.
  - Involvement can be sub-endocardial or transmural depending on the type of amyloid disease.

- There are two types of amyloid disease which make up 95% of cardiac amyloidosis:
  - Light Chain Amyloidosis (AL)
  - Transthyretin Amyloidosis (ATTR)

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Cardiac Amyloidosis

- Amyloid cardiomyopathy typically manifests as symptoms of heart failure, which include:
  - Dyspnea, fatigue, edema, angina, pre-syncope.

- Severe cardiac amyloidosis does lead to HFpEF because the thickened ventricular myocardium results in impaired diastolic function/filling.

- Additional cardiac manifestations include small vessel disease, conduction system disease, and pericardial disease.

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Cardiac Amyloidosis

- Echocardiogram shows LV wall thickness and EKG shows low voltage amplitudes.

- Cardiac MRI is also useful in diagnosis.
  - Typical findings include late gadolinium enhancement with diffuse, patchy involvement.
  - Cardiac MRI is highly sensitive (93%) and specific (70%).

- Endo-myocardial biopsy is nearly 100% sensitive, and is used for definitive diagnosis.

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Cardiac Amyloidosis

- Management of the disease depends on the type of amyloidosis the patient has.
  - AL can be treated with alkylating agents, proteasome inhibitors, immuno-modulators, and anti-CD 38 monoclonal antibody.
  - ATTR can be treated with TTR blockers, silencers, and fibril disrupters.
- Regardless of the type of amyloid disease, patients with HFpEF are treated symptomatically.
  - Treatment includes salt restriction and diuresis.

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References

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3. Dubrey, et al. “Clinical Features of Immunoglobulin Light-Chain (AL) Amyloidosis with Heart Involvement.” *OUP Academic*, Oxford University Press, 1 Feb. 1998, academic.oup.com/qjmed/article/91/2/141/1582352.
4. Serdah, M., Kincaid, B., Gillis E. Cardiac Amyloidosis. Radiology Online 2020. University of Connecticut Health.

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