46 year old man with fatigue and lower extremity swelling

Rahul Dey, MD
Clifford Yang, MD

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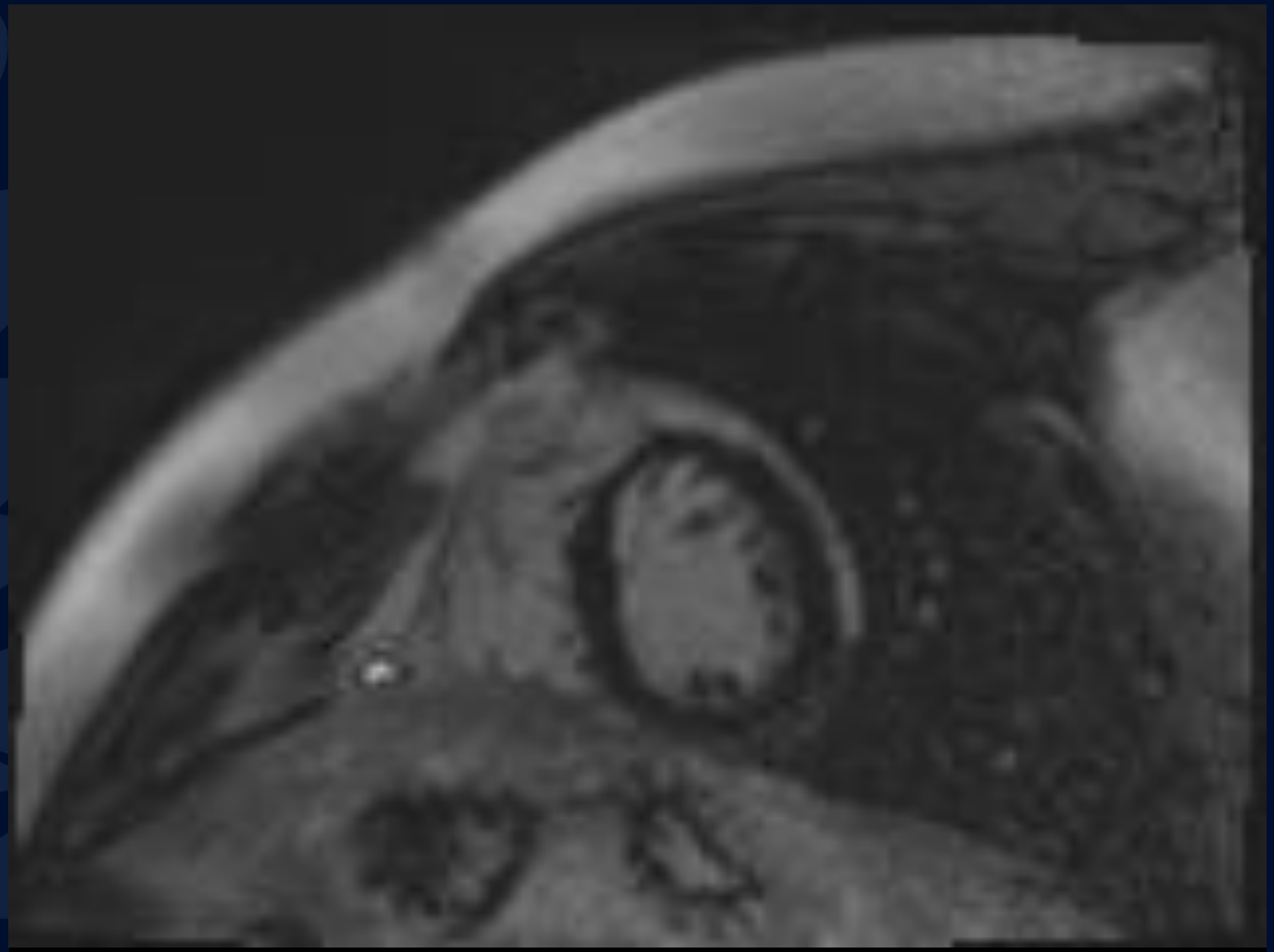

Short Axis Late Gadolinium Enhancement MR

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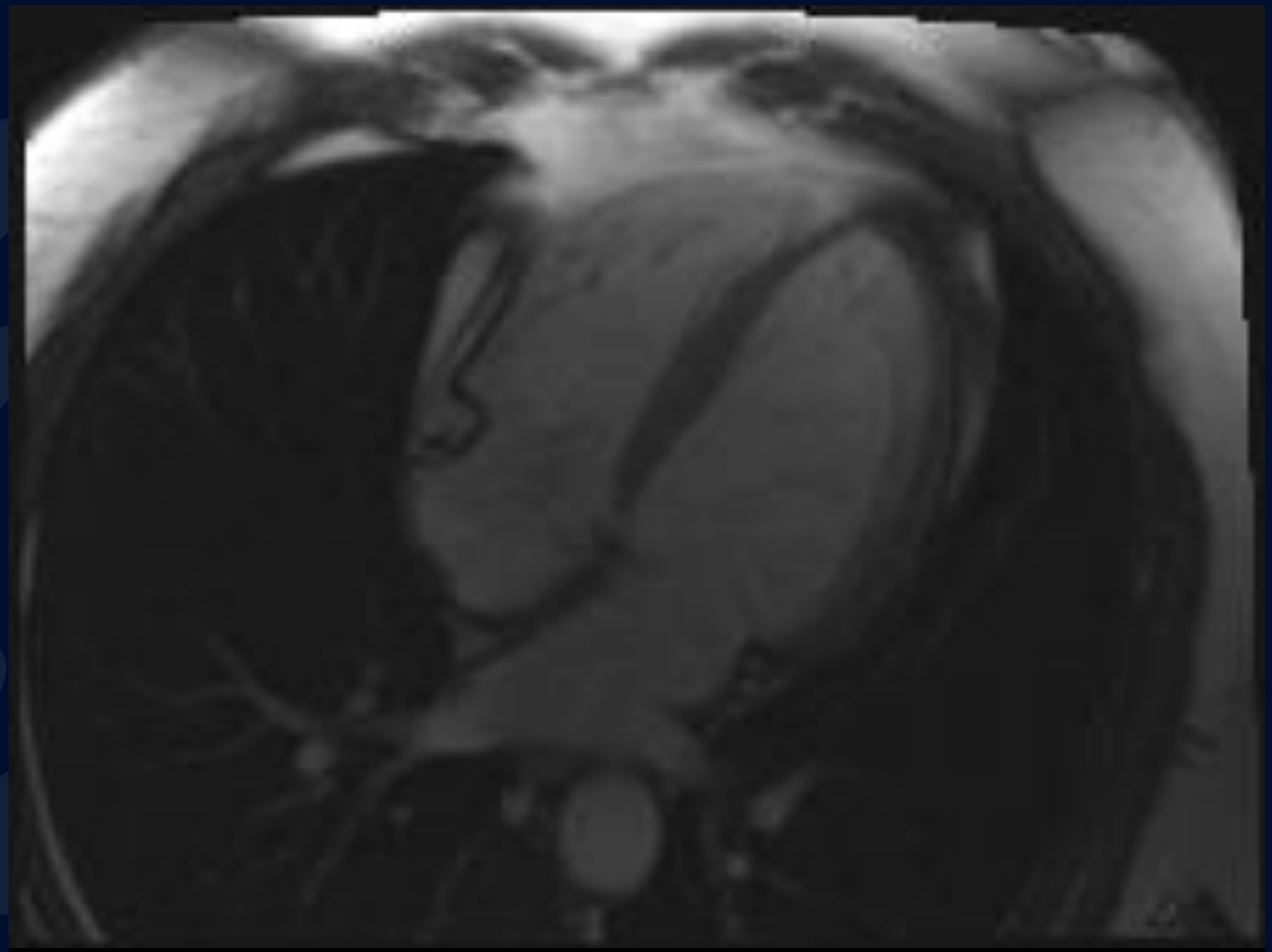

4 Chamber Steady State Free Precession MR

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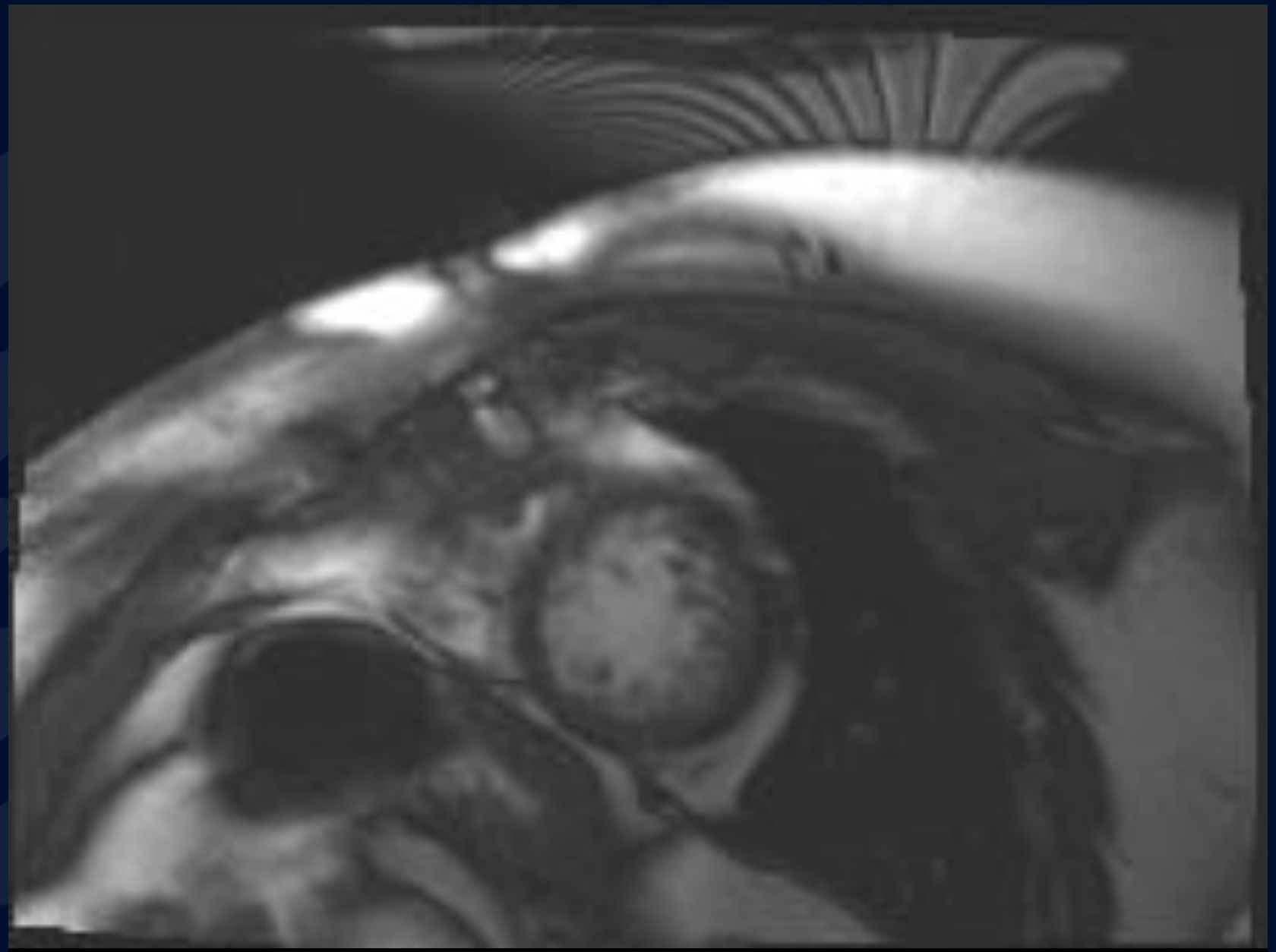

Short Axis Steady State Free Precession MR

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Non-compaction Cardiomyopathy

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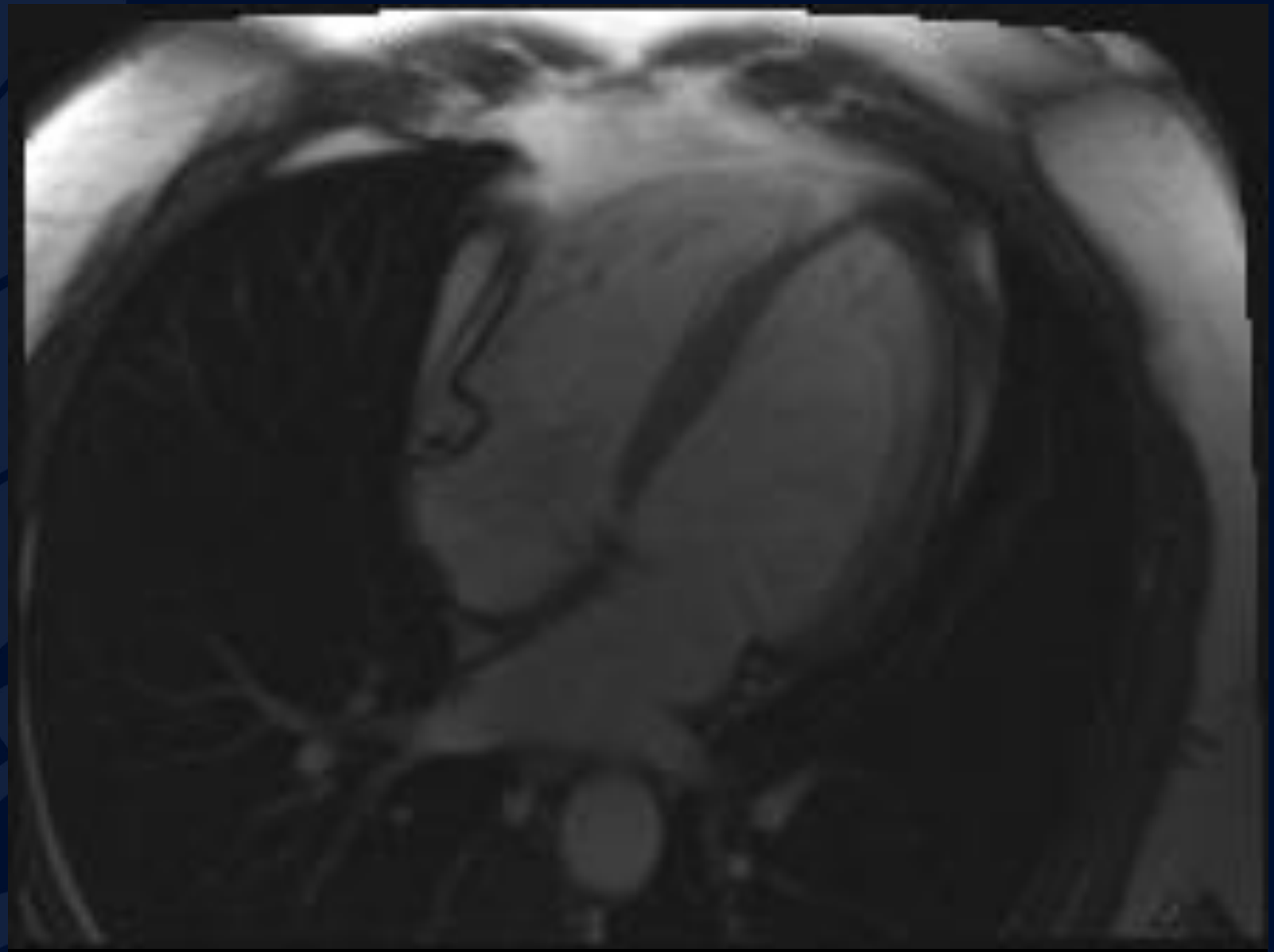

4 Chamber Steady State Free Precession MR

Relatively thickened trabeculae relative to myocardial wall at apex

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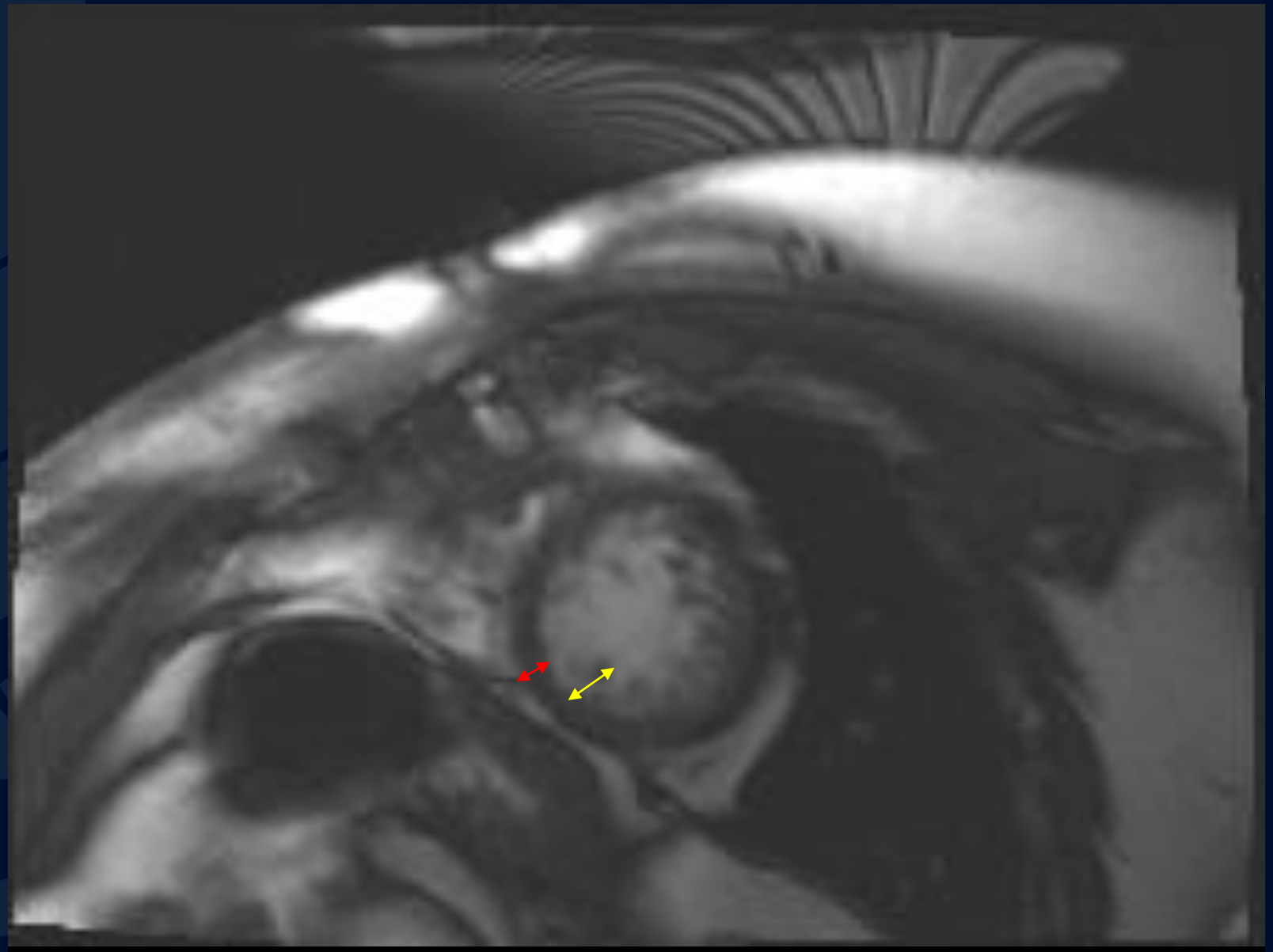

Short Axis Steady State Free Precession MR

Ratio of non compacted myocardium (yellow arrow) to compacted myocardium (red arrow) is >2.3

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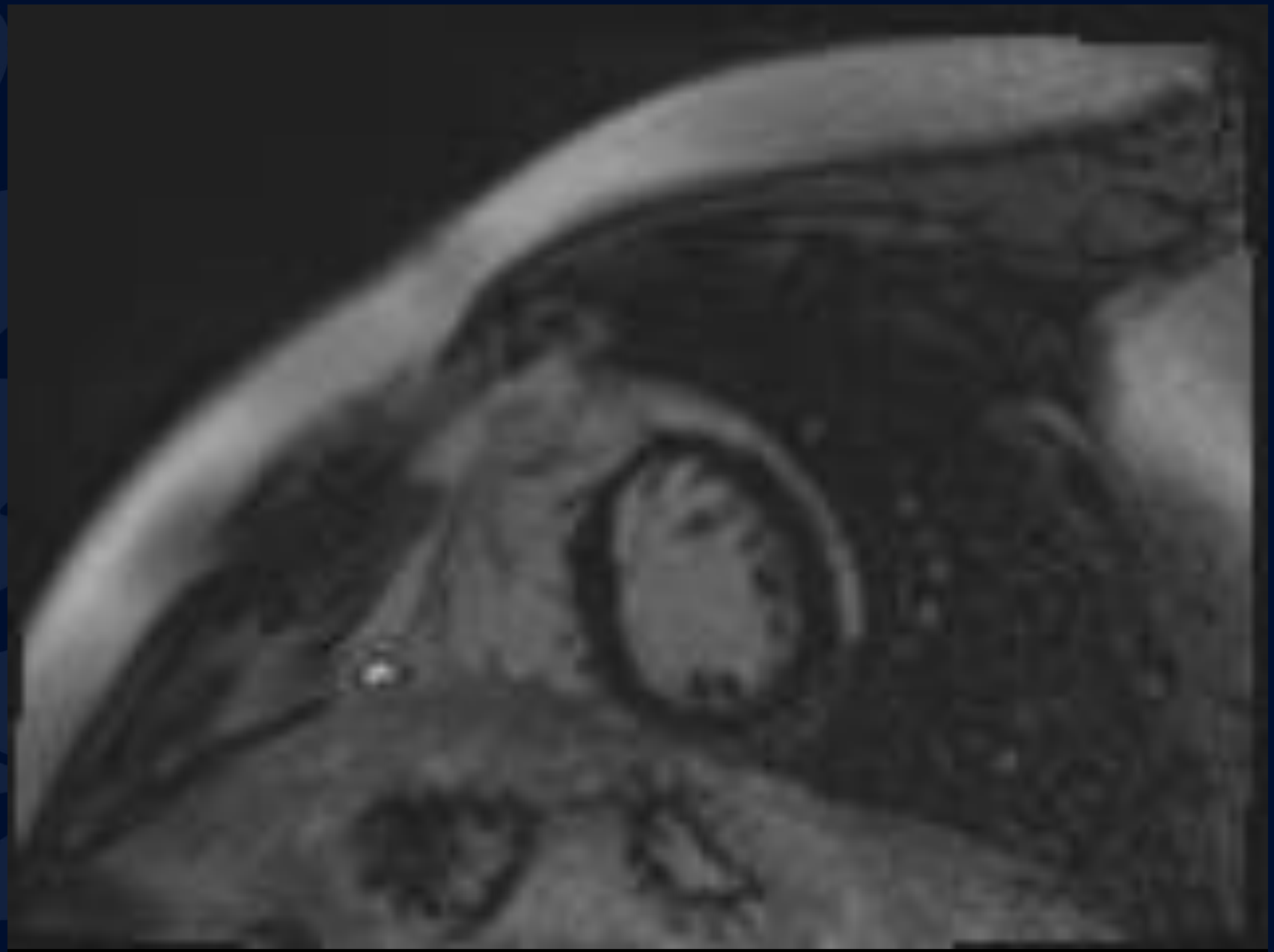

Short Axis Late Gadolinium Enhancement MR

Normal late gadolinium enhancement without fibrosis

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Non-compaction cardiomyopathy

- Rare cardiomyopathy characterized by prominent left ventricular trabeculation, deep intertrabecular recesses, and a two layered myocardium consisting of compacted and non compacted layers
- Thought to occur due to abnormal intrauterine trabecular compaction, in which the primitive ventricular trabeculation network does not properly undergo remodeling to form the specialized contractile structure
- Largely genetic (in one systematic review, etiology was genetic in 83% of patients)
- Classically presents with a triad of heart failure (HF), arrythmias, and cardio-thromboembolic events
  - Often asymptomatic (up to 50%)
- Treatment: based on existing recommendations for HF/treated symptomatically
  - ACE-Is, Beta blockers, Aldosterone antagonists
  - Implantable cardioverter-defibrillator in cases with ventricular tachyarrythmias

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Imaging findings

- Transthoracic echocardiogram: compacted (C) thin epicardial band and a much thicker non compacted (NC) endocardial layer
  - Maximal NC/C ratio >2 at end systole on short axis parasternal view
- Cardiac MR: NC/C myocardium ration of >2.3 at end diastole
  - Late Gadolinium Enhancement (LGE) and T1 mapping identifies myocardial fibrosis (T1 mapping more sensitive)
- Cardiac CT useful for functional and anatomic assessment of ventricles and for excluding coronary artery disease

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References

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