MR vid myokardit och kardiomyopati

Fredensborg 2025-09-30, Jan Engvall

Region Östergötland

Översikt från guidelines mm

- Initial referens till presentation från NYC 2019
- Lake Louise kriterier 2009 och 2018
- Guideline från ESC 2025

- MR-signalen pga ödem och cellskada, gadolinium förstärker
- Vid misstänkt myokardit har ca 15% infarktskada/coronarsjuka

Myokardit

Consensus Criteria for the Diagnosis of Myocarditis

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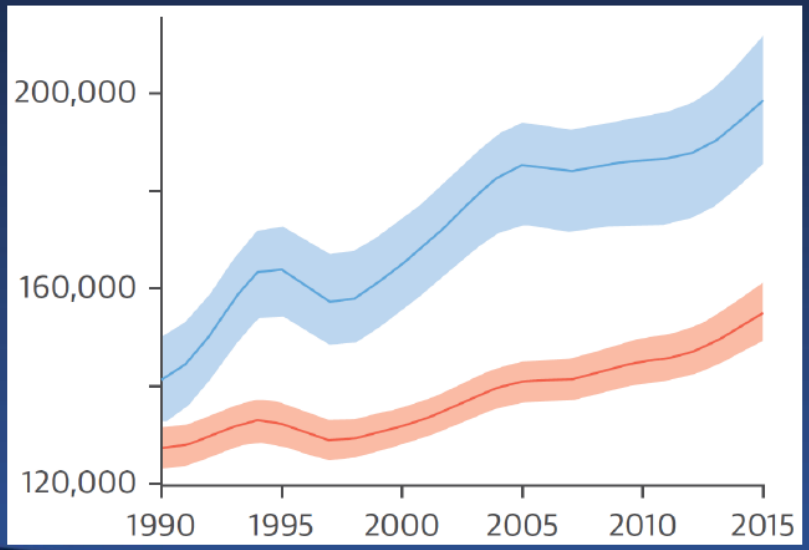
Prevalence, Etiology, Time Course of Myocarditis

Prevalence= 220/million/year
= 358 in Manhattan/year

Mortality of Myocarditis or Cardiomyopathy



Etiology
Virus, Trypanosoma cruzi (Chagas disease), Giant cell myocarditis, Autoimmune disease, Pheochromocytoma, Cardiac sarcoidosis, Hyperthyroidism

Time course in virus myocarditis

1. Acute (1-3 days) myocyte necrosis induced by virus replication
2. Subacute (few weeks-months) auto-immune myocarditis (activated T-lymphocytes) aimed to eliminate the virus in the myocardium
3. Chronic phase (DCM)

Heymans S. et al. JACC 2016;68:2348-64.

Sjukdomsmekanismer

Myocardial Inflammation

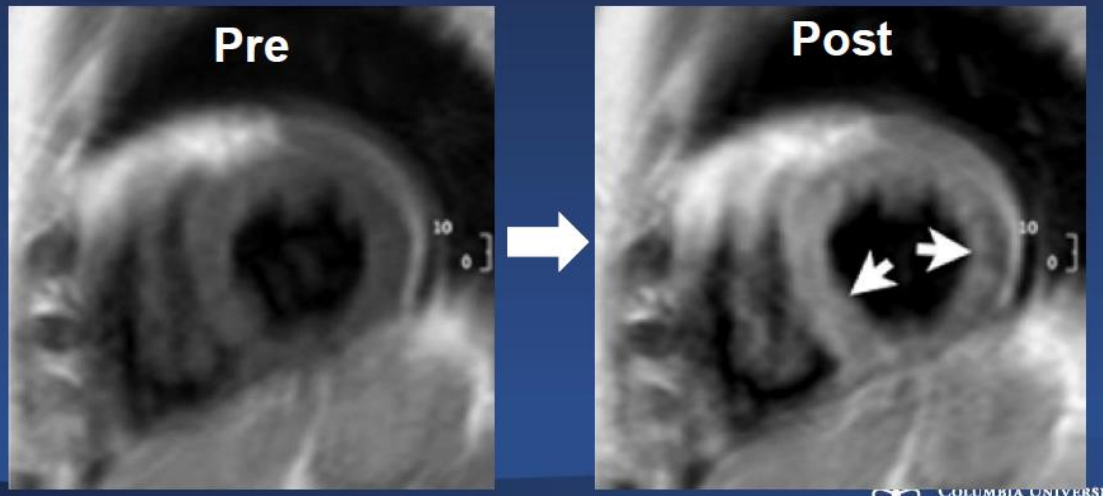
1. Dilatation of the myocardial vascular bed with hyperemia, increased vascular permeability or capillary leak, edema (intracellular, extracellular space) →**edema, hyperemia, increase of extracellular volume → T2 relaxation ↑ > T1 relaxation**
2. Myocyte injury with loss of cell membrane integrity, myocyte necrosis, accumulation of debris in the extracellular space, infiltration of inflammatory cells or macrophages →**myocardial damage, increase of extracellular volume → LGE (+)**
3. Collagen deposition with formation of interstitial fibrosis and scar →**increase of extracellular volume → LGE (+)**

Kriteriediagnos, Lake Louise 2009

Lake Louise Criteria

1. Regional or global signal intensity increase in T2
2. Increased myocardial early enhancement ration between myocardium and skeletal muscle in T1 with gadolinium.
3. ≥1 focal nonischemic LGE

Early enhancement:
A global SI enhancement ration of myocardium/skeletal muscle≥4.0 or absolute myocardial enhancement ≥45%



Friedrich MG. et al. JACC 2009;53:1475-87.

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Lake Louise är en skidort nära Banff och Calgary

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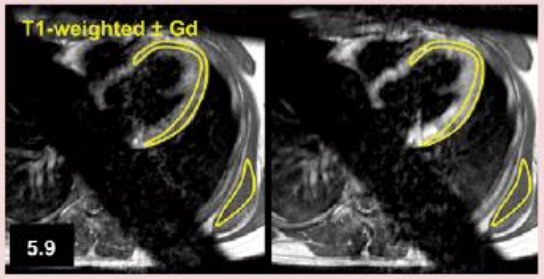
Reviderade Lake Louise 2018

Original Lake Louise criteria

T2-weighted

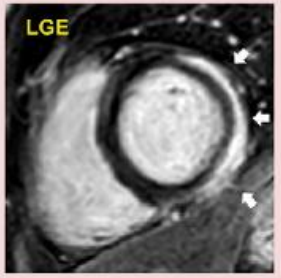
Regional high T2 SI or high T2 SI ratio

Early gadolinium enhancement



Increased early gadolinium enhancement ratio

LGE

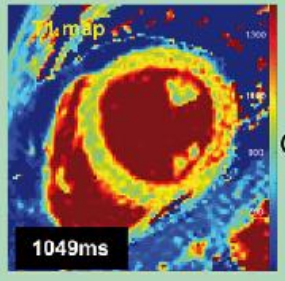


Positive nonischemic LGE

(any 2 out of 3)

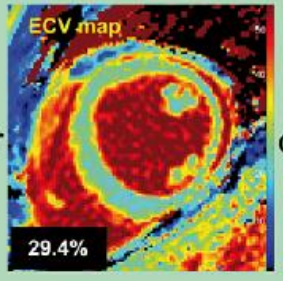
2018 Lake Louise criteria

T1-based imaging



Increase of native T1

or

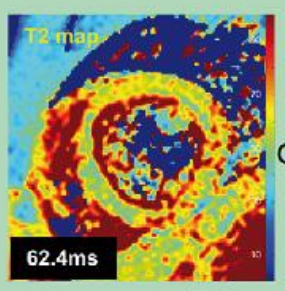


Increase of ECV

or

Positive nonischemic LGE

T2-based imaging



Increase of T2

or

Regional high T2 SI or high T2 SI ratio

(2 out of 2)

Inflammation med CMR

CMR Criteria of Myocardial Inflammation

Diagnostic Target		Original Lake Louise Criteria (Any 2 out of 3)	Updated Lake Louise Criteria (Any 2 out of 2)
Myocardial Edema		T2-weighted imaging Signal intensity↑	T2-weighted imaging Signal intensity↑ Relaxation time↑
Myocardial Injury	Hyperemia (Intra/extra cellular edema, capillary leak)	Early Gadolinium Enhancement	T1-weighted imaging Native (non-contrast) relaxation time ↑ Extracellular volume↑ Non-ischemic pattern LGE
	Myocardial Necrosis, scar	Late Gadolinium Enhancement	
Supportive Criteria		Pericardial effusion Systolic LV wall motion abnormality	Pericardial effusion High signal intensity of pericardium in LGE, T1, T2 mapping Systolic LV wall motion abnormality

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Ferreira VM. et al. JACC 2018;72:3158-76.

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Antal pt i studier av Gd, T1 och T2

Diagnostic Performance Of CMR Criteria for Acute Myocarditis

Ferreira VM. et al. JACC 2018;72:3158-76.

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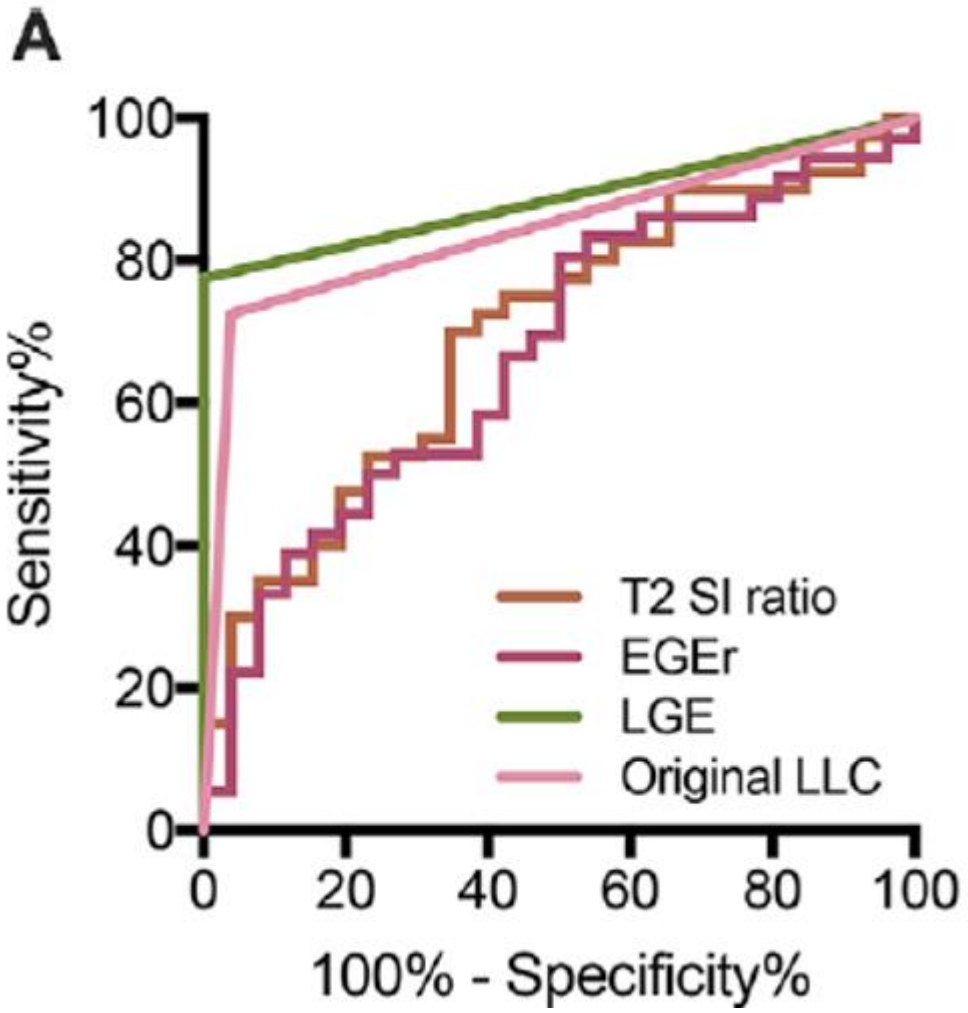
Vad är Area-Under-Curve, AUC?



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Vad säger guidelines?

- 2025 ESC likställer hjärt-MR med biopsi, för diagnos och 6 mo F/U

Guideline Recommendation

- **2016 ESC for the acute and chronic heart failure**
  - **CMR is recommendation for the characterization of myocardial tissue in case of suspected myocarditis, amyloidosis, sarcoidosis, Chagas disease, Fabry disease, non-compaction cardiomyopathy, and haemochromatosis. (Class I, Level C Evidence).**
- **2016 AHA Scientific Statement for specific DCM**
  - **CMR is reasonable for the diagnosis of myocarditis in clinically stable patients with clinically suspected myocarditis (Moderate level consensus, Level C Evidence).**
- **2013 ESC Myocardial and Pericardial Disease**
  - **Reasonable to perform CMR in clinically stable patients prior to biopsy.**

Caforio AL, et al. EHJ 2013;34:2636-48. Bozkurt B, et al. Circulation 2016; 134:e579-646. Ponikowski P, et al. EHJ 2016;37:2129-200.

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2025 ESC Guidelines for the management of myocarditis and pericarditis

Ska alla patienter med akut troponinläckage utredas med inneliggande hjärt-MR?

15.4. Prognosis and outcomes

The prognosis of IMPS varies. Most patients presenting with chest pain have a favourable outcome, although recurrences, especially for pericarditis, may severely affect the quality of life of patients and require long-term follow-up. For complicated cases of myocarditis with HF and arrhythmias, a tailored and individualized approach is warranted. Such patients require long-term follow-up, usually lifetime monitoring.

Nevertheless, a specific challenge for myocarditis is related to the low frequency of complications and the high rate of spontaneous remission. On this basis, clinical trials on medical therapy for myocarditis might be currently underpowered to assess the clinical benefit of a spe-

Recommendation Table 3 — Recommendations for the use of cardiovascular magnetic resonance imaging (see Evidence Table 3)

Recommendations	Class[a]	Level[b]
Myocarditis		
CMR is recommended in patients with suspected myocarditis to reach a clinical diagnosis and to determine the cause of acute myocardial injury, including assessment of oedema, ischaemia, and necrosis/fibrosis/scarring.[115,164,169–183]	I	B
CMR is recommended for follow-up at least within the first 6 months in patients with myocarditis to identify a healed or ongoing process, for risk stratification and personalized therapy, and to enable a return to exercise.[10,62,184–186]	I	C
Pericarditis		
CMR is recommended in patients with suspected pericarditis when a diagnosis cannot be made using clinical criteria to assess evidence of pericardial thickening, oedema, LGE, and to assess the persistence of disease during follow-up in selected cases.[110,129,165,187–194]	I	B

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Fynd vid misstanke om myokardit

MyoRacer-Trial

129 pts with suspicious myocarditis with biventricular biopsy, CMR (1.5T, 3T)

	Acute Group (<2 weeks)	Chronic Group (≥2 weeks)
Age, yrs	40 (29, 56)	46 (35, 57)
Days since symptom onset	6 (1, 13)	30 (21, 138)
Symptoms, chest pain	64%	41%
ECG T-wave changes	44%	49%
Troponin positive	77%	75%
Biopsy findings		
Myocardial inflammation	70%	71%
Presence of virus genome	26%	28%

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Lurz P. et al. JACC 2016;67:1800-11.

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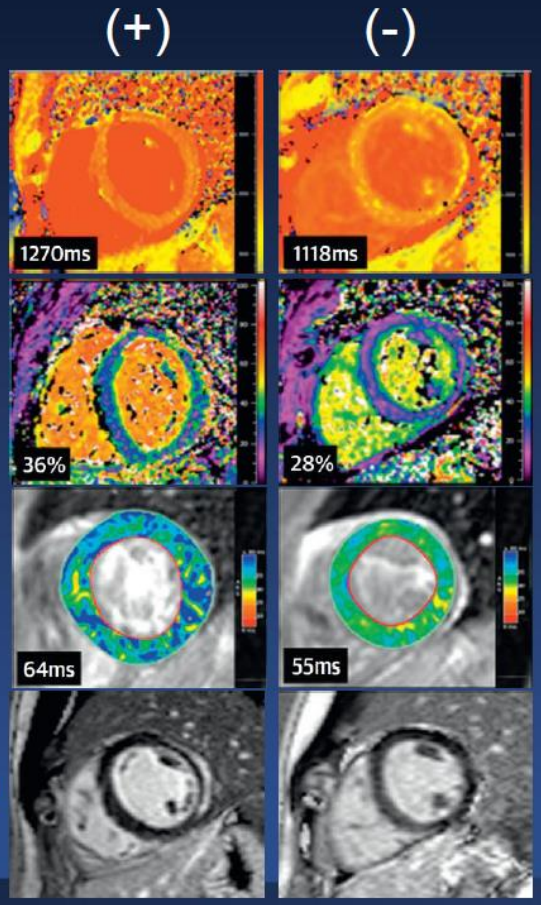
Fynd i CMR

Acute Myocarditis



Lurz P. et al. JACC 2016;67:1800-11.

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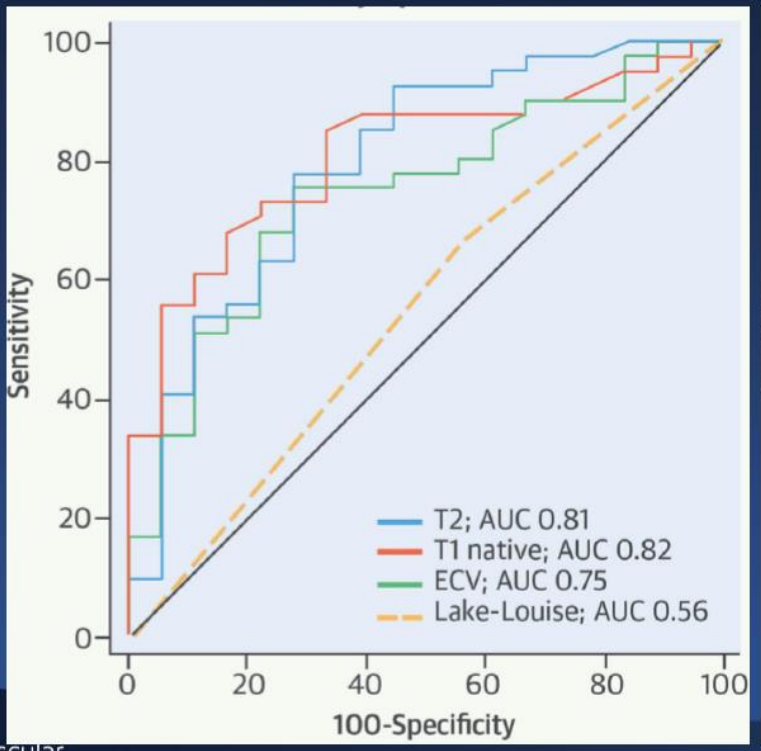
ROC-analys av klinisk studie

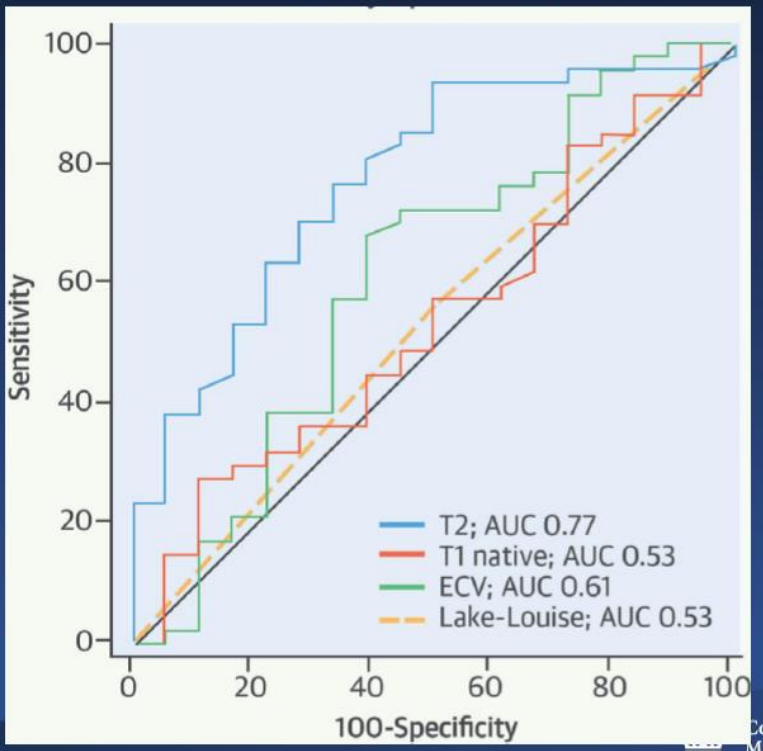
MyoRacer-Trial, Diagnostic Accuracy

1.5T vs 3T (n=111) shows similar diagnostic accuracy.

Acute Symptoms (n=61)

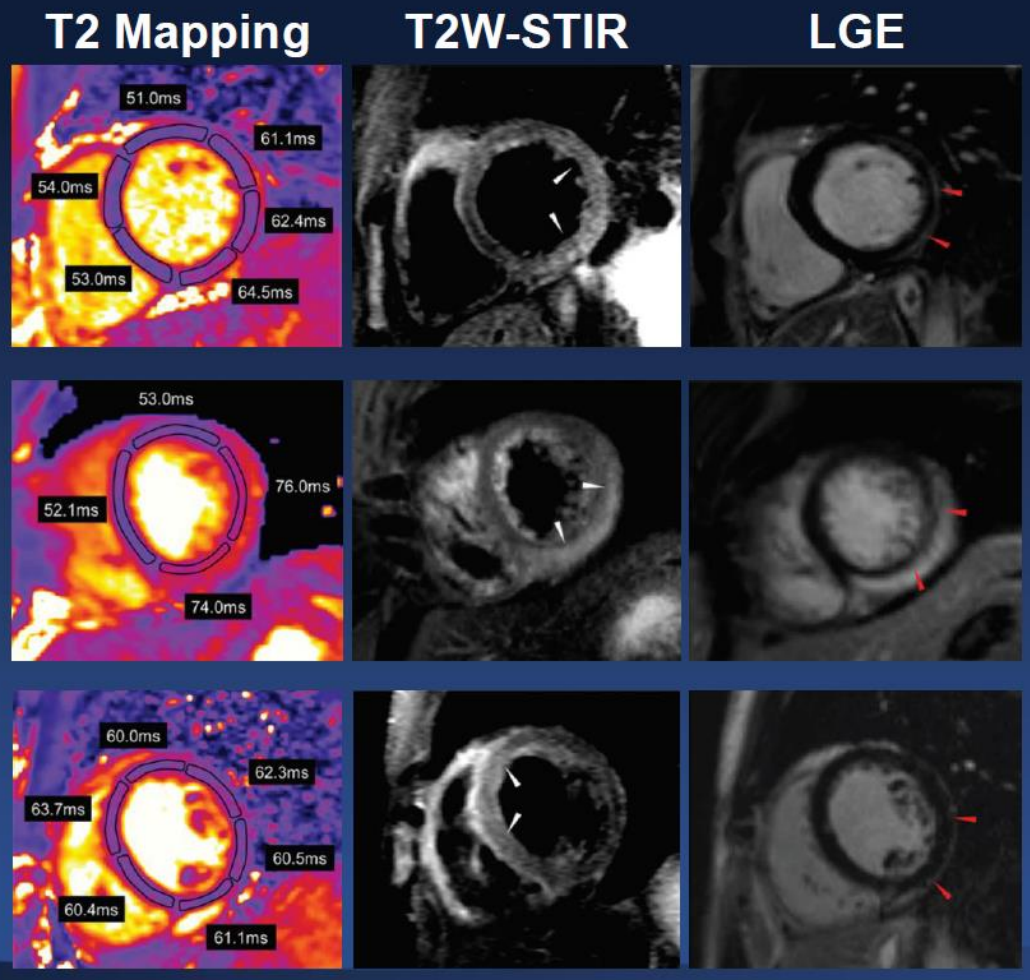


Chronic Symptoms (n=61)



Lurz P. et al. JACC 2016;67:1800-11.

T2 map – STIR - LGE



Thavendiranathan P. et al. Circ Img 2012;5:102-110.

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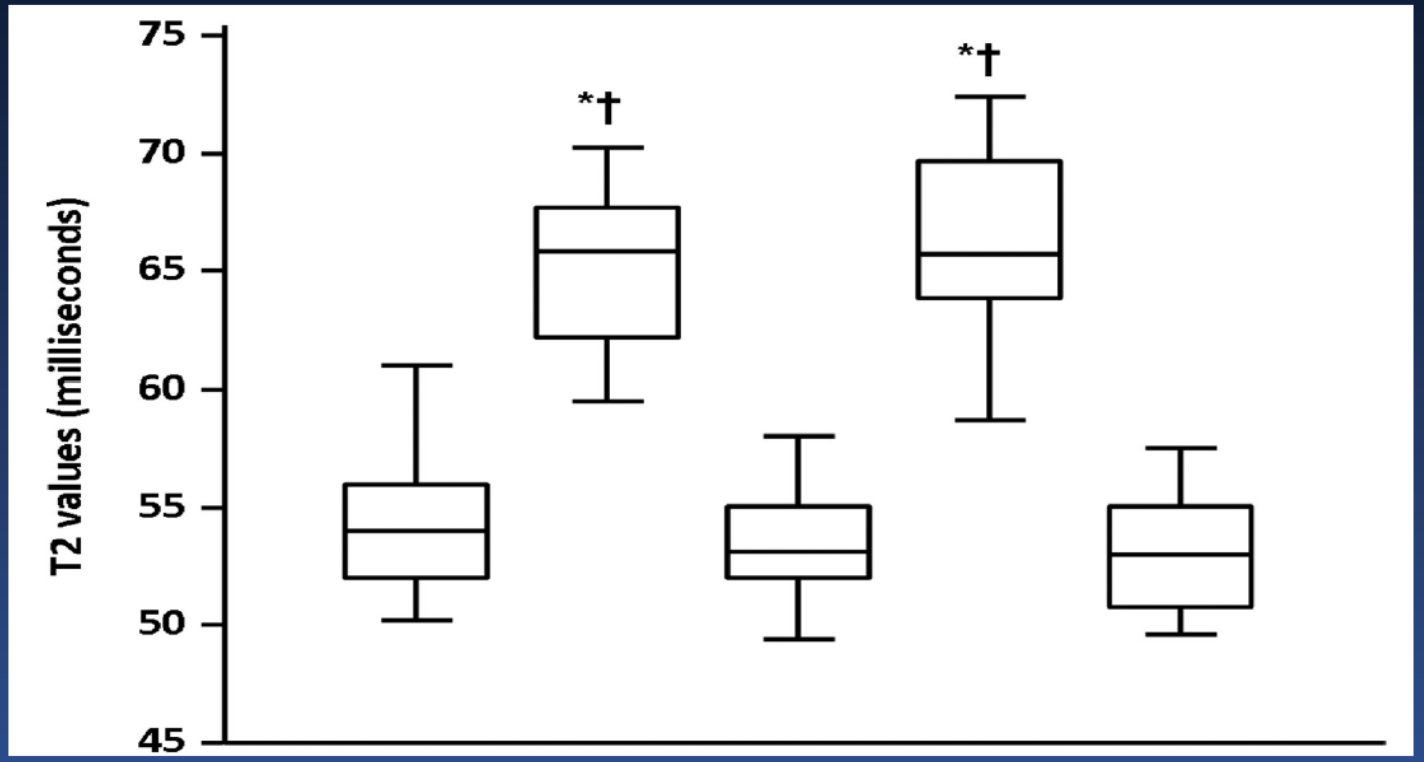
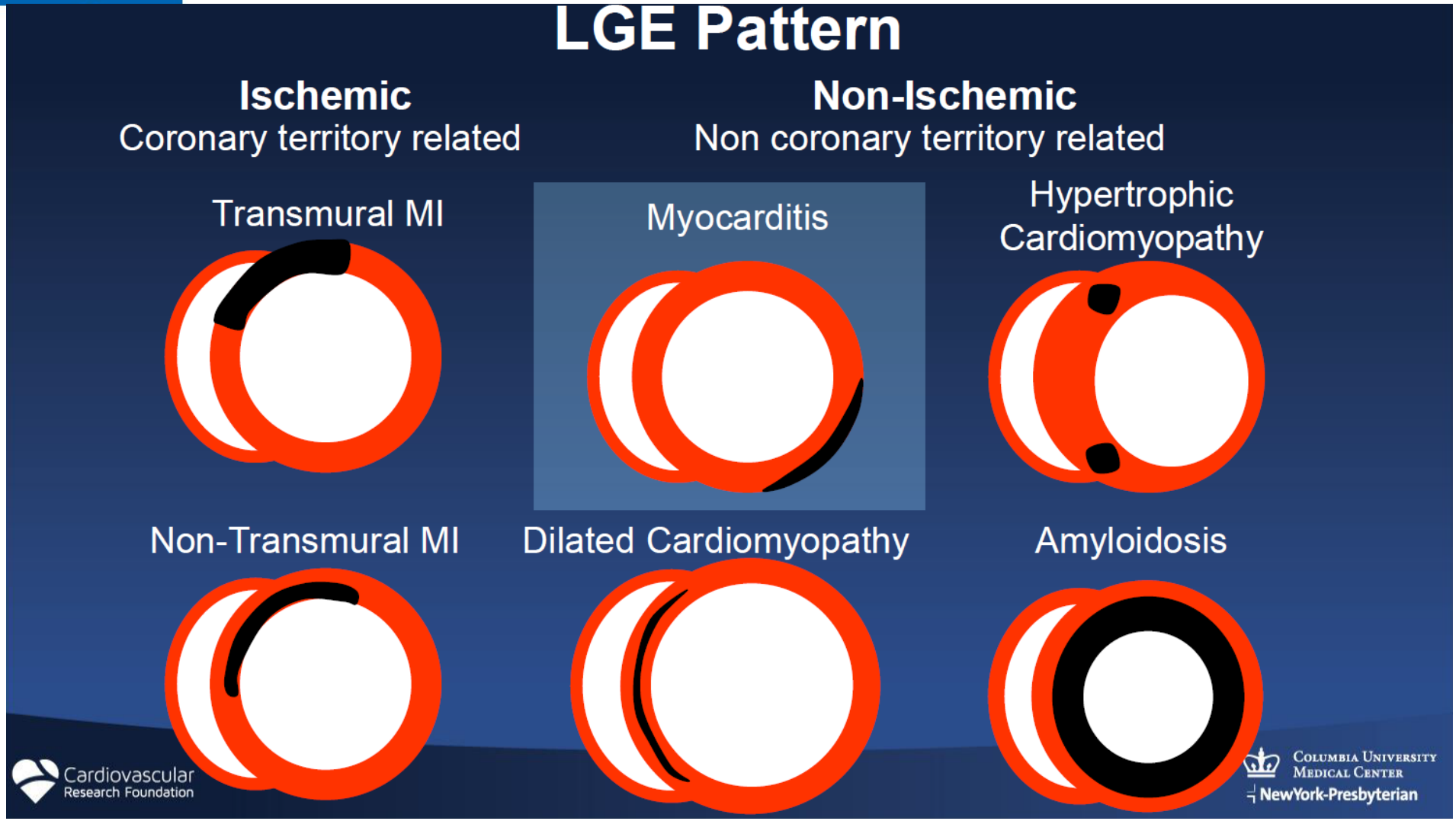
T2 vid myokardit och Takotsubo



Thavendiranathan P. et al. Circ Img 2012;5:102-110.

LGE mönster


LGE Pattern
Ischemic
Coronary territory related
Non-Ischemic
Non coronary territory related
Transmural MI
Myocarditis
Hypertrophic Cardiomyopathy
Non-Transmural MI
Dilated Cardiomyopathy
Amyloidosis
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Sens och spec beror av prevalens

Summary

1. New Lake Louise Criteria for myocarditis includes 1) edema by T2 and 2) myocardial injury by T1 image.
2. Diagnostic accuracy (area under the curve) of CMR for acute myocarditis is about 80% in the previous report.
3. However, this depends on the pre-test probability and timing/severity of myocarditis.

Accuracy ny vs gammal LLC

Radiology: Cardiothoracic Imaging

Comparison of Original and 2018 Lake Louise Criteria for Diagnosis of Acute Myocarditis: Results of a Validation Cohort

Julian A. Luetkens, MD • Anton Faron, MD • Alexander Isaak, MD • Darius Dabir, MD • Daniel Kuetting, MD
Andreas Feisst, MD • Frederic C. Schmeel, MD • Alois M. Sprinkart, MSc • Daniel Thomas, MD

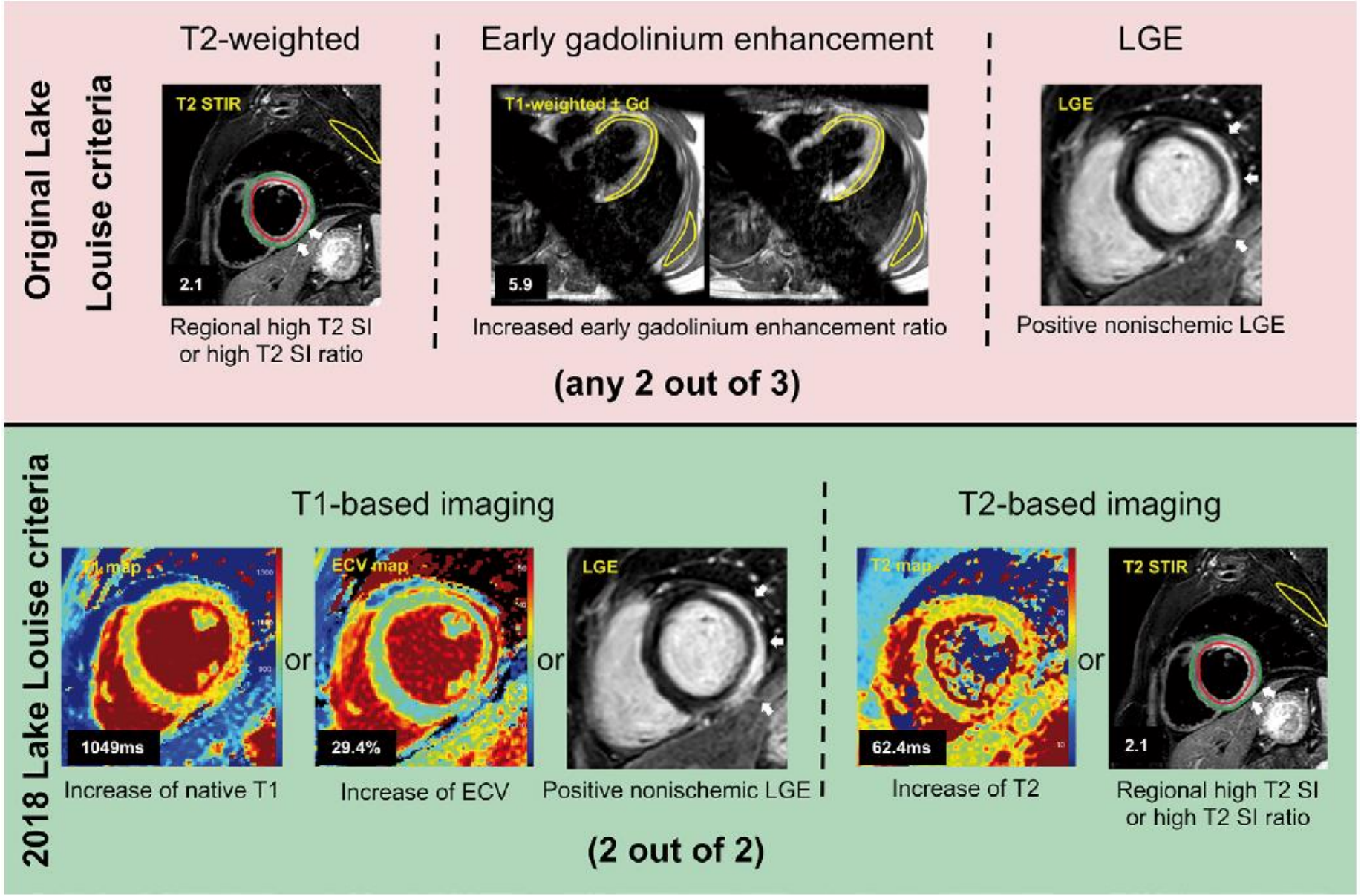


Figure 1: Illustration shows original and 2018 Lake Louise criteria (LLC) in a 24-year-old man with acute myocarditis. Original LLC consisted of three main criteria: regional high T2 signal intensities on T2-weighted images (white arrows) or increased global T2 signal intensity ratio, increased early gadolinium enhancement ratio on T1-weighted images, and areas with high signal intensities in nonischemic distribution pattern on late gadolinium enhancement (LGE) images (white arrows). 2018 LLC consist of two main criteria (T1-based criterion and T2-based criterion). T1-based criterion is considered to be positive if increase of native T1 relaxation times, increase of extracellular volume (ECV), or positive LGE (white arrows) exist. T2-based criterion is positive in cases of increased T2 relaxation times or in cases with regional high T2 signal intensities on T2-weighted images (white arrows) or increased global T2 signal intensity ratio. Gd = gadolinium, SI = signal intensity, STIR = short tau inversion recovery.

Table 2: Cardiac MRI Characteristics of Patients with Myocarditis and Healthy Control Participants

Variable	Patients with Myocarditis (n = 40)	Healthy Control Participants (n = 26)	P Value
General parameters			
Left ventricular ejection fraction (%)	52.4 ± 11.4	61.4 ± 3.6	<.001
Left ventricular end-diastolic volume index (mL/m^2)	86.8 ± 32.6	74.0 ± 12.6	.070
Interventricular septal thickness (mm)	9.8 ± 1.7	9.4 ± 1.9	.389
T2 signal intensity ratio	1.7 ± 0.3	1.5 ± 0.3	.006
Visible myocardial edema*	25 (62.5)	0 (0.0)	<.001
Early gadolinium enhancement ratio	3.6 ± 3.5	2.1 ± 1.4	.036
Visible late gadolinium enhancement*	31 (77.5)	0 (0.0)	<.001
Mapping parameters			
T1 native (msec)	1047.0 ± 53.8	965.8 ± 25.1	<.001
Extracellular volume fraction (%)	28.6 ± 5.3	26.1 ± 4.2	.038
T2 (msec)	61.8 ± 8.2	52.8 ± 2.4	<.001
Lake Louise criteria*			
Original Lake Louise criteria	29 (72.5)	1 (3.8)	<.001
2018 Lake Louise criteria	35 (87.5)	1 (3.8)	<.001

Note.—Unless otherwise specified, data are means ± standard deviations.
* Data are absolute frequencies, with percentages in parentheses.

Table 3: Diagnostic Performance of Different Cardiac MRI Parameters for Diagnosis of Acute Myocarditis

Variable	Cutoff Value	Sensitivity (%)	Specificity (%)	PPV (%)	NPV (%)	Accuracy (%)
Qualitative or semiquantitative parameter						
T2 signal intensity ratio	≥1.9	35 (22.1, 50.5)	88.5 (71.0, 86.0)	82.4 (59.0, 93.8)	46.9 (33.7, 60.6)	56.1 (44.1, 67.4)
Early gadolinium enhancement ratio	≥1.95	66.7 (50.3, 79.8)	57.7 (38.9, 74.5)	68.6 (52.0, 81.4)	55.6 (37.3, 72.4)	62.9 (50.5, 73.8)
Late gadolinium enhancement		77.5 (62.5, 87.7)	100.0 (87.1, 100.0)	100.0 (89.0, 100.0)	74.3 (57.9, 85.8)	86.4 (76.1, 92.7)
Quantitative parameter						
T1 native (msec)	≥1000	77.5 (62.5, 87.7)	96.2 (81.1, 99.3)	96.9 (84.3, 99.4)	73.5 (56.9, 85.4)	84.8 (74.3, 91.6)
Extracellular volume fraction (%)	≥28.8	45 (30.7, 60.2)	84.0 (65.3, 93.6)	81.8 (61.5, 92.7)	48.8 (34.6, 63.2)	60.0 (47.9, 71.0)
T2 (msec)	≥55.9	79.5 (64.5, 89.2)	92.3 (75.9, 97.9)	93.9 (80.4, 98.3)	75.0 (57.9, 86.7)	84.6 (73.9, 91.4)
Lake Louise criteria						
Original Lake Louise criteria		72.5 (57.2, 83.9)	96.2 (81.1, 99.3)	96.7 (83.3, 99.4)	73.5 (56.9, 85.4)	81.8 (70.9, 89.3)
2018 Lake Louise criteria		87.5 (73.9, 94.5)	96.2 (81.1, 99.3)	97.2 (85.8, 99.5)	83.3 (66.4, 92.7)	90.9 (81.6, 95.8)

Note.—Data in parentheses are 95% confidence intervals. Cutoff values were adopted from an initial study cohort (7). NPV = negative predictive value, PPV = positive predictive value.

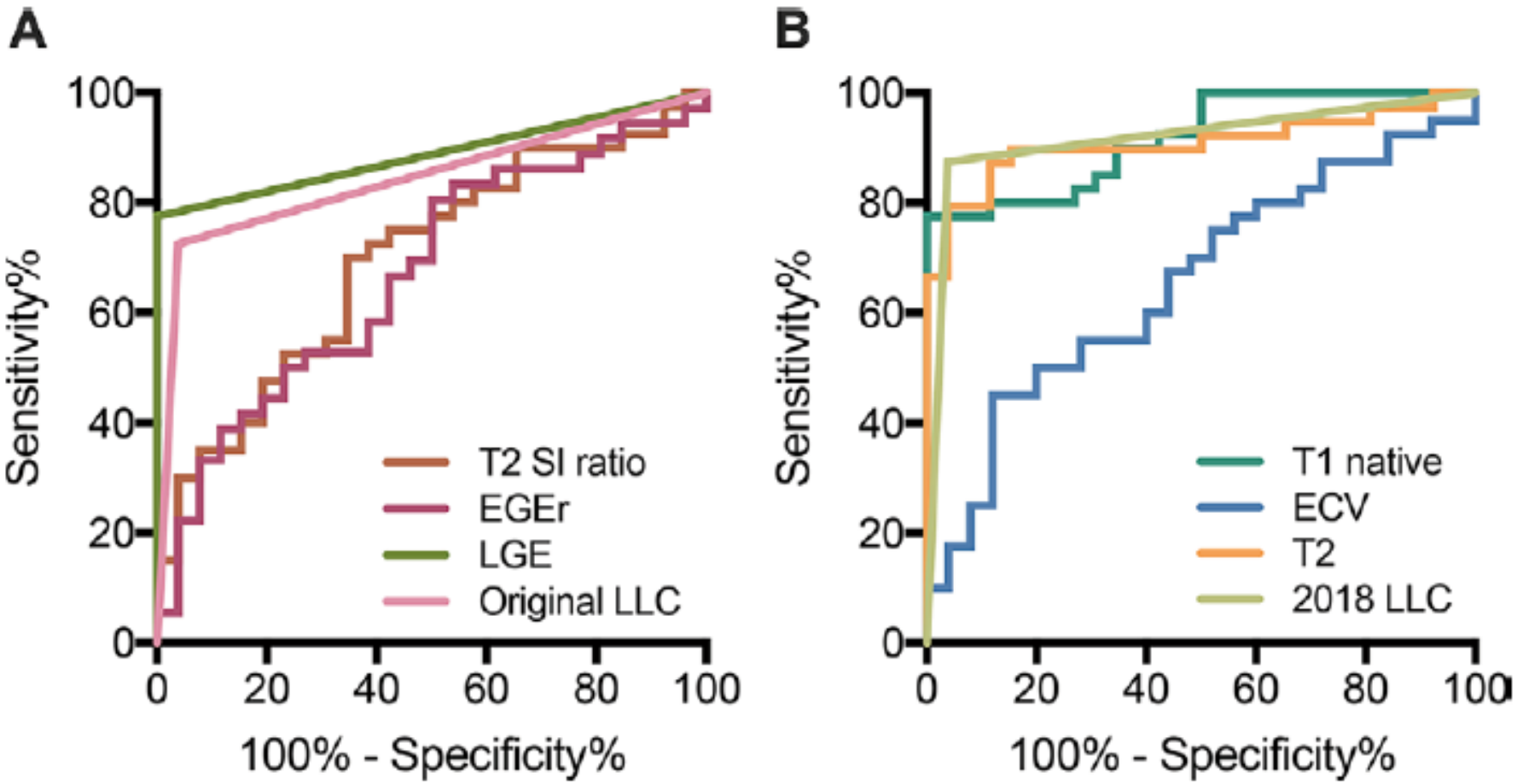


Figure 2: Graphs show receiver operating characteristic curves for, *A*, T2 signal intensity (SI) ratio (area under curve [AUC], 0.70), early gadolinium enhancement ratio (EGEr) (AUC, 0.67), late gadolinium enhancement (LGE) (AUC, 0.89), and Lake Louise criteria (LLC) (AUC, 0.84) and for, *B*, native T1 relaxation times (AUC, 0.92), extracellular volume (ECV) (AUC, 0.66), T2 relaxation times (AUC, 0.91), and 2018 LLC (AUC, 0.92).

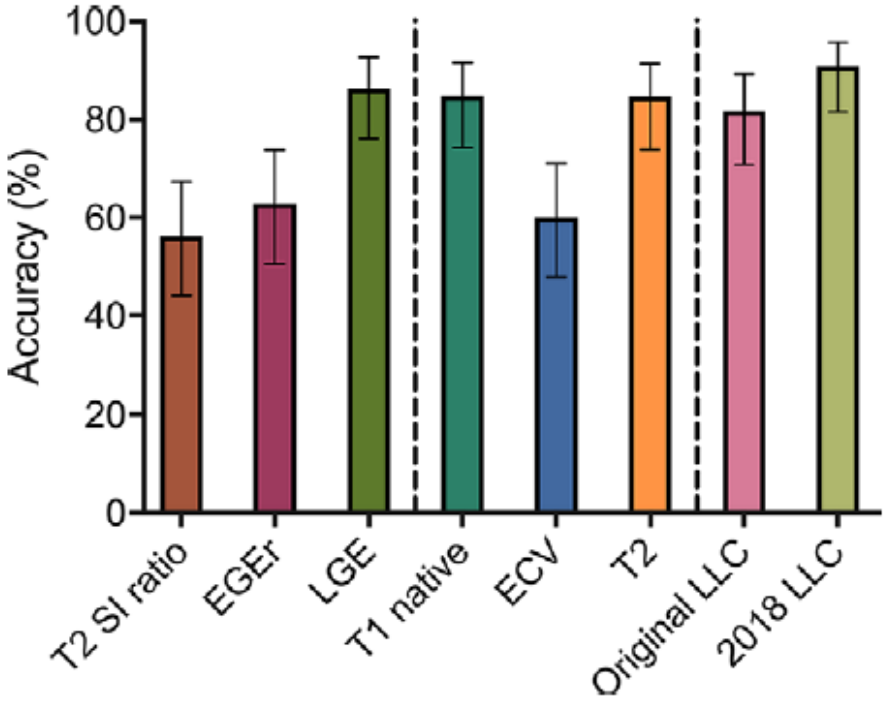


Figure 3: Column graph shows overall diagnostic accuracies for single parameters and for Lake Louise criteria (LLC). Error bars indicate 95% confidence intervals. ECV = extracellular volume fraction, EGEr = early gadolinium enhancement ratio, LGE = late gadolinium enhancement, SI = signal intensity.

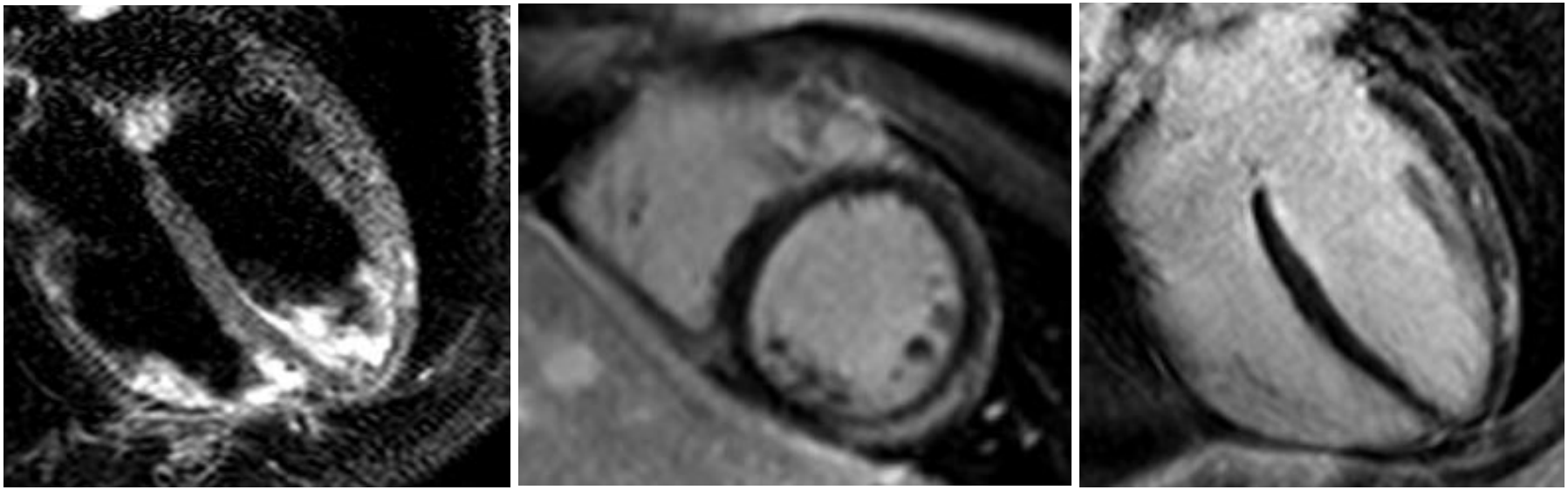
Summary

The 2018 Lake Louise criteria provide a high diagnostic accuracy for the diagnosis of acute myocarditis and significantly increase the sensitivity compared with the original score.

Key Points

- Multiparametric cardiac MRI has a high diagnostic value for the diagnosis of patients with acute myocarditis.
- The 2018 Lake Louise criteria improve the diagnostic performance of cardiac MRI by significantly increasing its sensitivity compared with the original criteria.

Myokardit, 25 åå, man

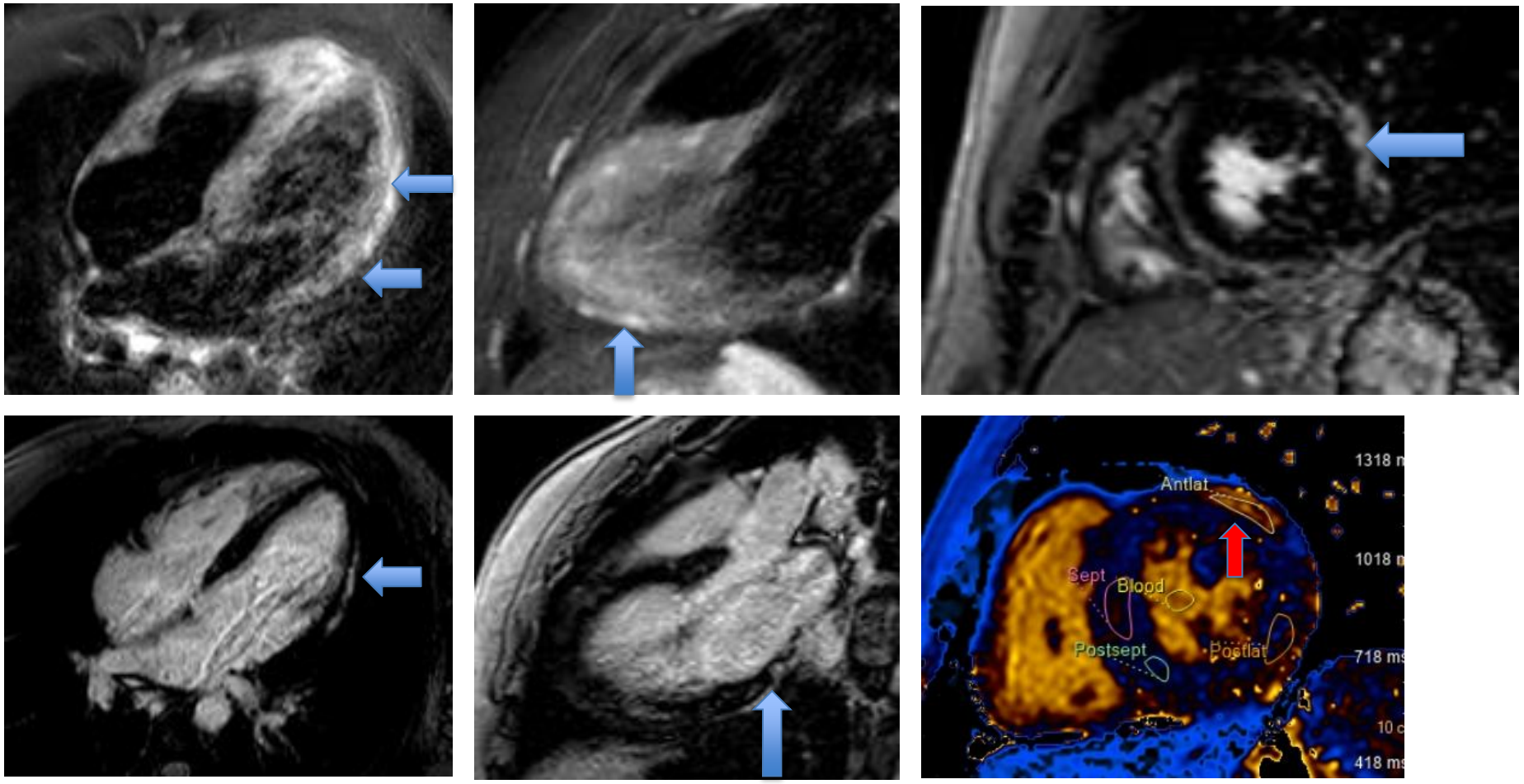


2009 hade vi inte samma bildkvalitet och inte tillgång till T-mapping-tekniker

Recidiv av myokardit

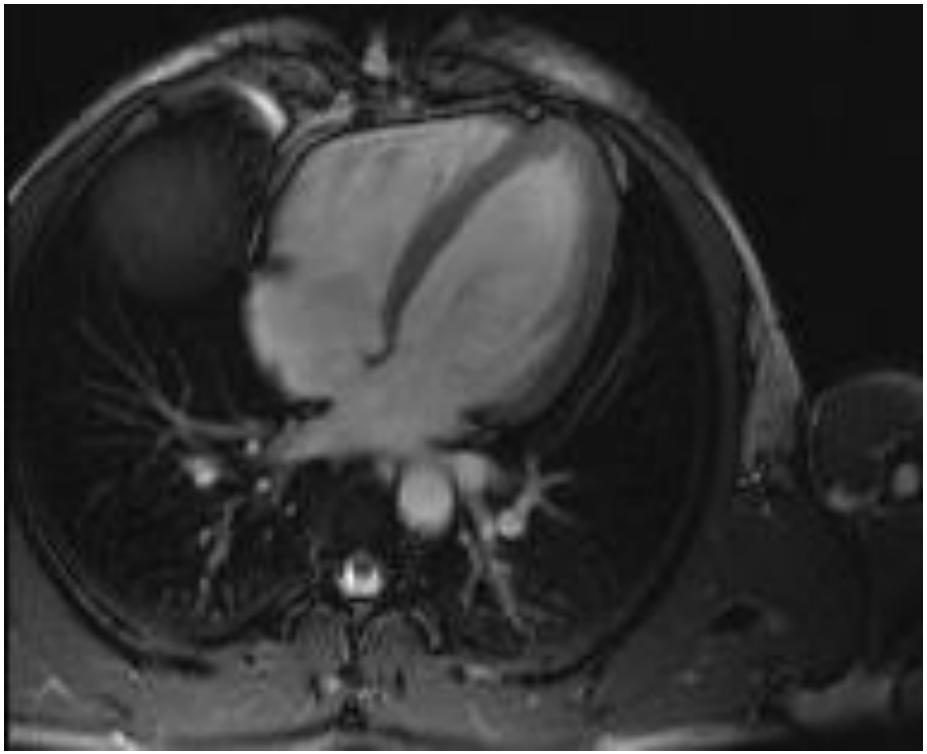
19-åring med trop 1400, proBNP 650, förkyld, bröstont

ECV antlat 39%
T1 antlat 1226 ms

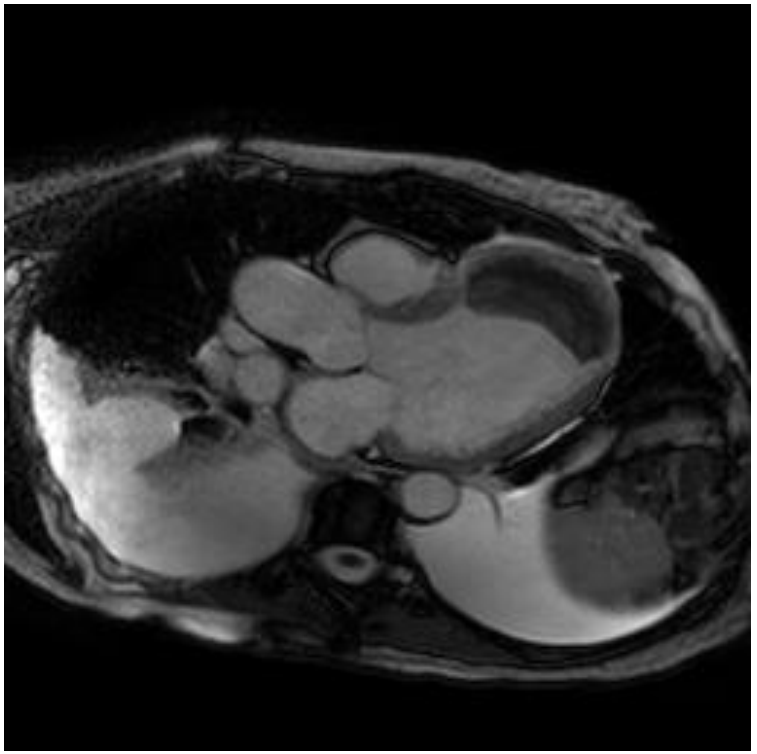


Systolisk vänsterkammarfunktion

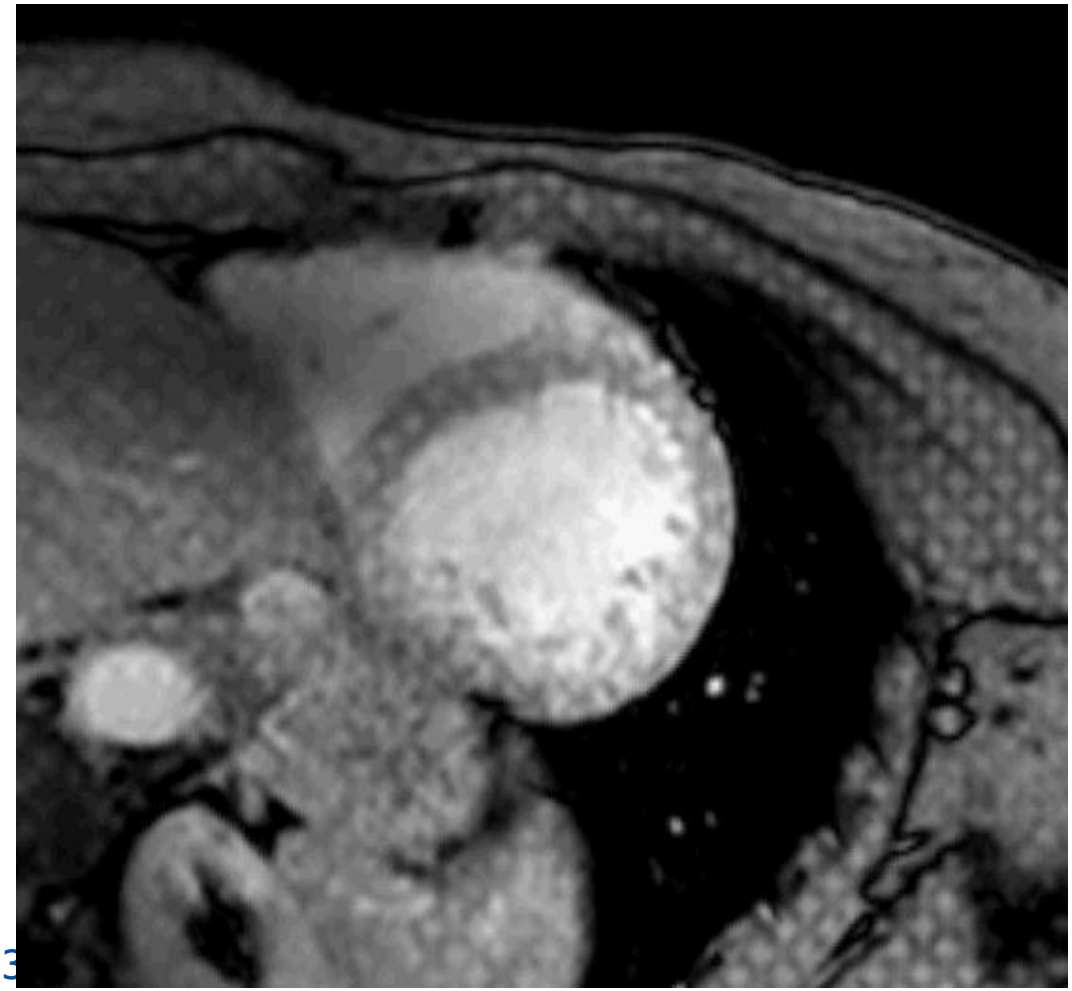
- Normal kammarstorlek
- Normal långaxelförkortning
- Normal väggförtjockning
- Normal ejektionsfraktion



Ischemisk kardiomyopati
Nedsatt väggrörlighet
Regionala skillnader
Tromb

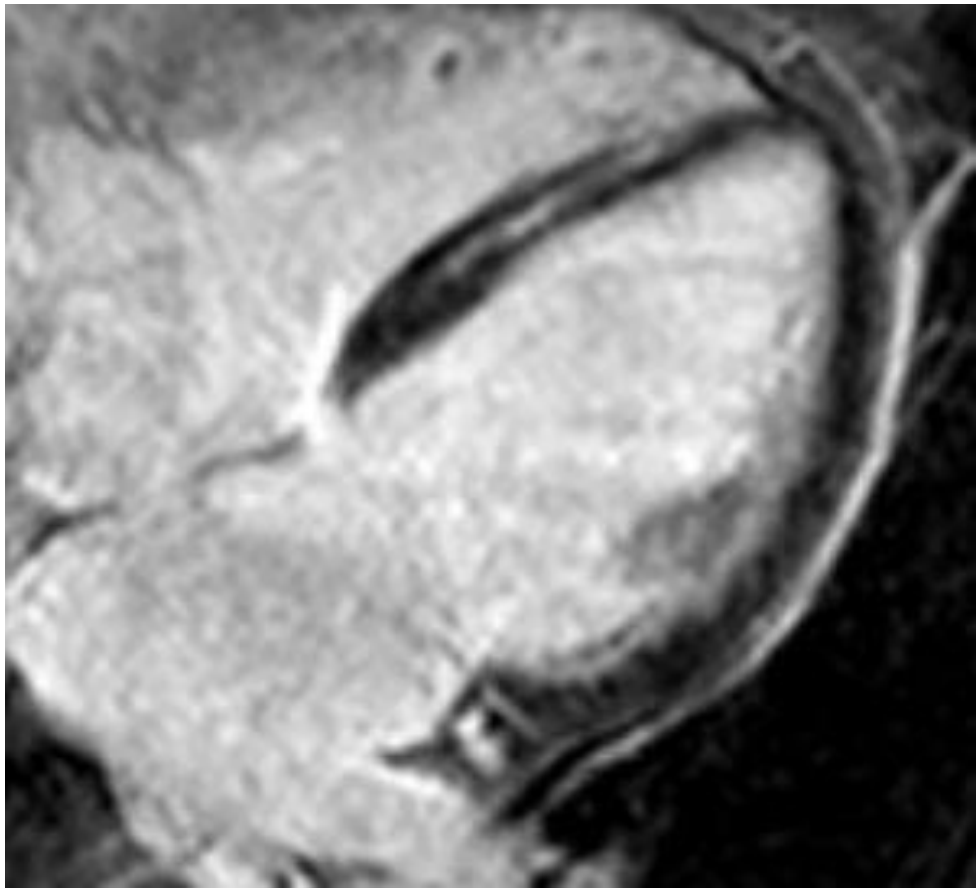


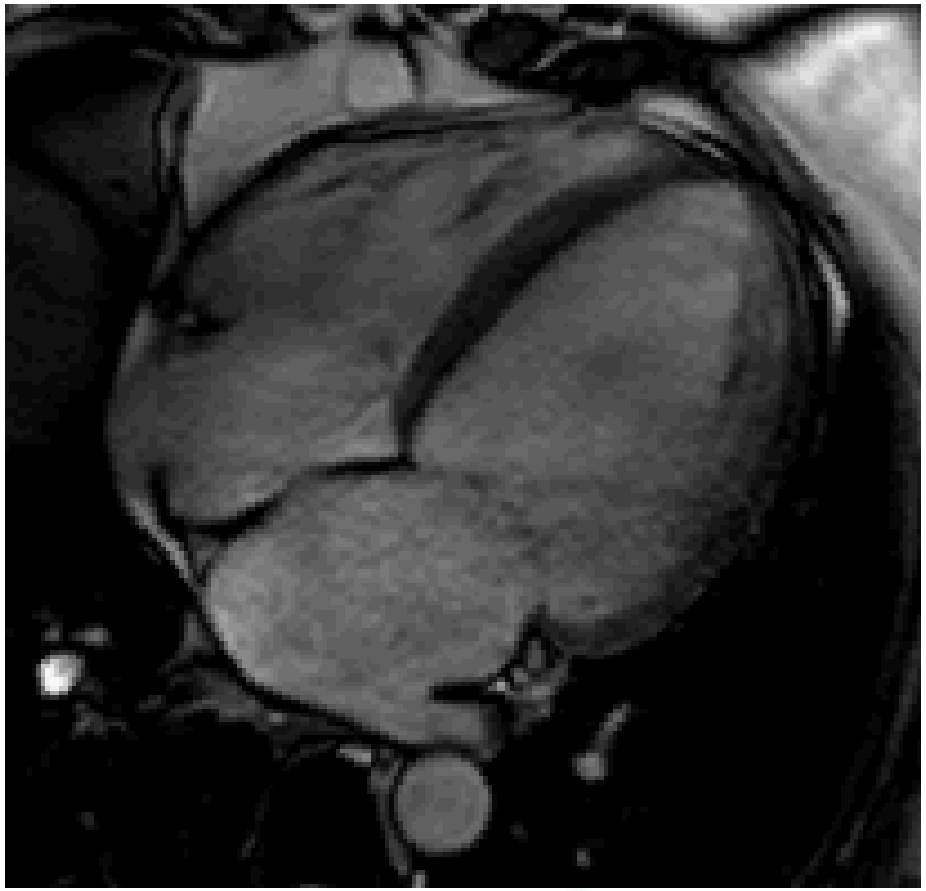
Normal funktion med tagging



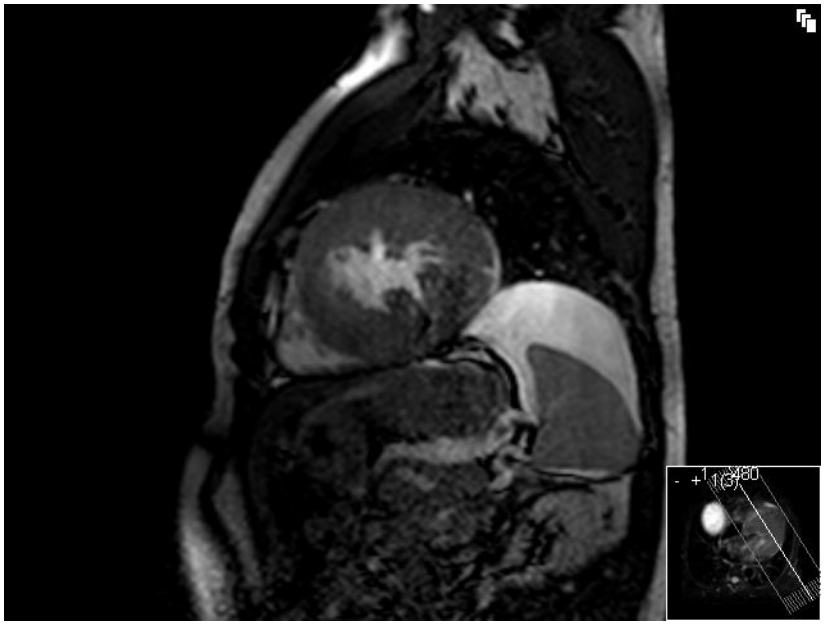
Region Östergötland

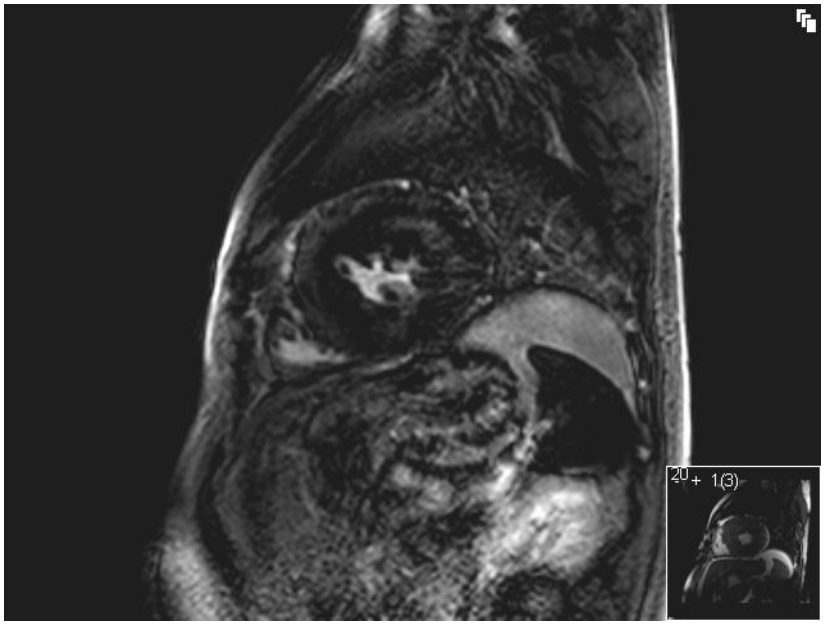
Dilaterad kardiomyopati

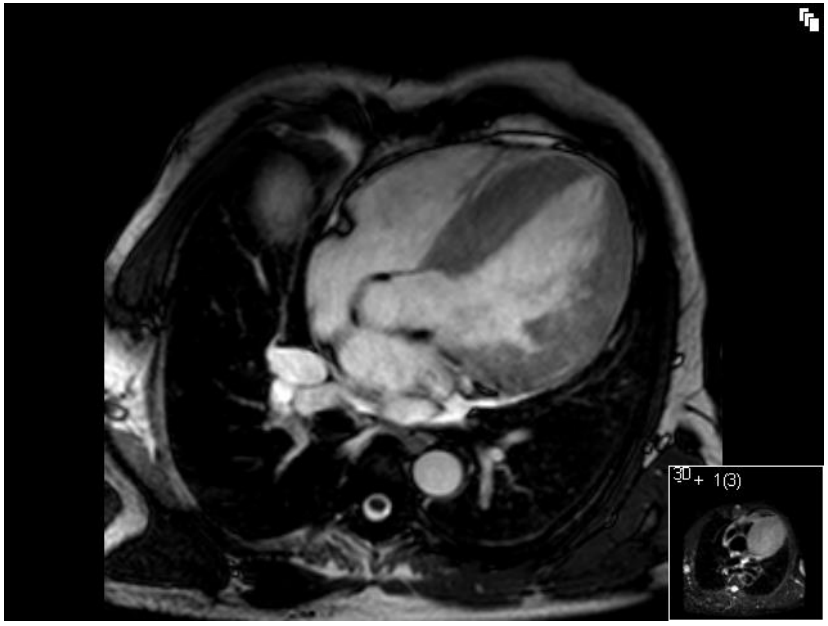




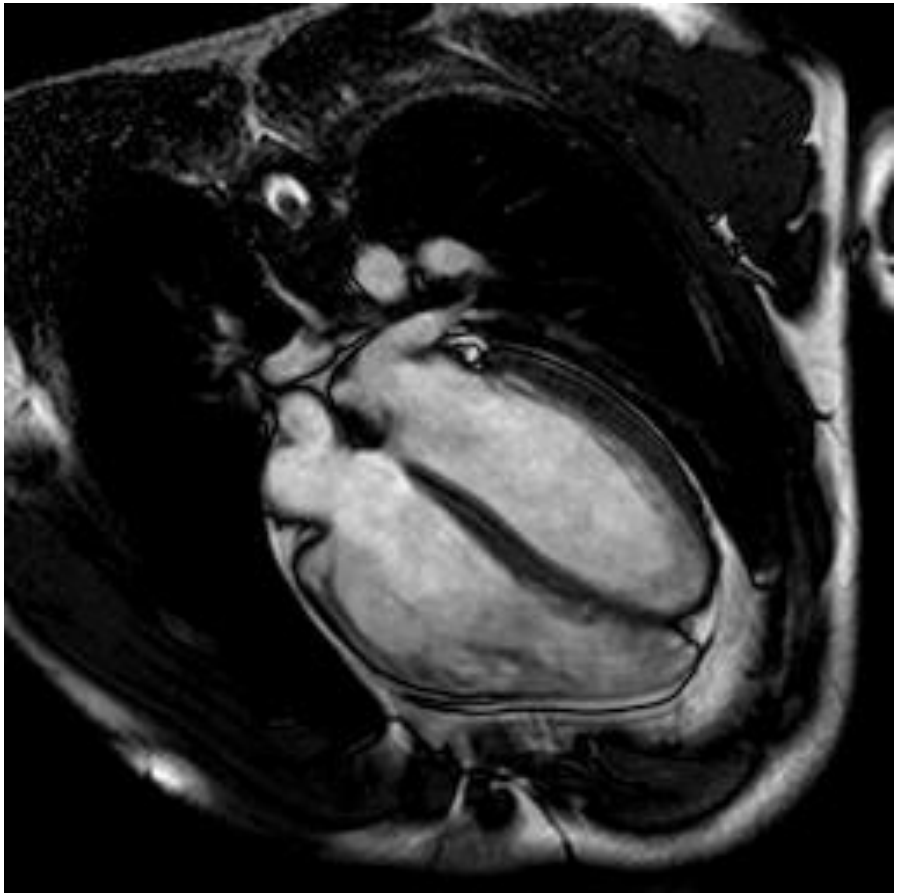
Hypertrof kardiomyopati - inlagring?

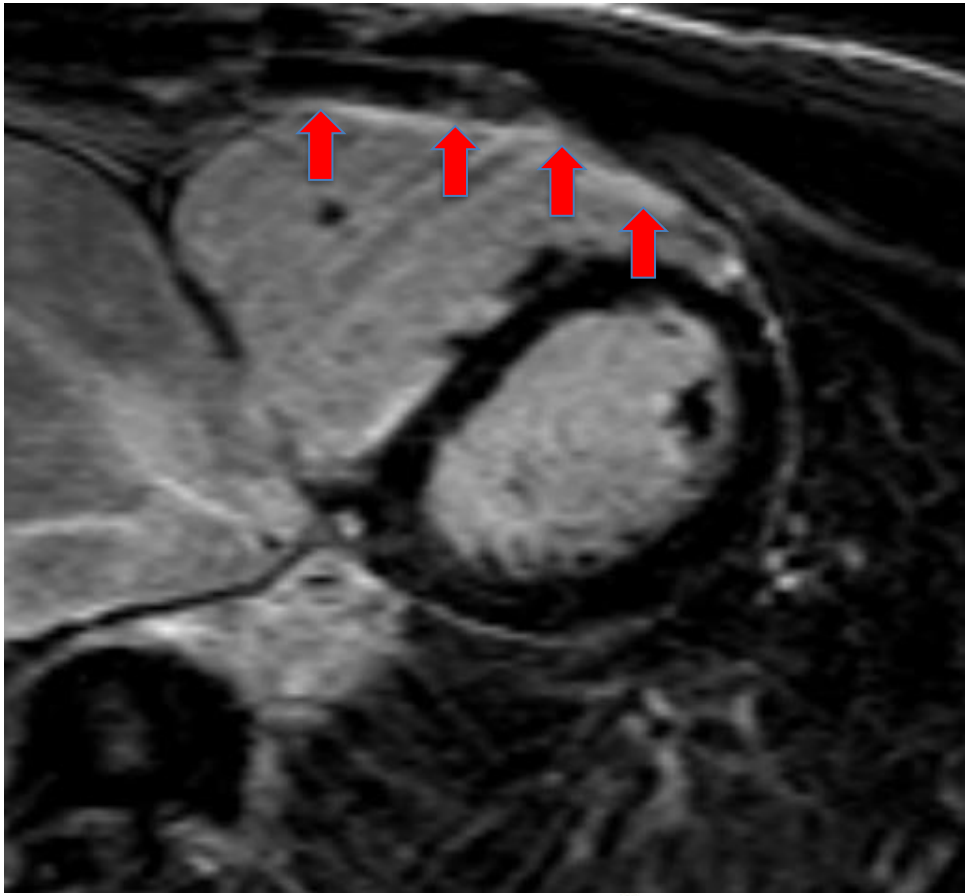






Arytmogen kardiomyopati



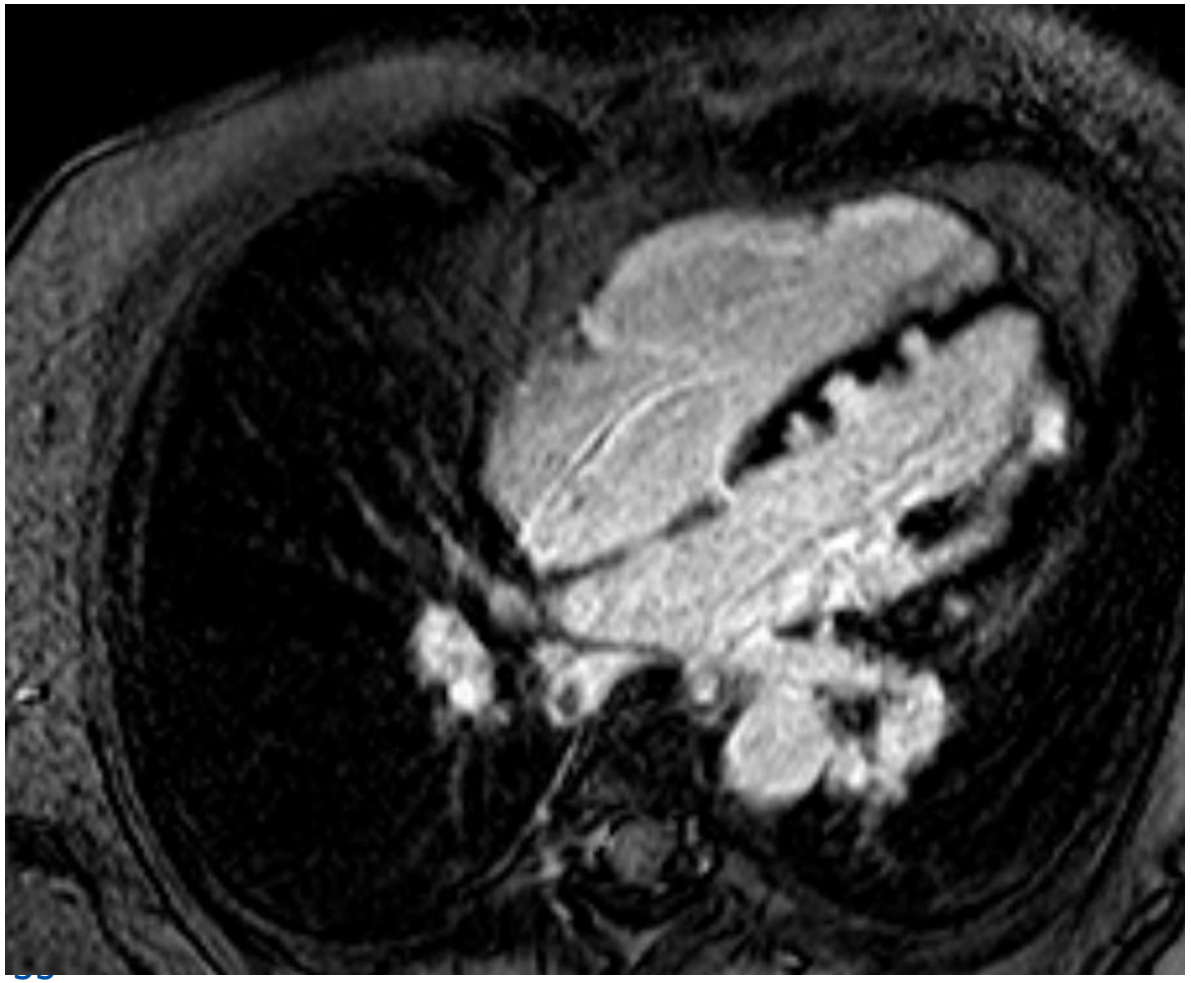


Inlagring (amyloid)

Amyloid kännetecknas av svårighet att ställa in inversionstid. Kan också ha LGE makro.

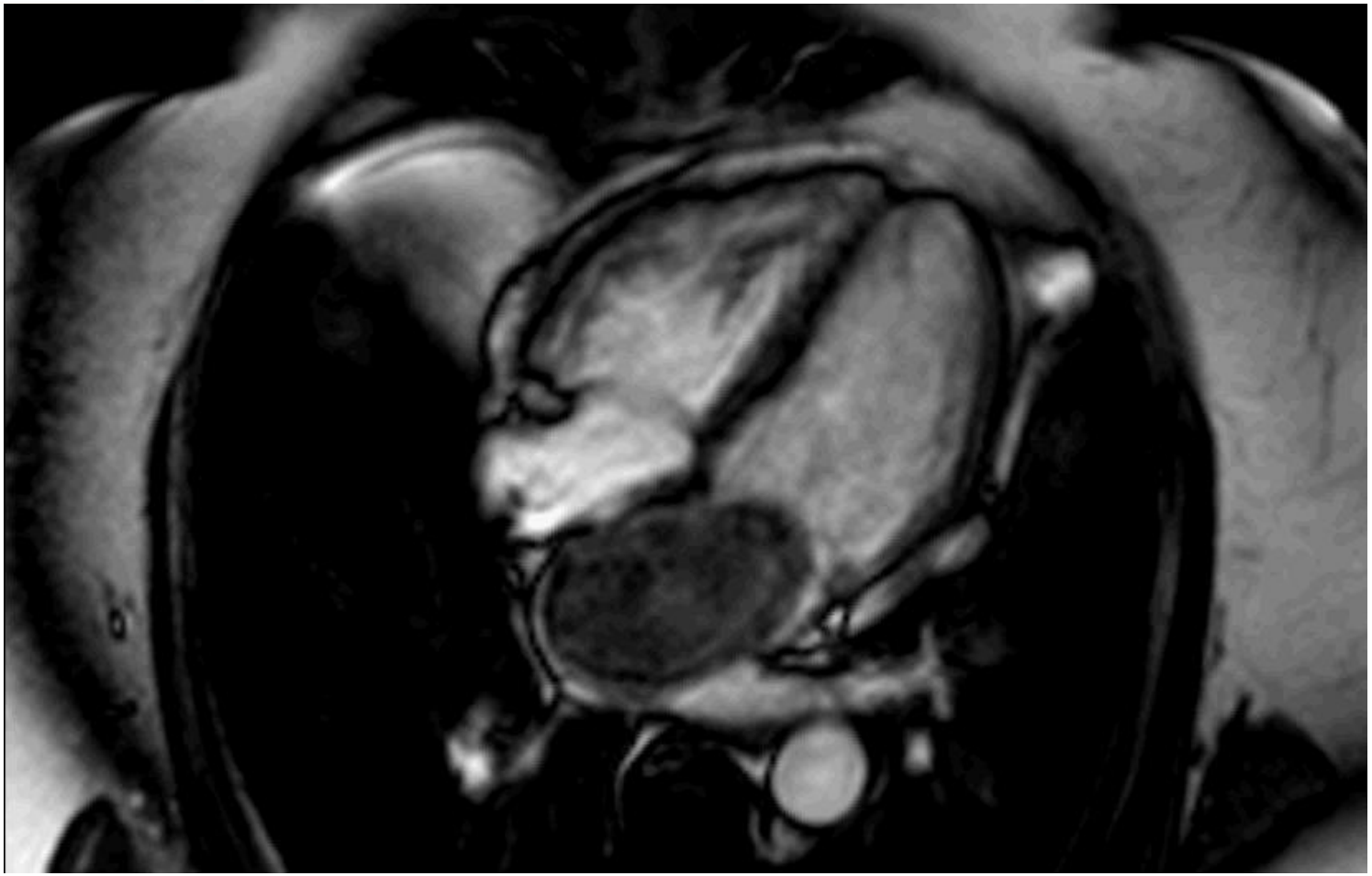
Region Östergötland

Sarkoidos



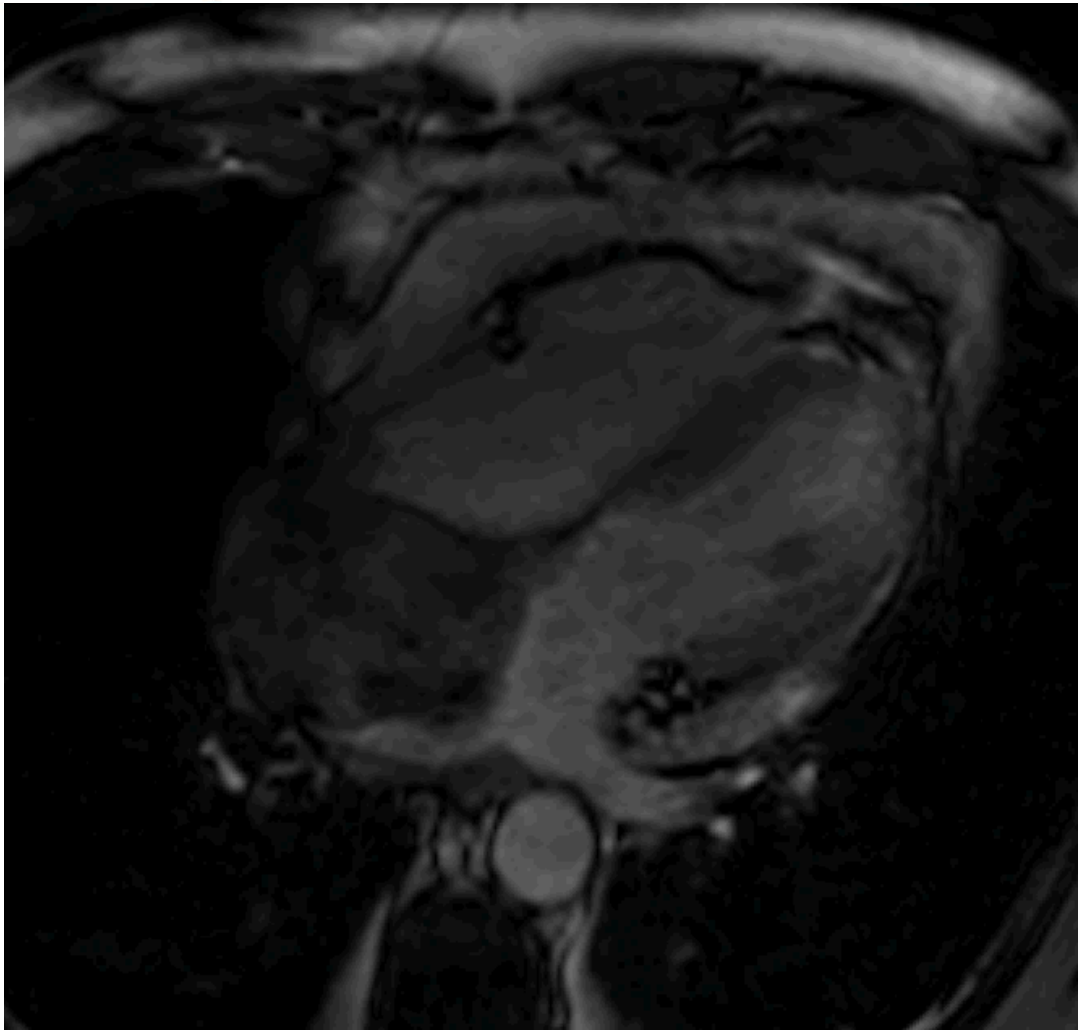
Sarkoidos kan se ut på alla möjliga sätt vid LGE

Hjärttumör – vilken?



Region Östergötland

Hjärttumör 2



Sarkom i förmaksseptum

Region Östergötland