“Rapid fire rhythm”: Multimodal Imaging Uncovers the Culprit

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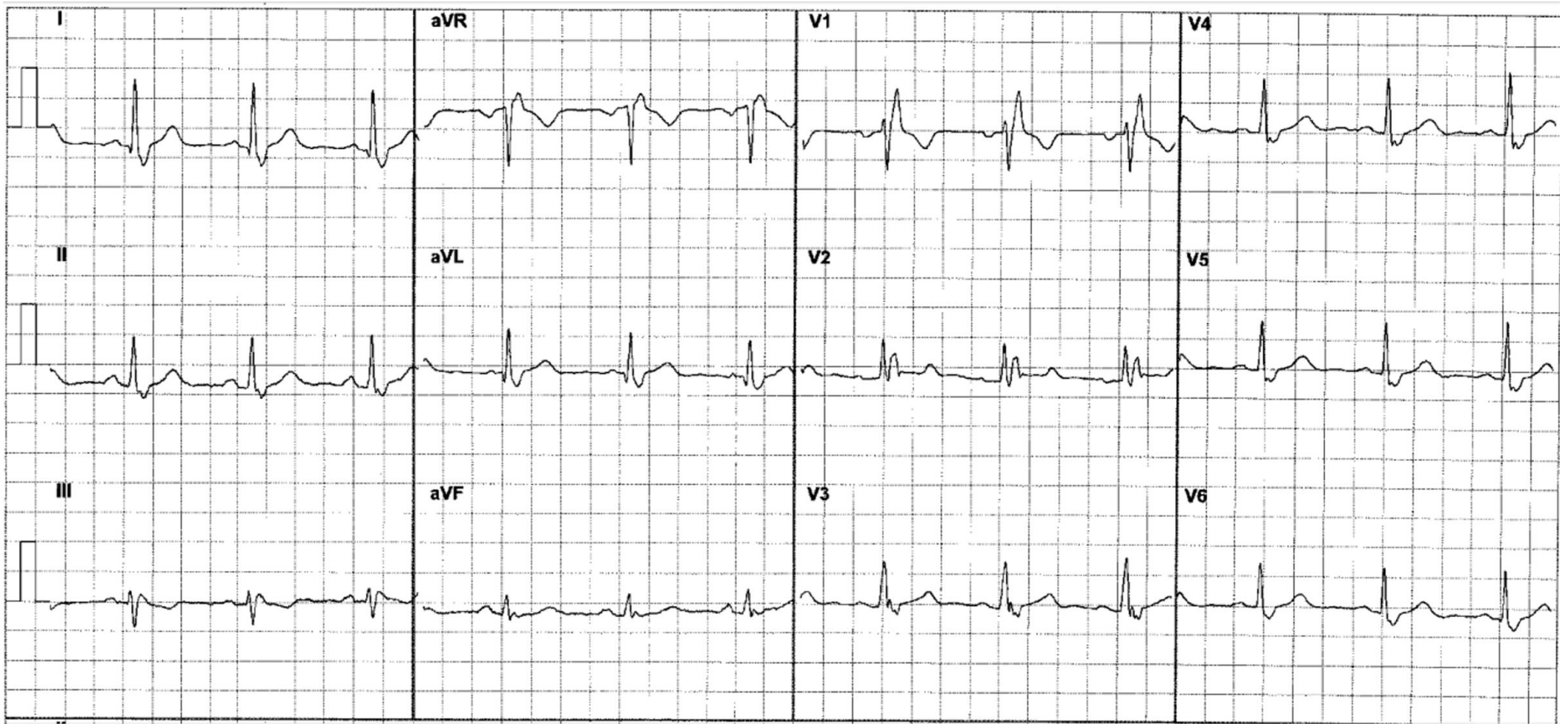
Disclosures

- None relevant to presentation

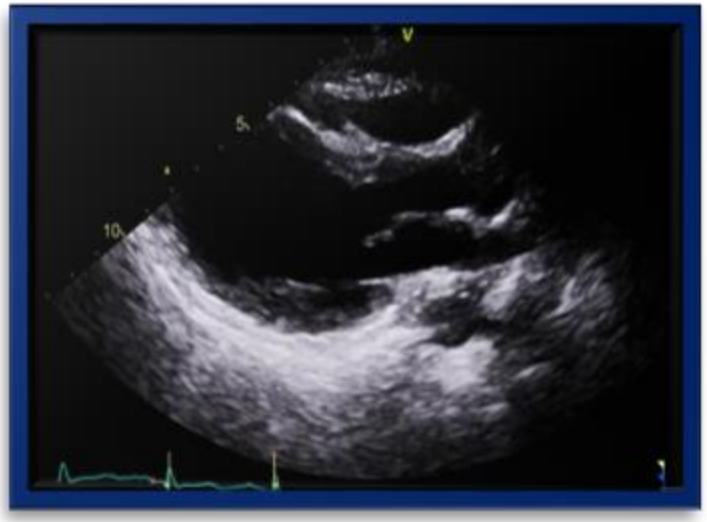
Case

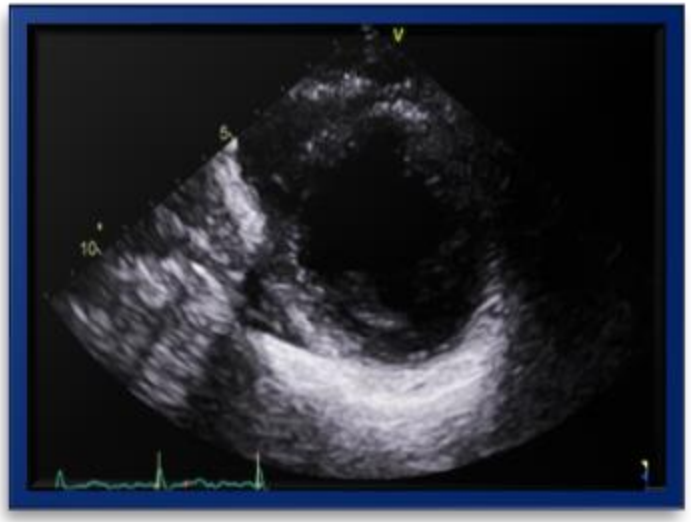
- 17-year-old female presented with out of hospital cardiac arrest and 20 minute down time
- B/G of Appendicectomy 1 week prior
  - Neuroendocrine tumour detected on Histo
- No regular medications
- Intubated and ventilated
- TnI 900->3100

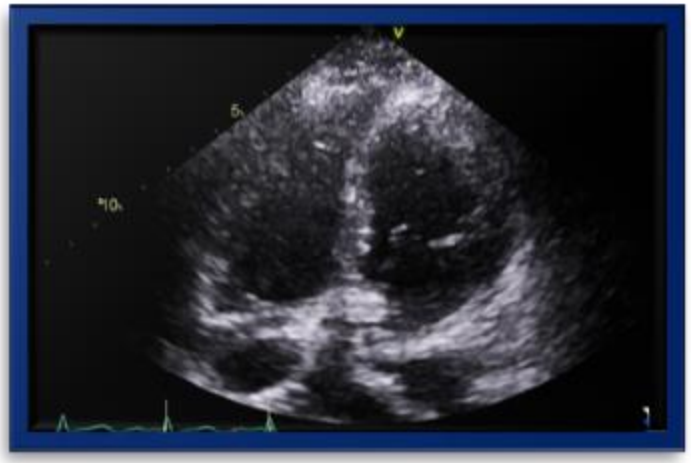
ECG

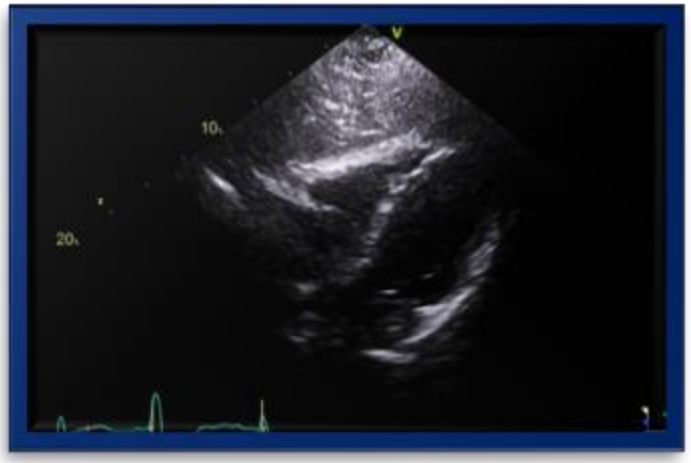


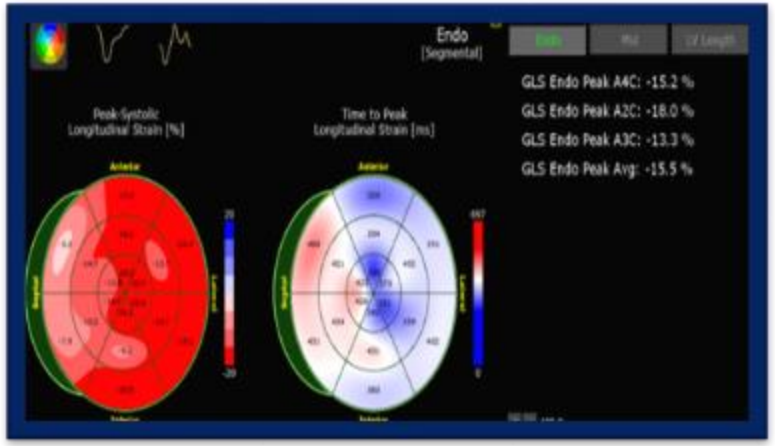
Echocardiogram



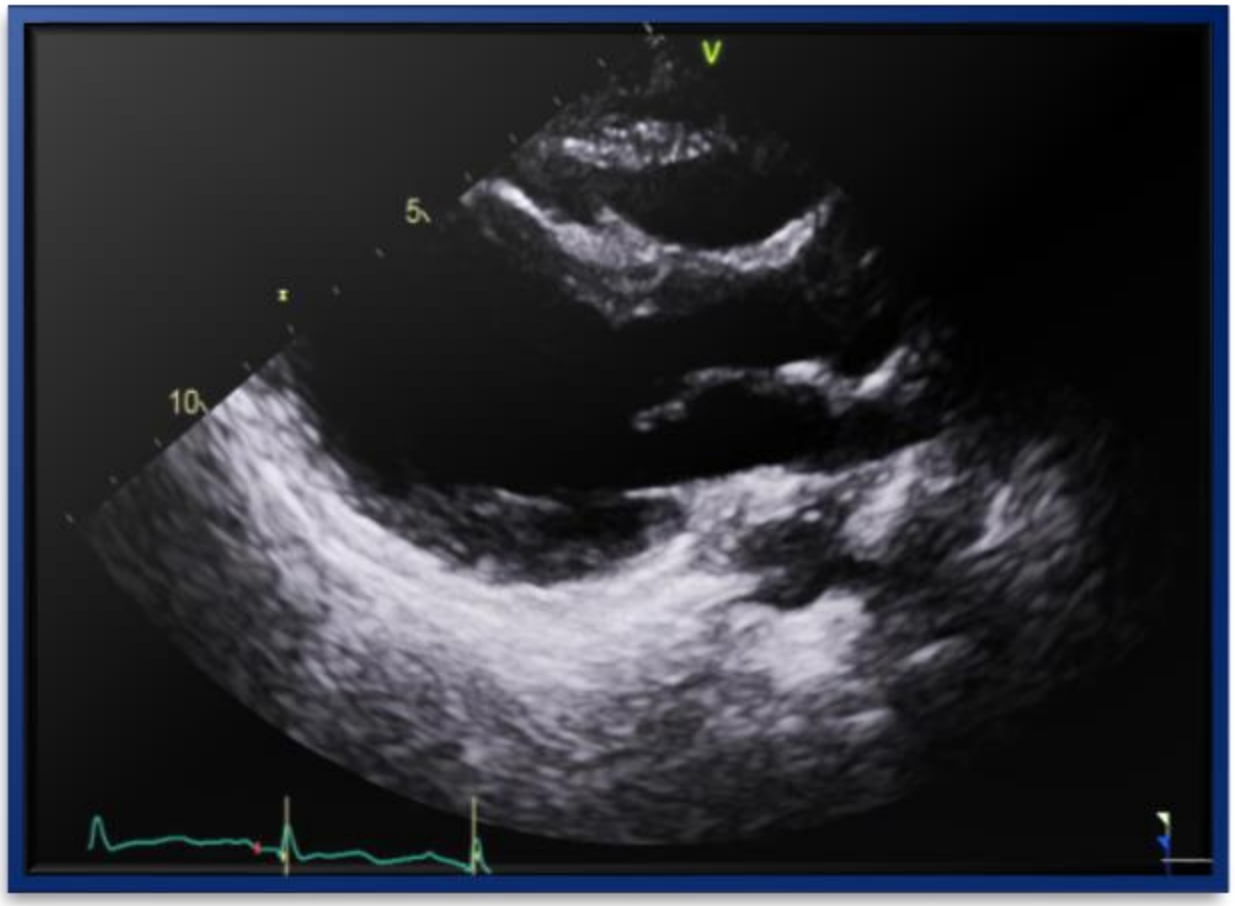


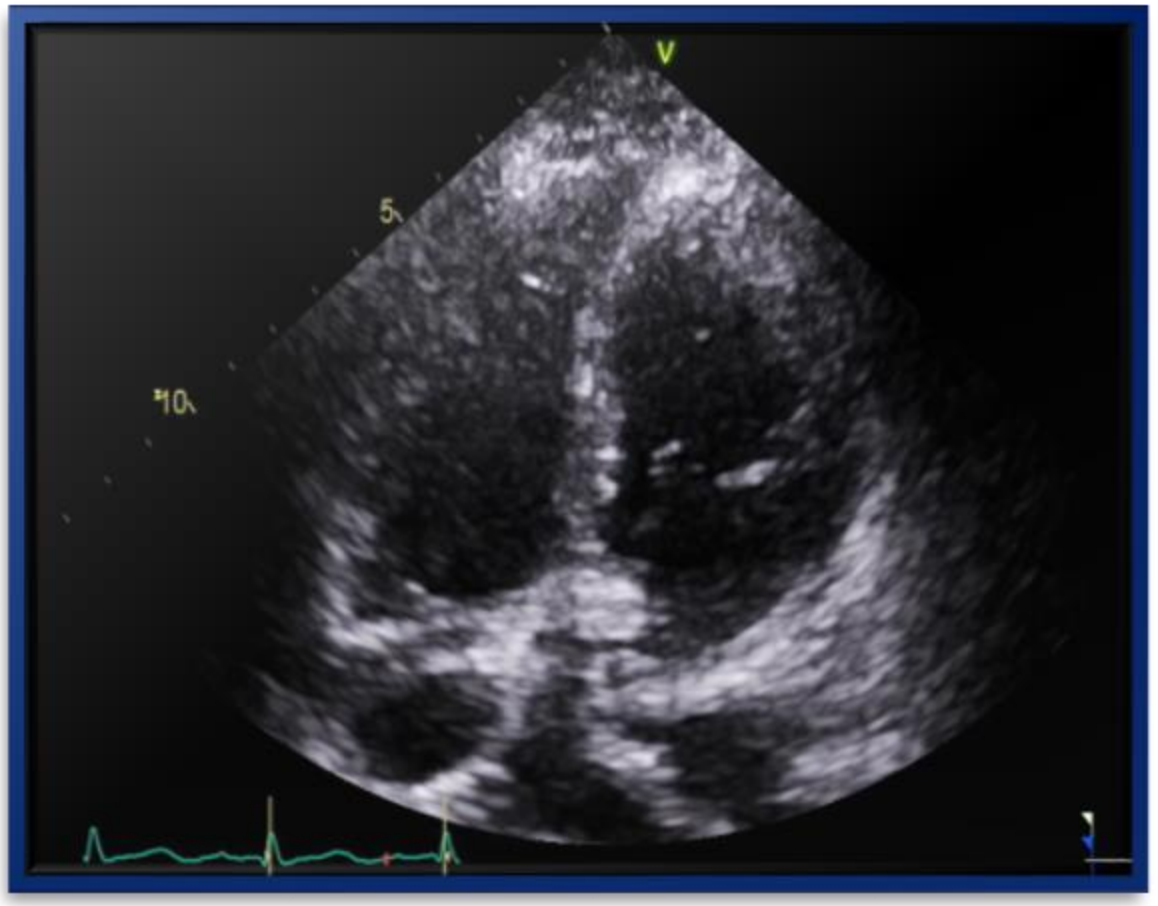




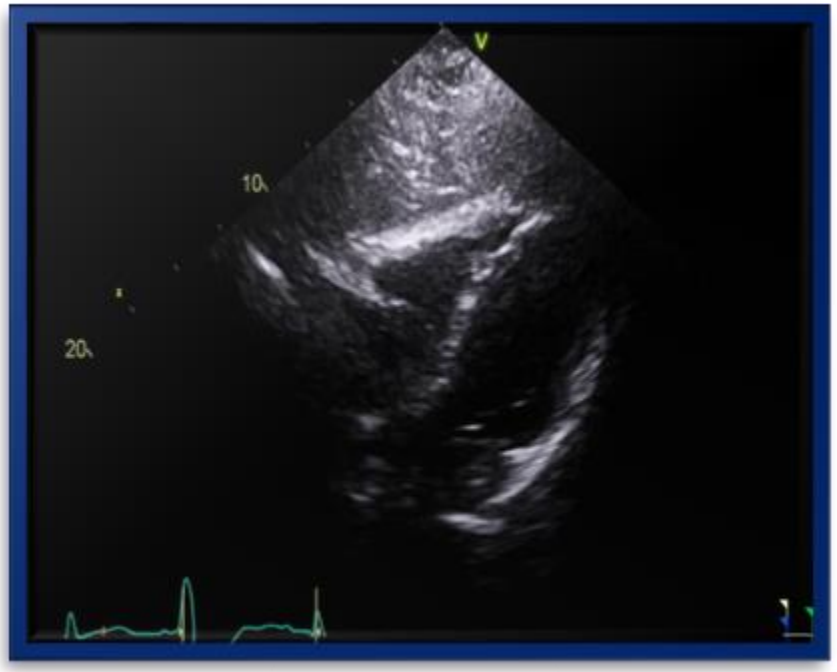


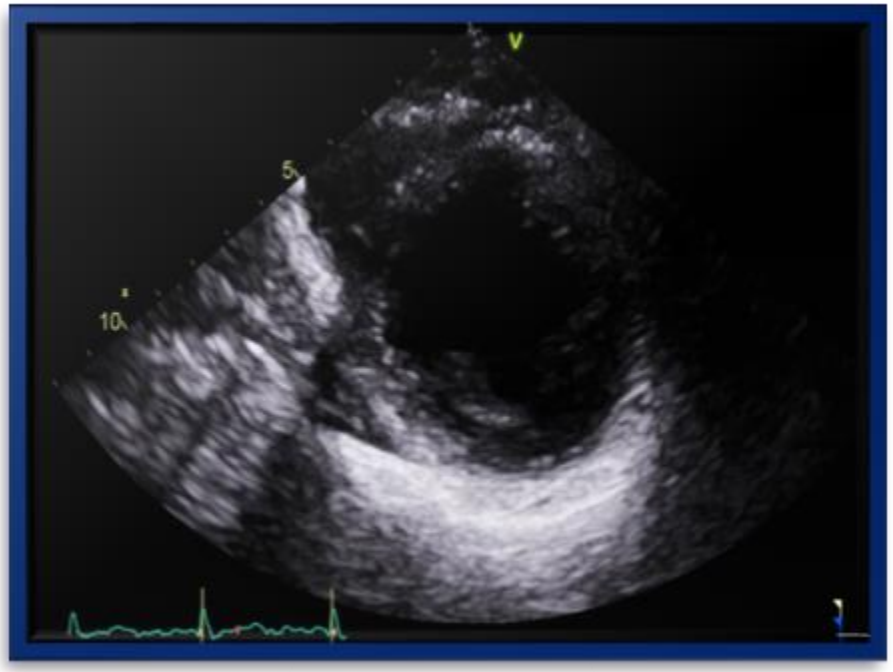
Echocardiogram

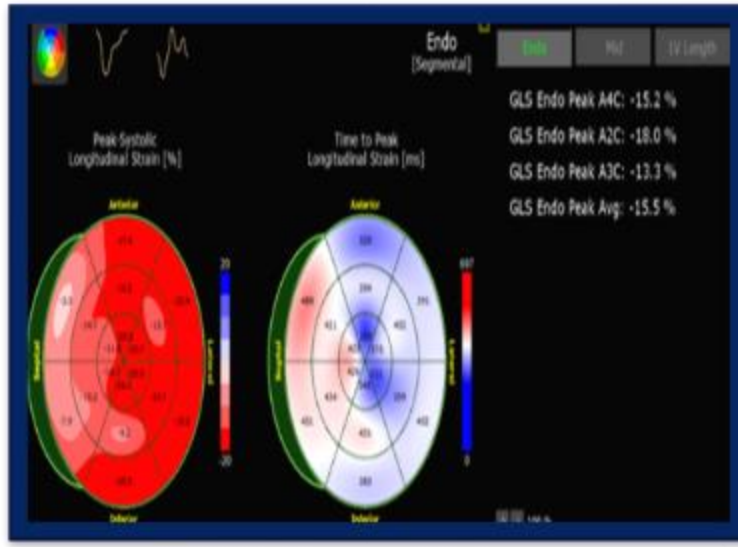




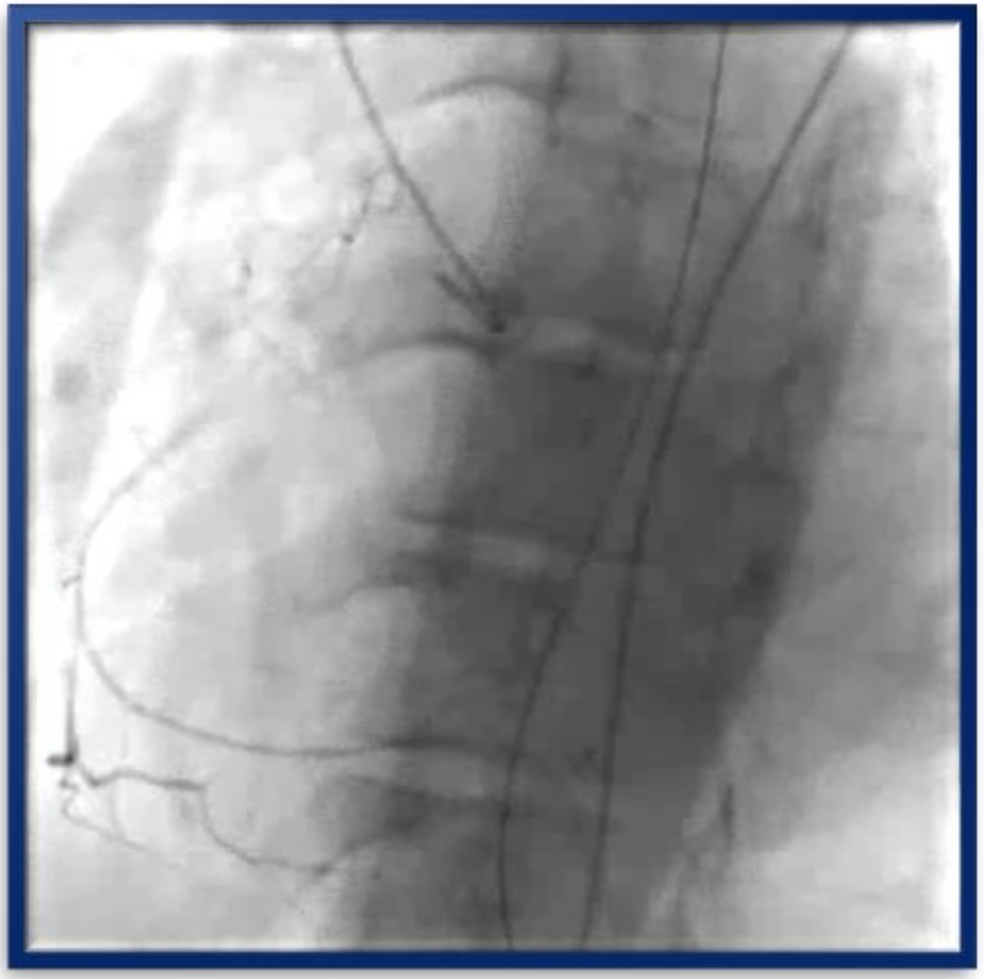
Echocardiogram



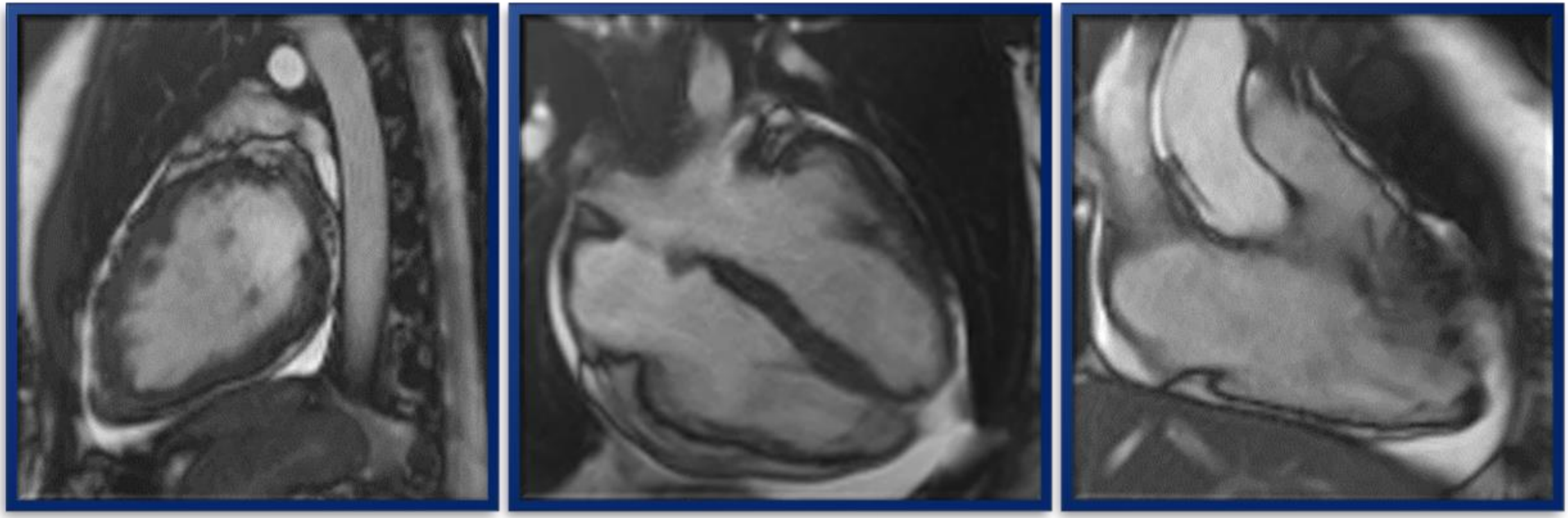




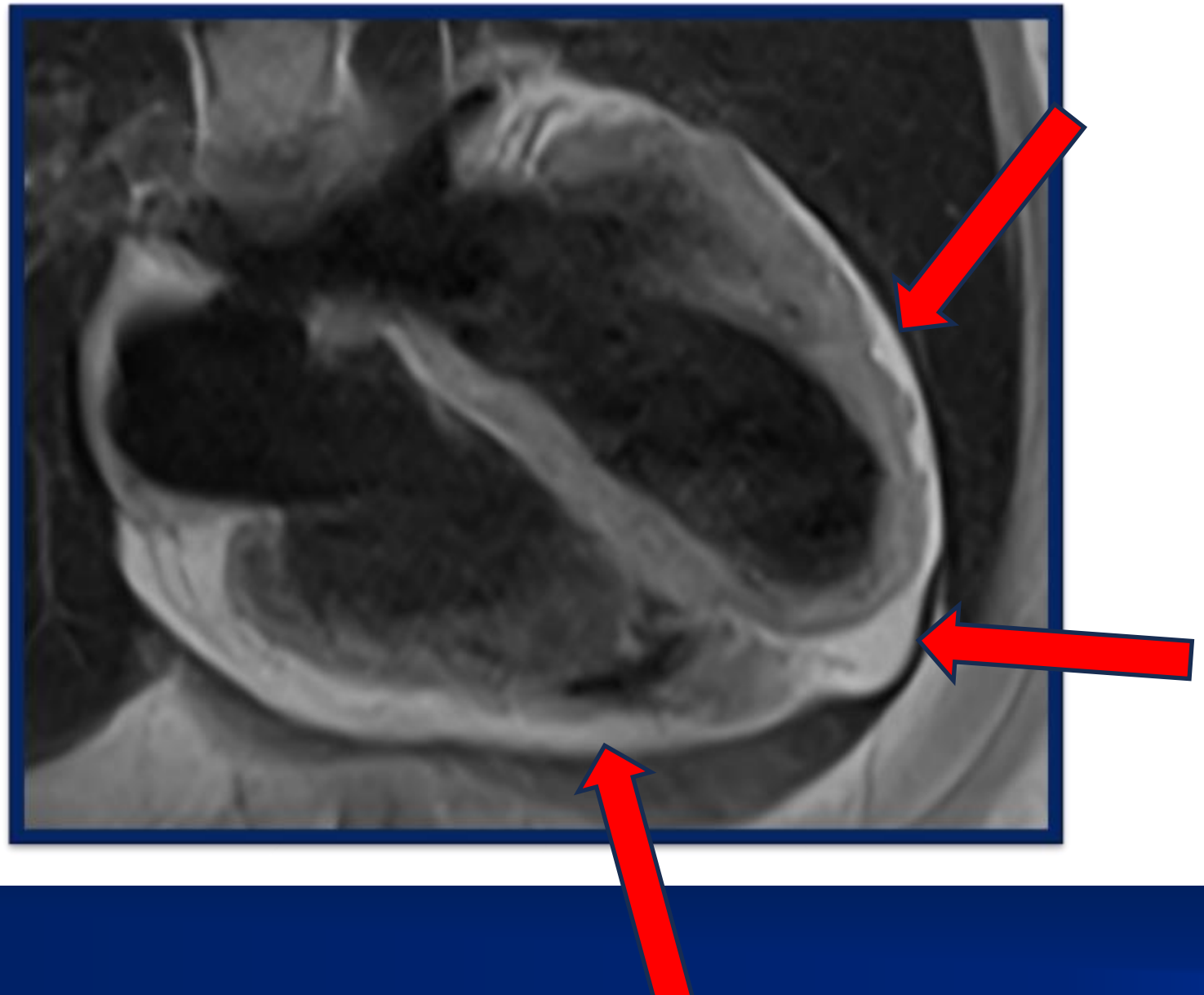
Coronary angiogram



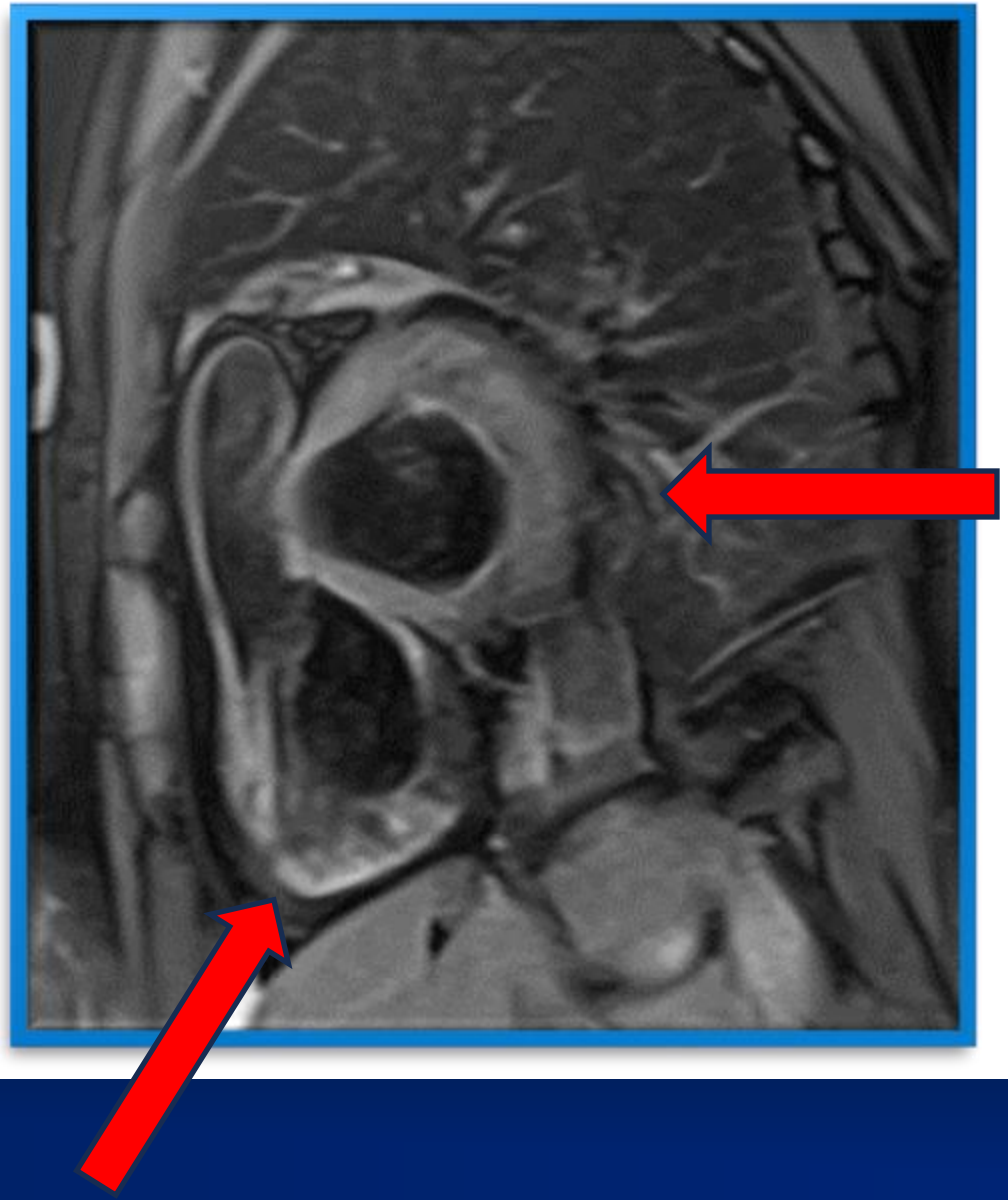
CMR - SSFP



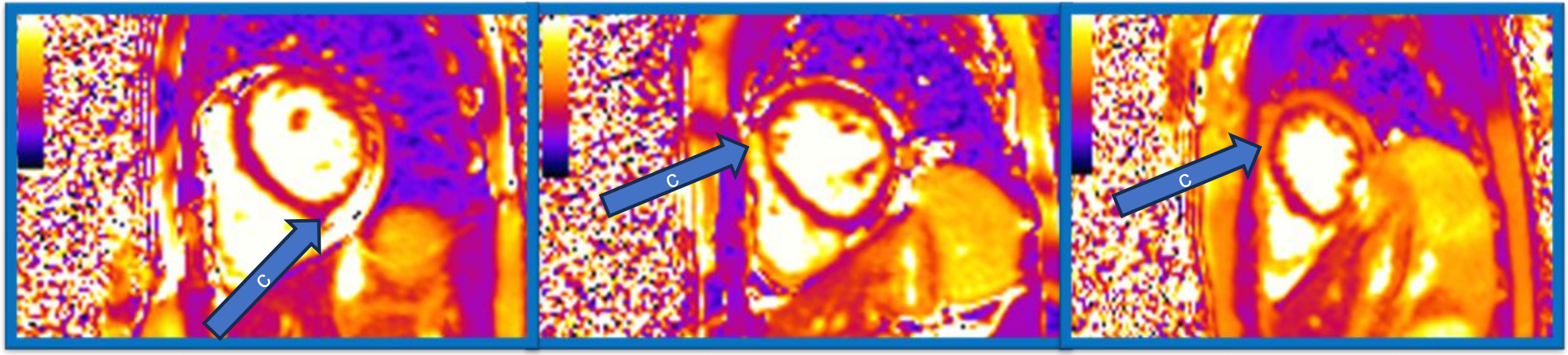
CMR - T1 HASTE Dark blood



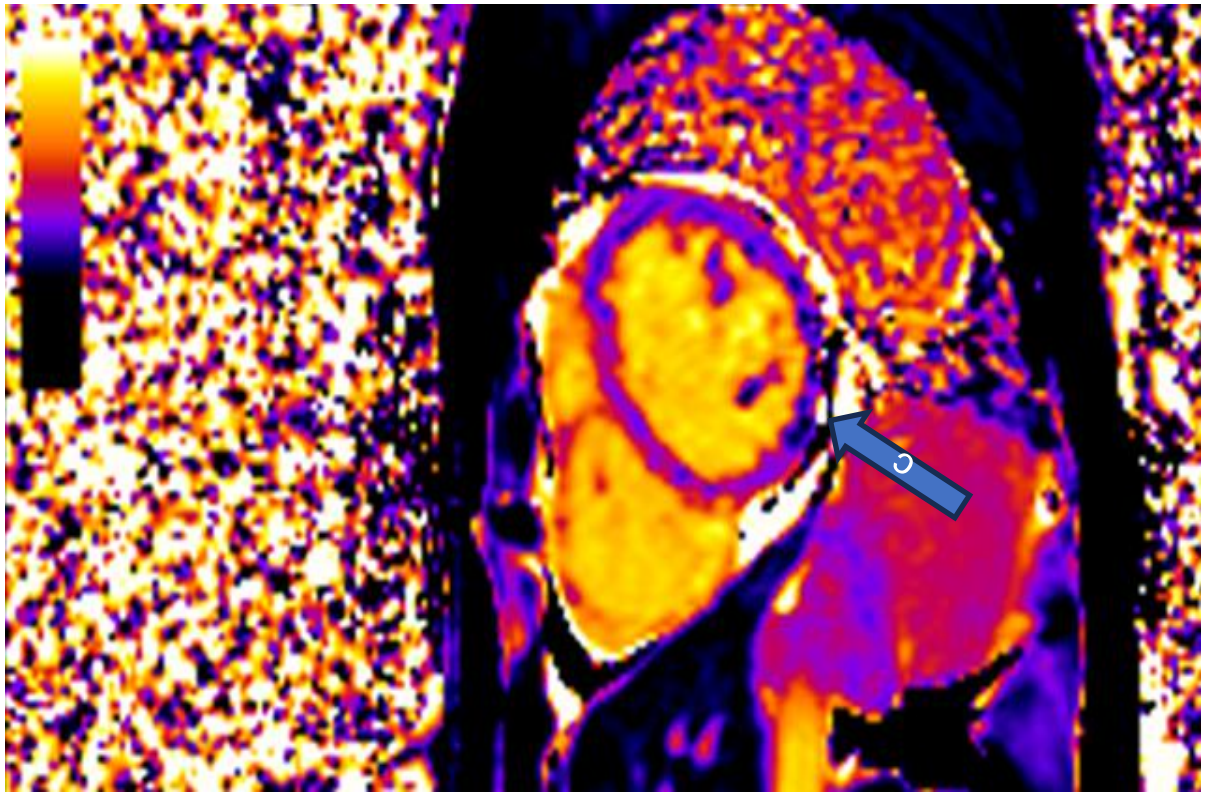
CMR - T2 SPAIR DB

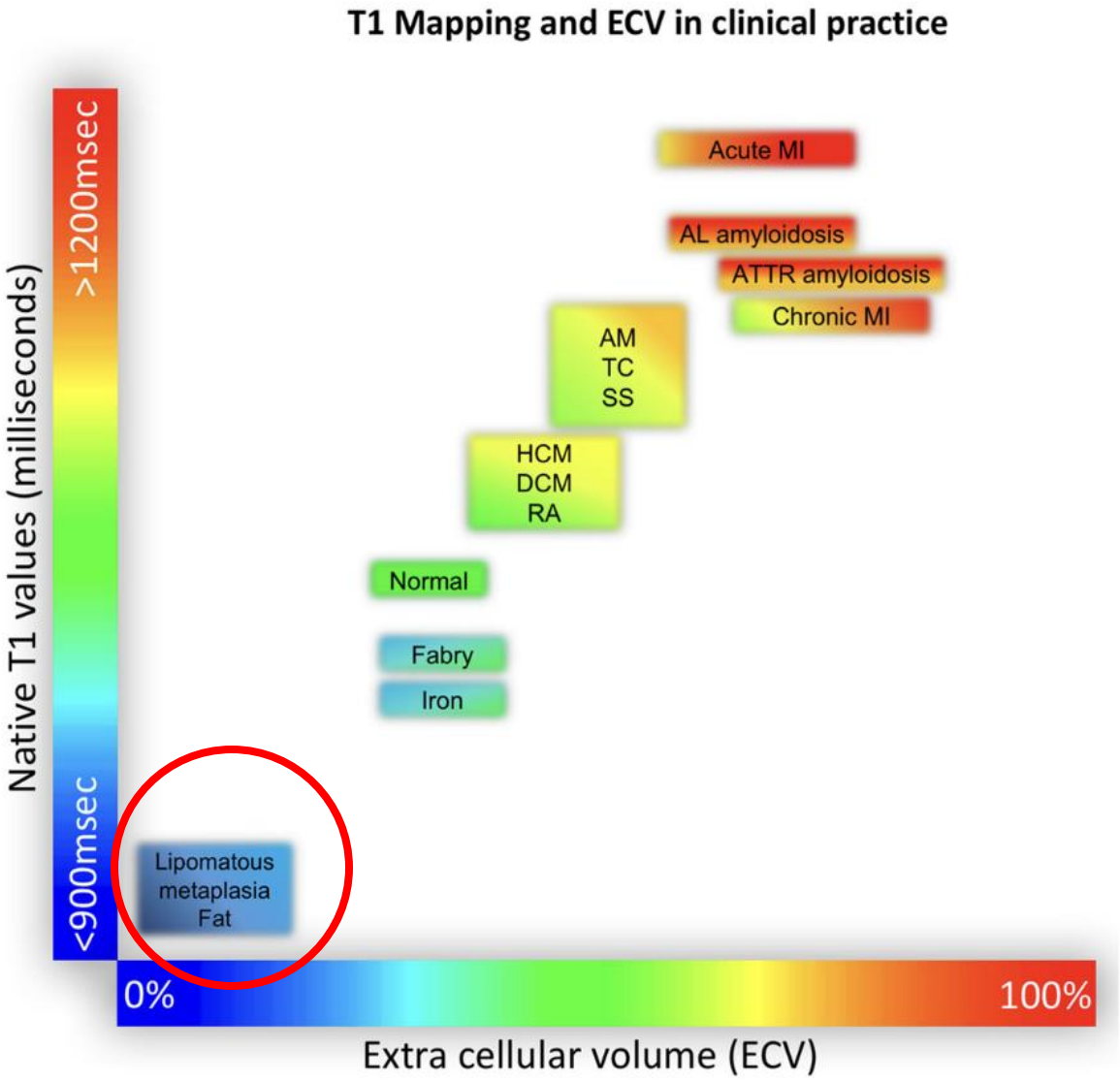


CMR T2 Maps



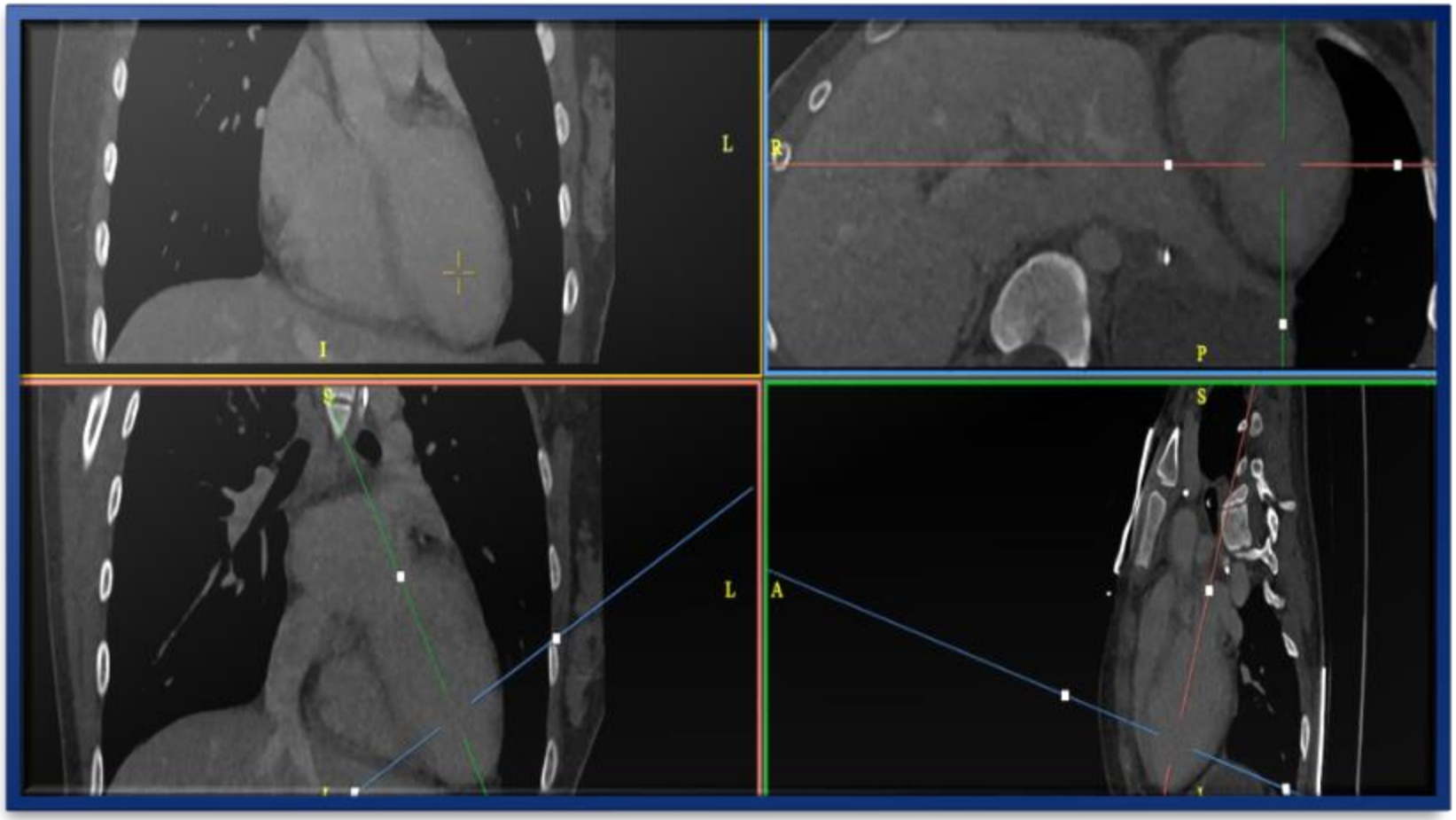
CMR – T1 Map



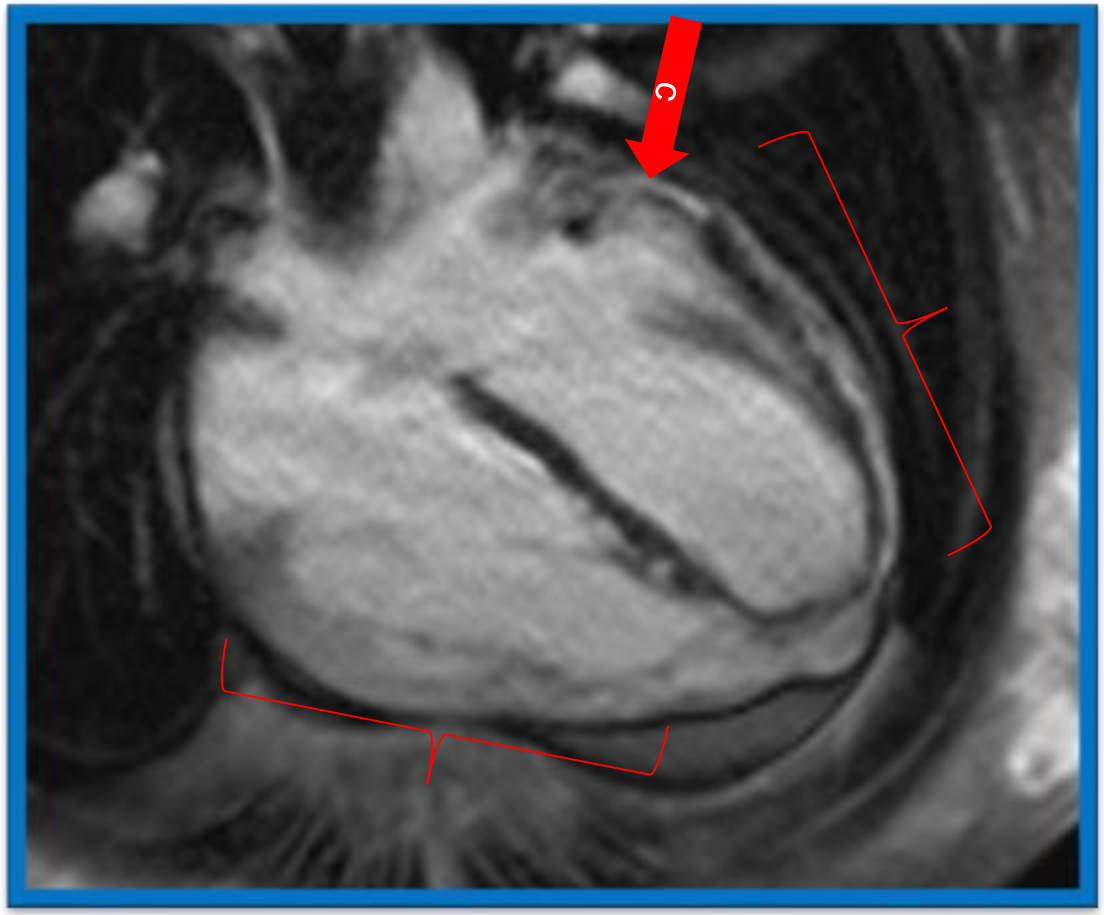


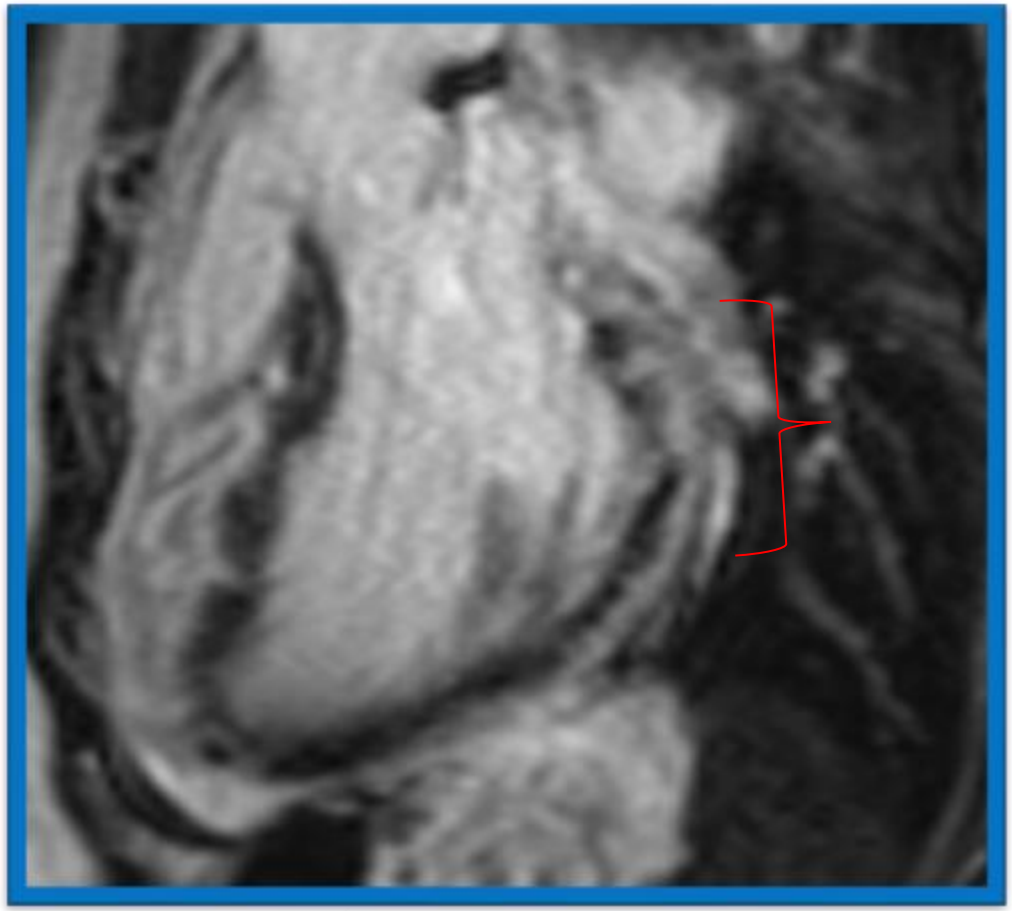
1. Messroghli et al. 2017 JCMR;19(75) DOI: https://doi.org/10.1186/s12968-017-0389-8

CT Chest/Abdo/Pelvis

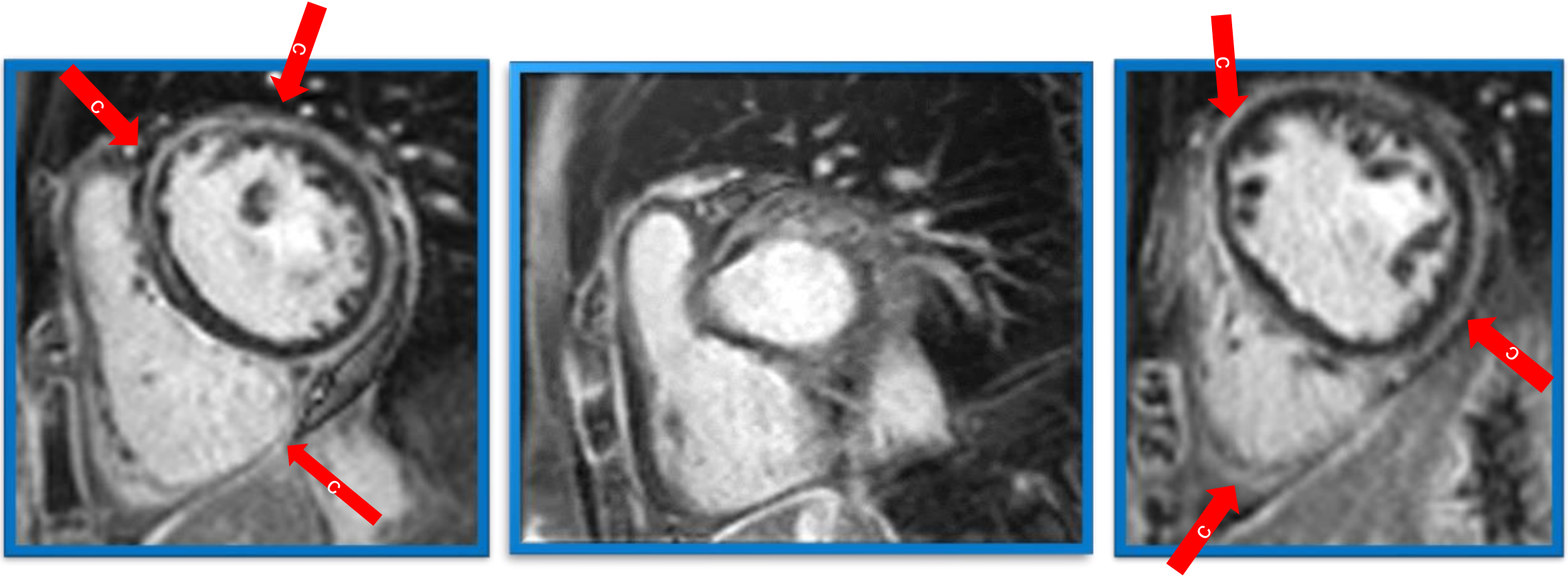


CMR - LGE

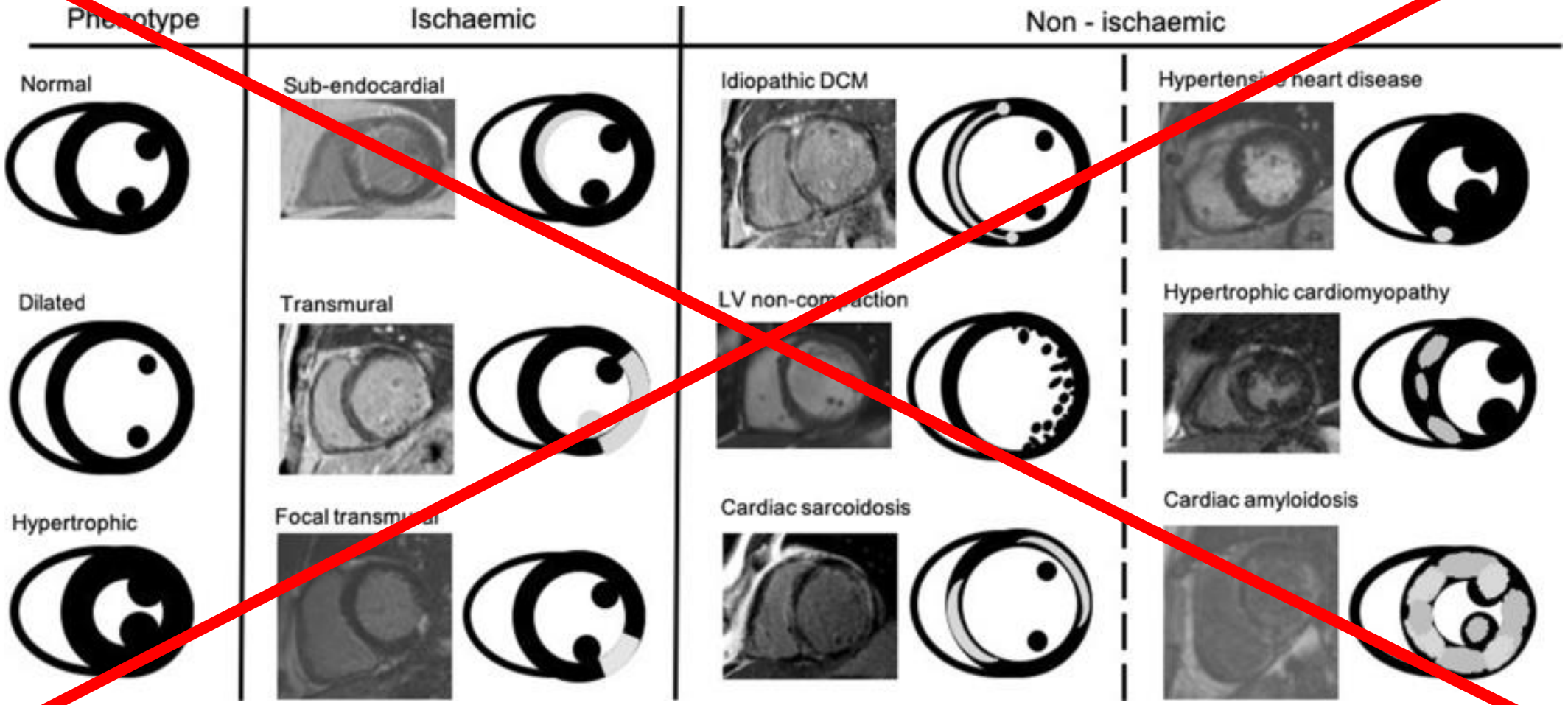




CMR - LGE



So finally, what are we working with?



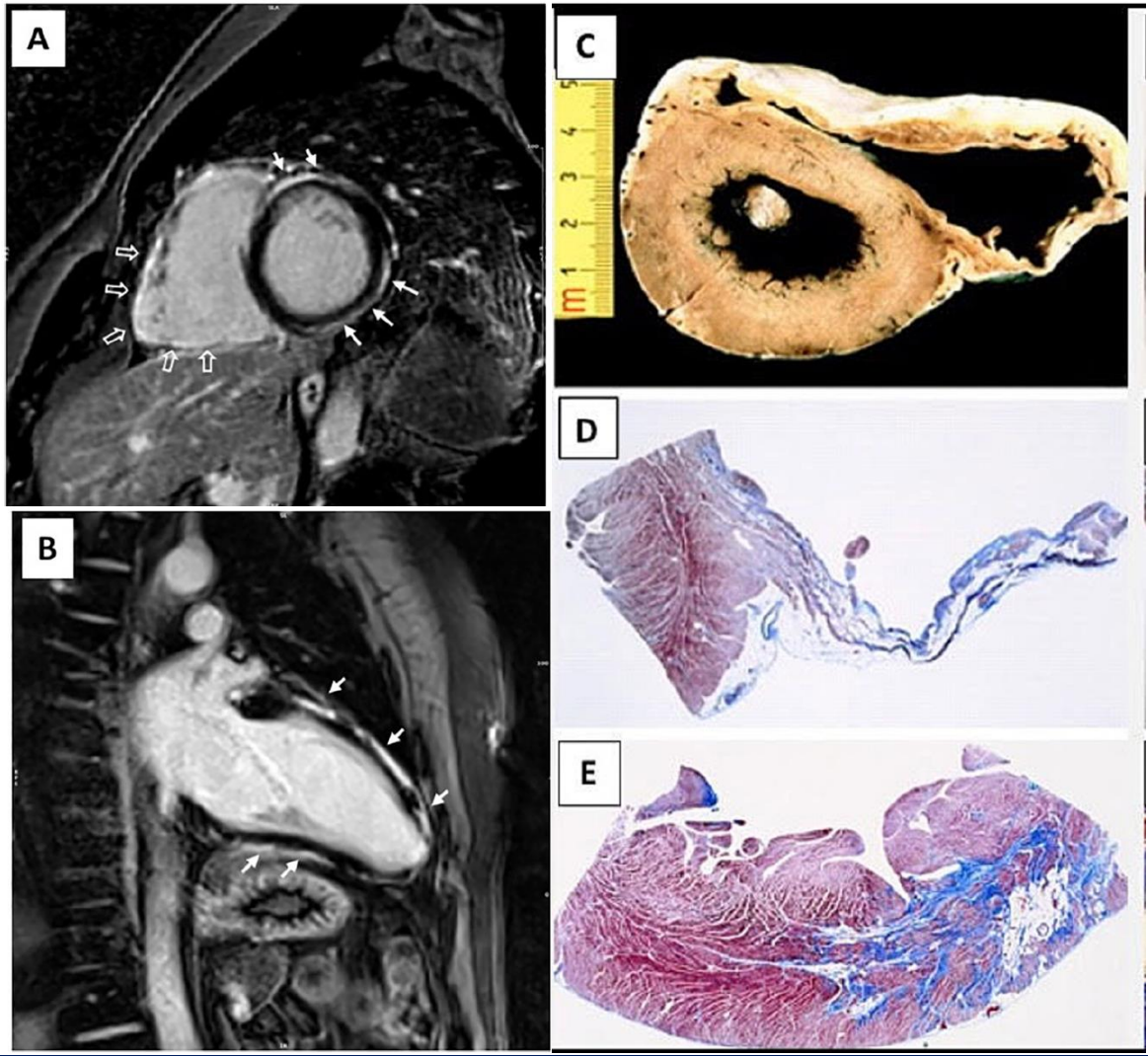
1. Berlot et al., Heart Failure Reviews 2020.

So finally, what are we working with?

- Berlot et al., Heart Failure Reviews 2020.

Arrhythmogenic Cardiomyopathy

- Our patient has ABCM (arrhythmogenic biventricular cardiomyopathy) presenting with likely “hot-phase”
- Large shift in approaching these patients from a multi-modality imaging perspective



Corrado et al. Int J Card 2024;395;131447

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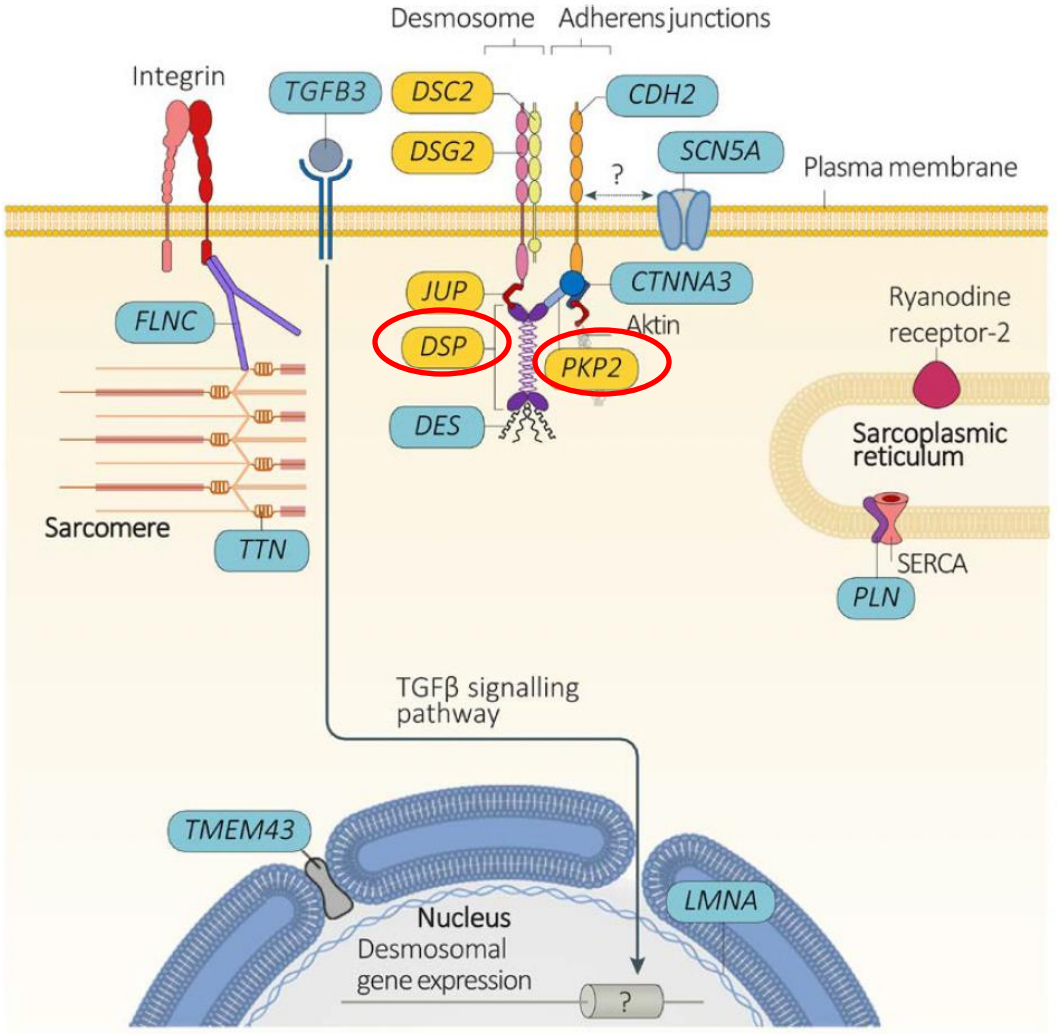
Hot-phase ACM

- An inflammatory, active stage of ACM, leading to severe arrhythmias and heart failure
  - Can commonly be mistaken for myocarditis
  - SCD is an extreme presentation of this phase
  - Elevated T2 (oedema/inflammation) in areas of LGE
- Strong genetic component with abnormalities in the DSP/PKP2 desmosomal genes
  - Commonly found in ACM



2024 Updated European Guidelines for ACM

Table 1

European Task Force criteria for diagnosis of Arrhythmogenic Cardiomyopathy.

Category	RV Phenotype	LV Phenotype
I. Morpho-functional ventricular abnormalities	***Major*** • Regional RV akinesia, dyskinesia, or aneurysm *plus* one of the following: • global RV dilatation (increase of RV EDV according to the imaging test specific nomograms for age, sex and BSA)* or • global RV systolic dysfunction (reduction of RV EF according to the imaging test specific nomograms for age and sex)* ***Minor*** • Regional RV akinesia, dyskinesia or aneurysm of RV free wall	***Minor*** • Global LV systolic dysfunction, with or without LV dilatation (increase of LV EDV according to the imaging test specific nomograms for age, sex, and BSA)*
II. Structural alterations	***Major*** • Fibrous replacement of the myocardium in ≥1 sample, with or without fatty tissue, at histology ***Minor*** • Unequivocal RV LGE (confirmed in 2 orthogonal views) in ≥1 RV region(s) (excluding tricuspid valve)	***Major*** • "Ring-like" LV LGE (subepicardial or midmyocardial stria pattern) of ≥3 segments (confirmed in 2 orthogonal views), ***Minor*** • LV LGE (subepicardial or midmyocardial stria pattern) of 1 or 2 Bull's Eye segment(s) (in 2 orthogonal views) of the free wall, septum, or both (excluding patchy, focal or septal junctional LGE**)

Back to our patient

- Arrhythmogenic biventricular cardiomyopathy presenting with “hot phase” characteristics
- ICD implanted
- 4 pillar heart failure therapy
- Extubated and discharged 10 days after admission with no significant deficits
- Genetic testing pending

Thank you

Questions?

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