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Cardiac involvement in secondary disease

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Conflict of interest

I am a founder/director of MycardiumAI (for corelab work)

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Introduction

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1) Vasculitis

2) Muscular Dystrophies

3) Sarcoidosis

4) Amyloidosis

5) Iron Overload Cardiomyopathies

6) Athlete's Heart

7) Endomyocardial Fibrosis

8) Chagas Disease

9) Fabry's Disease

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1) Vasculitis

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1/ Vasculitis
- LV dysfunction
- LGE pattern

Rahman et al, JCMR, 2012; 14(1): 82

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Vasculitis

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Inflammation of blood vessels

Leads to

- Blindness
- Renal failure
- Aortic rupture
- Heart Fai

Look for vascular imaging

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Vasculitis

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Rare

High morbidity/mortality

Diagnosed late

Several classifications

- Just use the size of vessels
  - Large – Giant Cell, Takayasu's, Idiopathic
  - Mid – Kawasaki Disease, Polyarteris Nodosa, Behçet's
  - Small – Churg Strauss

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Vasculitis - Types

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Table 2

Typical arterial segments involved in the major primary vasculitides

	Thoracic Aorta	Abdominal Aorta	Pulmonary Arteries	Carotid Arteries	Upper Extremities	Mesenteric arteries	Renal arteries	Lower Extremities	Coronary Arteries
Giant cell arteritis	X	X		X	X				
Takayasu arteritis	X	X	X	X	X				
Polyarteritis nodosa						X	X		
Kawasaki disease									X
Behçet disease			X					X	

The most common conditions prompting referral for MR examination are shown with X indicating typically-involved segments of the extracranial arterial tree. This scope warrants consideration when prescribing the imaging protocol. Note that atypical manifestations have been reported in virtually all vessel territories for these disorders.

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Vasculitis - Imaging

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Table 1

Components of the CMR Examination of Vasculitis

Technique	Comments
PRECONTRAST	
Dark blood stacks typically in axial, coronal and sagittal planes e.g. HASTE	Provides vessel wall imaging as well as complementary information to CE-MRA regarding lumen
Noncontrast bright blood stack(s) e.g. SSFP	
	CONTRAST
3D contrast-enhanced magnetic resonance angiography e.g. spoiled gradient echo	Appropriate vasculature should be covered depending on clinical questions and known or suspected diagnosis (see Table 2)
	POSTCONTRAST
T1-weighted vessel wall imaging e.g. VIBE or FAME	Additional vessel wall imaging, particularly useful to delineate thickening and thrombus
	CARDIAC ACQUISITIONS
Multiplane cine imaging e.g. SSFP Aortic valve velocity-encoded cine Myocardial imaging: T2 precontrast, T1W early post contrast, late post-gadolinium imaging	May be appropriate when aortic root disease involves the aortic valve or when myocardial inflammation is suspected, particularly in small-vessel vasculitides

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Vasculitis

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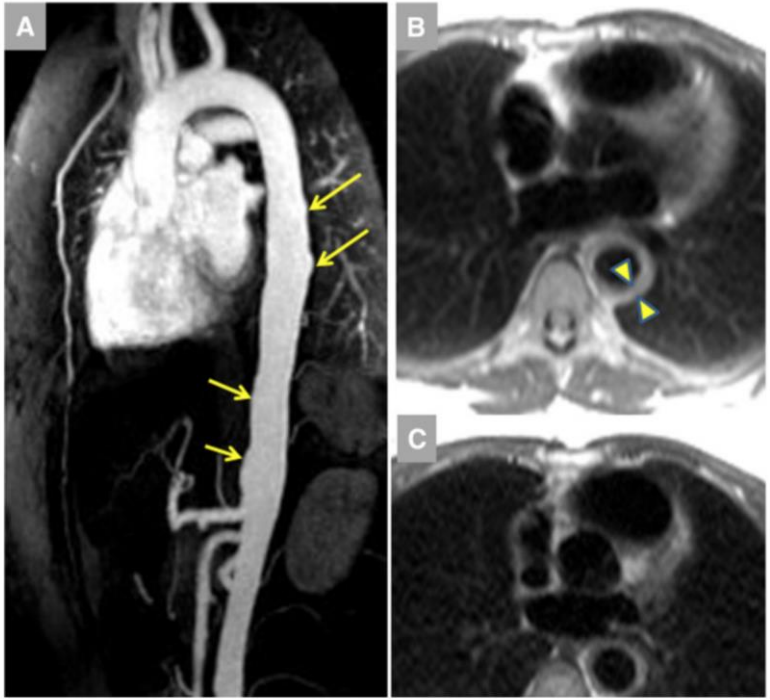

Vasculitis was identified in a 28 year-old female with unrelenting back pain initially referred for MR examination to rule out aortic dissection; additional history revealed recent unintentional 5 kg weight loss.A. Contrast-enhanced magnetic resonance angiography (CE-MRA) showed diffuse luminal irregularities (arrows). **B**. Pre-contrast dark blood imaging indicated marked aortic wall thickening to 9–10 mm (arrowheads). There was marked elevation of inflammatory markers including erythrocyte sedimentation rate (ESR, 94 mm/hr) and c-reactive protein (7.3 mg/L) levels. Symptoms markedly improved with prednisone, with reduced ESR (12 mm/hr) and aortic wall thickness at 12-month follow-up.

Vasculitis – Large - Takayasu

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Women > Men

> 50 yrs

Need angiographic findings AND one of:

- Decreased pulses/claudication
- Blood pressure differences
- Bruits
- hypertension

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Vasculitis – Large – Takayasu's

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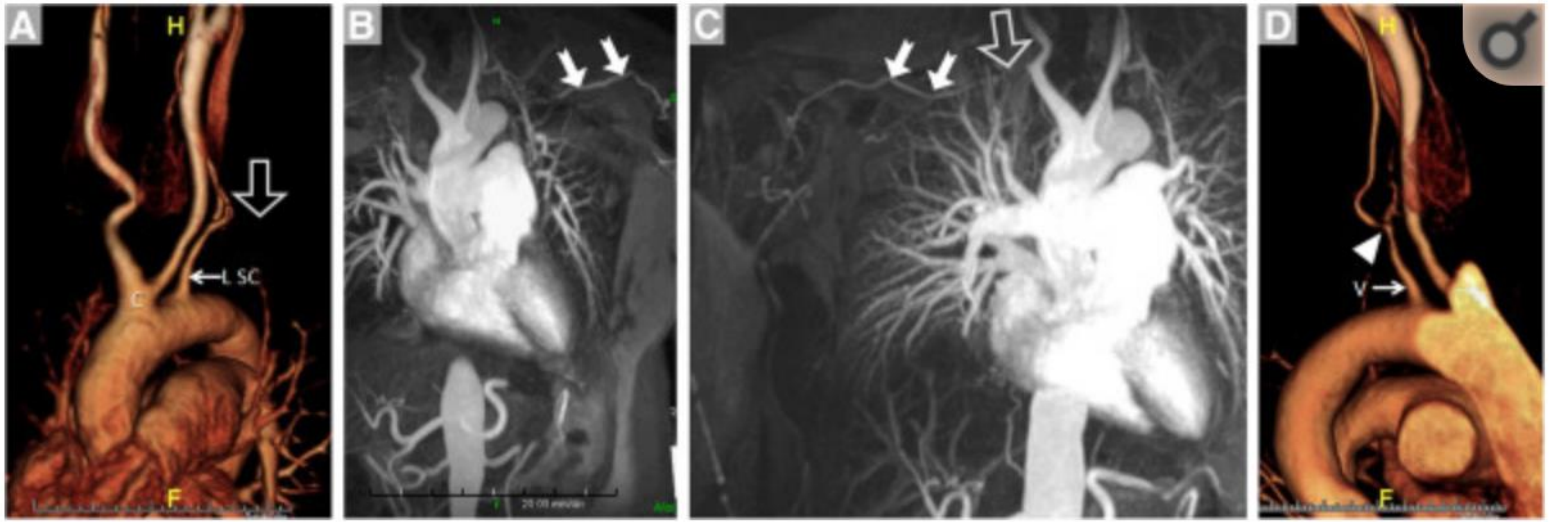

A 42 year-old female presented with bilateral arm fatigue, worse with lifting above the head. Physical examination showing absent radial pulses, and serum inflammatory markers including erythrocyte sedimentation rate and c-reactive protein levels were elevated. Anemia was also present (hematocrit 30%). With a presumptive diagnosis of Takayasu arteritis, treatment with prednisone was initiated and CE-MRA was requested. **A**. Volume rendering shows patency of the common brachiocephalic trunk (C); the proximal portion of the left subclavian artery (L SC, arrow) is patent while distally it is occluded (open arrow). **B**. Maximum intensity projection (MIP) shows reconstitution of the distal L SC (arrows) via collaterals. **C**. Similarly, a MIP image shows that the right subclavian artery is occluded (open arrow) and fills distally (filled arrows) via collaterals. **D**. Volume rendering demonstrates high-grade stenosis (arrow) of the left vertebral artery (V).

Vasculitis – Large – Takayasu's

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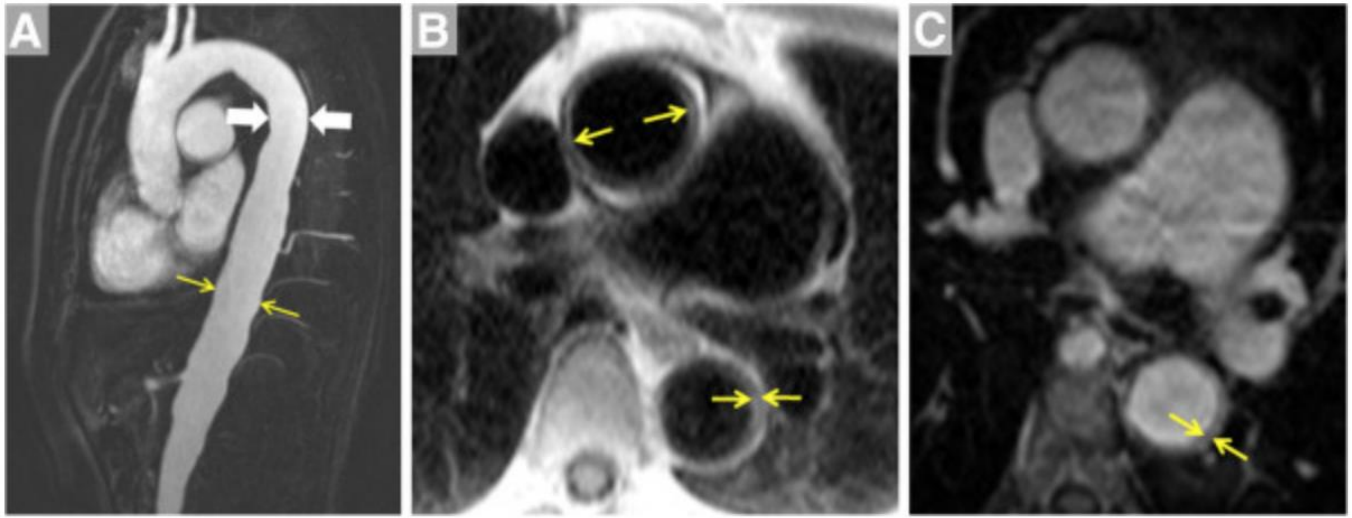

Images of the thoracic aorta in a patient with Takayasu arteritis are shown.A. CE-MRA in the sagittal plane demonstrates diffuse, mild dilatation of the descending aorta that measured 33 mm at the level indicated by arrows vs. 25 mm more proximally (arrowheads). Vessel wall thickening can be appreciated using techniques such as non-contrast inversion recovery dark blood imaging (**B**, showing thickening of 4–5 mm of the thoracic aorta wall, arrows). Additional post-contrast T1-weighted imaging such as the volumetric interpolated breathhold technique (**C**, same location as **B**) further confirm vessel wall thickening in this patient.

Vasculitis – Large – Idiopathic

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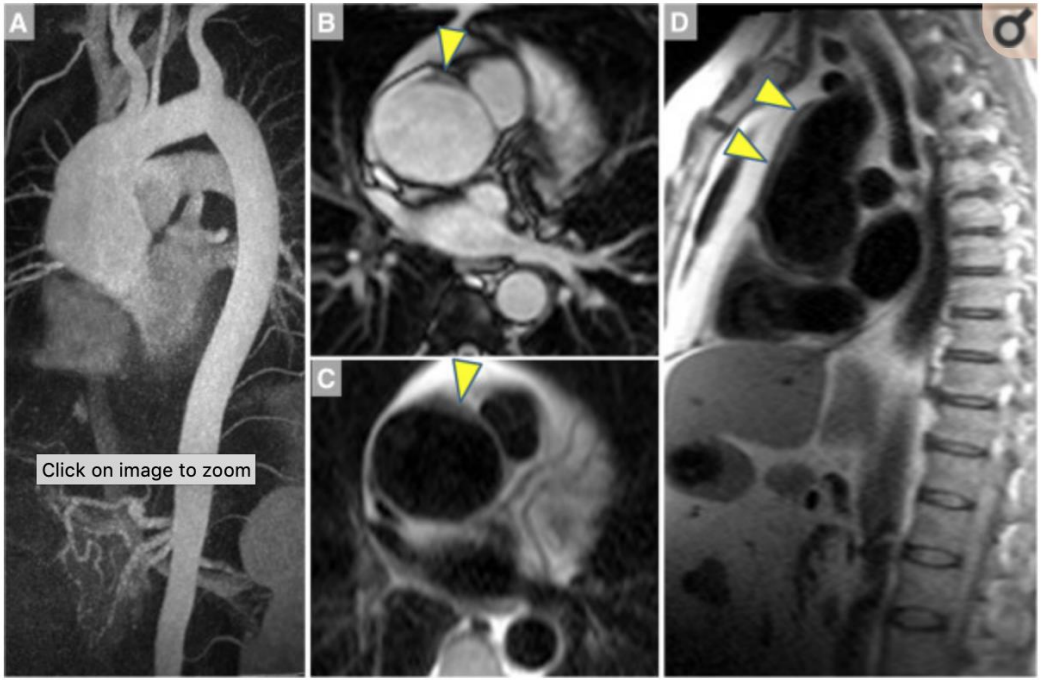

A 58 year-old male with fatigue and palpitations underwent transthoracic echocardiography that indicated dilatation of the aortic root. CMR was ordered to assess the aorta. **A**. MIP of the CE-MRA shows marked dilatation of the ascending aorta, which measured up to 6 cm in diameter compared to the 2.5 cm arch. **B**. Single heartbeat true FISP bright blood image shows thickening of the aortic wall (arrowhead), also evident on HASTE dark blood imaging in the axial (**C**) and sagittal planes (**D**).

Vasculitis – Mid - Kawasaki

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Children

Fever AND four of five from:

- Desquamating Rash (extremities/perineal area)
- Polymorphous Exanthema
- Bilateral conjunctival injection
- Injection of oral/pharyngeal mucosa
- Cervical lymphadenopathy

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Vasculitis – Mid - Kawasaki

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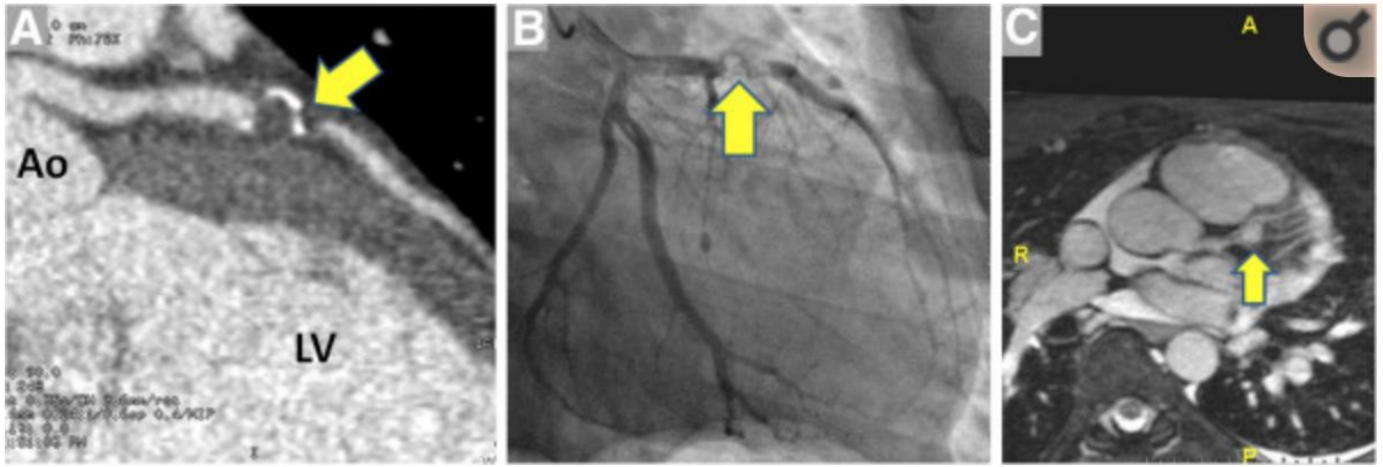

An 11 year-old boy presented to a pediatric hospital with chest and jaw pain while playing one year after a prolonged febrile illness. Initially, CT angiography was performed (**A**, image courtesy of Dr. Christopher Learn) that showed thrombus in a calcified aneurysm of the left anterior descending coronary artery (LAD, arrow). In the setting of elevation of the serum troponin and possible need for coronary intervention, the patient was transferred to a nearby adult hospital. Invasive angiography (**B**) showed thrombus nearly occluding LAD that was treated with angioplasty and stent placement. **C**. Coronary MRA performed in another patient with KD using a navigator-triggered slab prescribed perpendicular to the aortic root demonstrates a 9 mm proximal LAD aneurysm (arrow). LV = left ventricle Ao = aorta.

Vasculitis – Mid - Kawasaki

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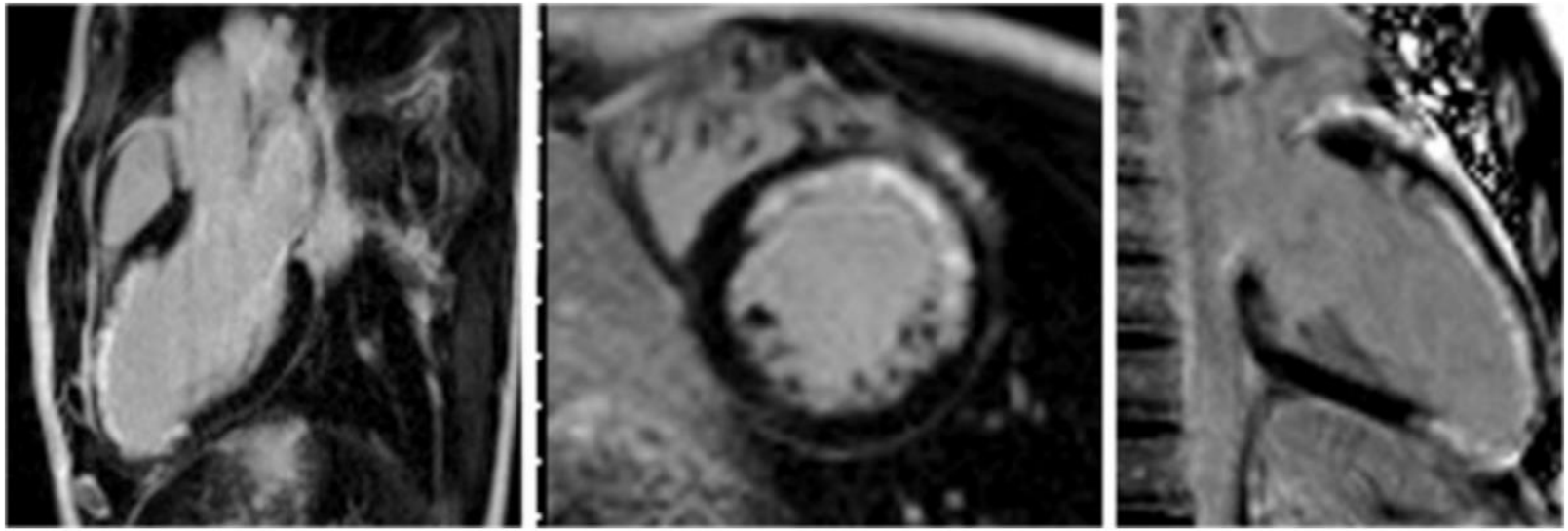

LGE-CMR in three-chamber (left), mid short-axis (center) and vertical long axis (right) planes show LAD-territory infarct scar in a boy with Kawasaki disease.

Vasculitis – Small – Churg Strauss

Significant constitutional symptoms

Clues include:

- Asthma
- Hypereosinophilia (we will do Loeffler's later!)
- Fever

ANCA positive

Vasculitis – Small – Churg Strauss

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Cardiac involvement

Pericarditis

Pericardial effusions

Myocarditis

Thrombi

Scarring

Subendocardial

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Vasculitis – Small – Churg Strauss

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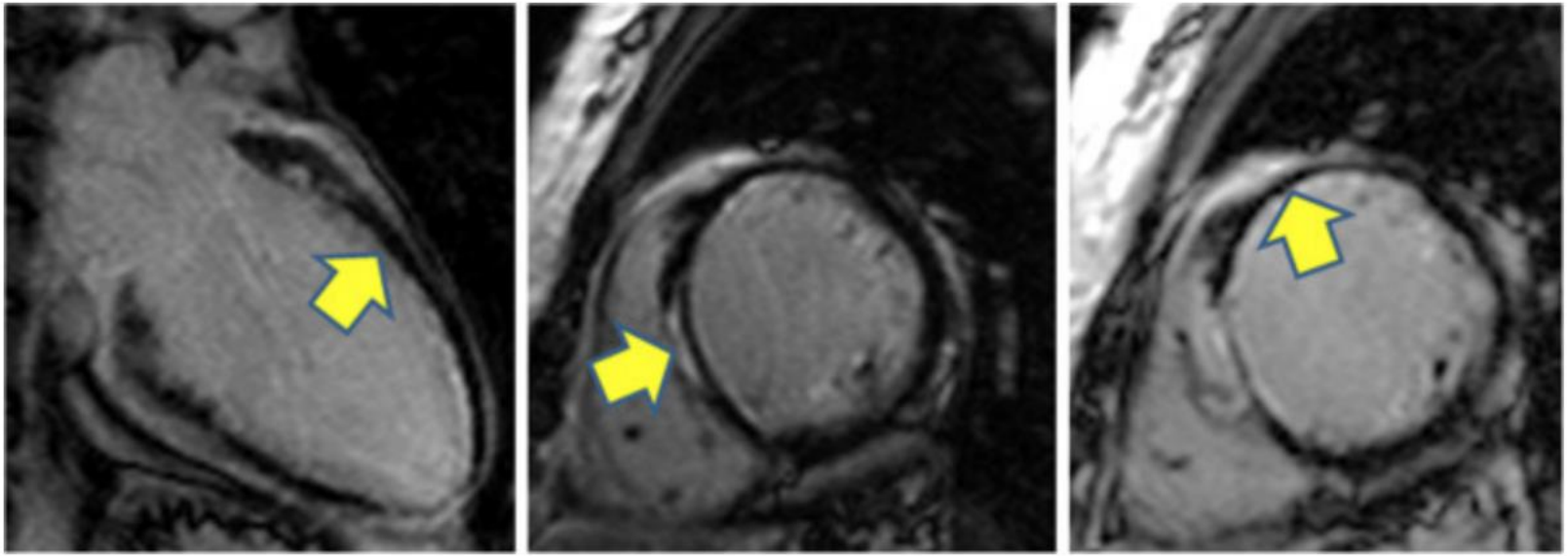

A 37-year-old female with biopsy-proven Churg-Strauss-vasculitis was referred for CMR examination The left ventricle was slightly enlarged with mild systolic dysfunction: LV ejection fraction was 45%. Late post-gadolinium myocardial enhancement images in various planes show septal intramural and anteroseptal and anterior subendocardial lesions. Images courtesy Drs. Ralf Waβmuth and Jeanette Schulz-Menger.

Vasculitis – Crib Sheet

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Large vessel and thick/inflammed

Takayasu

Child, medium vessel (coronaries), Infarction

Kawasaki

Generalised symptoms and diffuse subendocardial LGE

Churg Strauss

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2) Muscular Dystrophies

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2/ Muscle dystrophy

- LV enlargement, LV dysfunction
- LGE pattern
- Fatty infiltration

Verhaert et al. JACC Cardiovascular Imaging 2011 ; 4: 67

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2) Muscular Dystrophies

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Group of diseases

Weakening/breakdown of skeletal muscle

Genetic

Over 30 types including

- Duschenne Muscular Dystrophy
- Becker Muscular Dystrophy
- Lamin A/C

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2) Muscular Dystrophies - DMD

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X linked

Men only!

Cannot walk by age 12

Can affect the myocardium

Even if a female carrier

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2) Muscular Dystrophies - DMD

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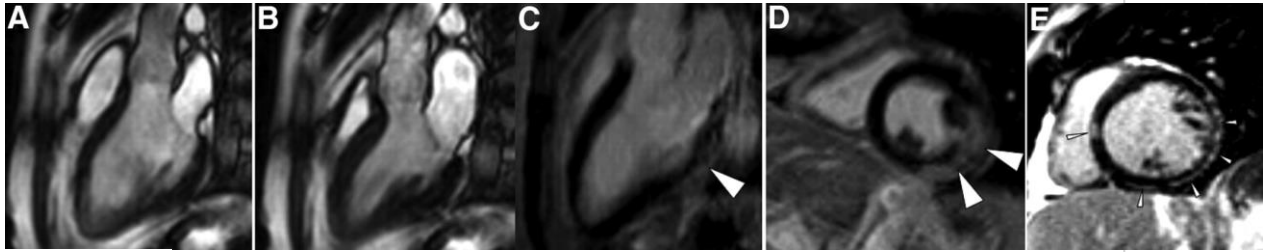

Figure 2. CMR findings in DMD in patients with different degrees of cardiac involvement. End-diastolic (A) and end-systolic (B) frames from a 3-chamber, long-axis cine acquisition (supplemental movie 1) show preserved LV systolic function in this 28-year-old man with DMD. LGE images (C, 3-chamber view; D, midventricular short-axis view) in the same patient show that despite preserved global LV systolic function, myocardial injury is evident as subepicardial fibrosis of the inferolateral wall (arrowheads). E, LGE in a 14-year-old boy with DMD shows more advanced cardiomyopathy with profound LV dilatation and systolic dysfunction (supplemental movie 2) and more extensive subepicardial scarring as well as septal fibrosis (arrowheads).

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2) Muscular Dystrophies - BMD

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X linked

Men only!

Less severe form of DMD

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2) Muscular Dystrophies - BMD

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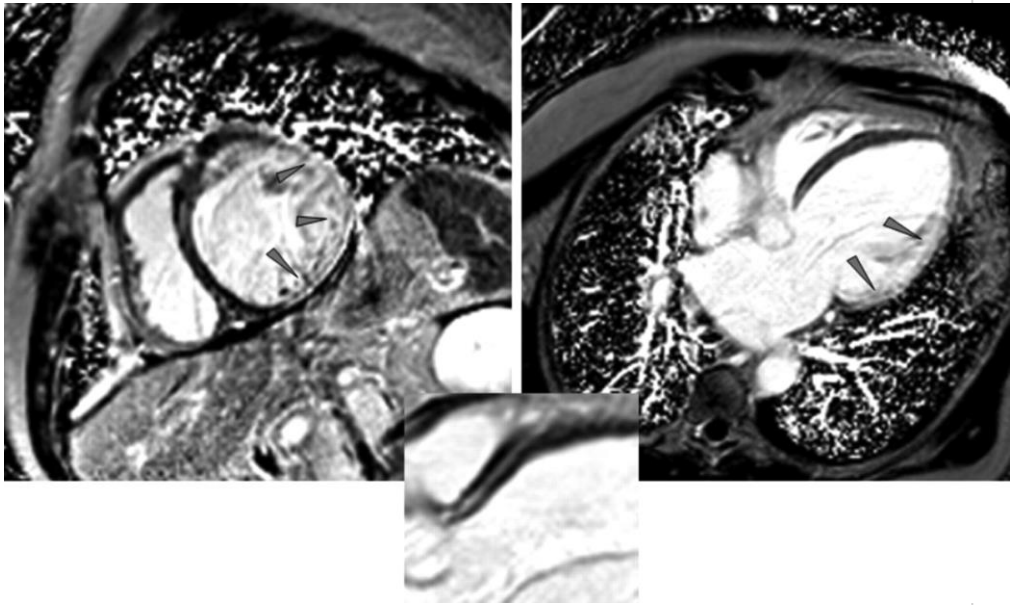
Figure 3. LGE findings in BMD. The pattern of myocardial injury in patients with BMD is similar to that seen in DMD, starting at the subepicardium of the inferolateral wall, with an age-dependent increase in the extent of fibrosis and progressive decline in systolic function. The left and right upper panels (short-axis and horizontal long-axis views, respectively) show almost transmural hyperenhancement of the entire anterolateral and inferolateral walls, consistent with advanced disease. In addition, this patient also had evidence of septal midwall fibrosis (lower middle panel), also seen in myocarditis and other nonischemic cardiomyopathies.



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2) Muscular Dystrophies – Lamin A/C

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Involved in nuclear membrane

Contractures

Muscle Weakening

Cardiac conduction defects

Fibrosous/adipose tissue replacement of myocardium

- Works from atrium down AV node to LV
- Leads to dilated impaired LV

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2) Muscular Dystrophies - BMD

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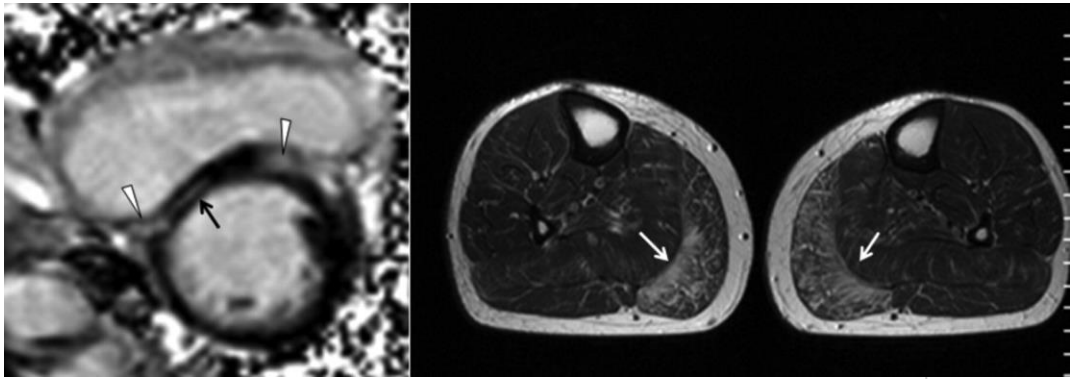

Figure 5. CMR findings in lamin A/C cardiomyopathy. Lamin A/C cardiomyopathy has been associated with midwall fibrosis of the midventricular septum (left panel, arrow) at an early stage of the disease. Note also the presence of fibrosis at the right ventricular–LV septal insertion sites (arrowheads) in this patient. Unlike patients with different types of *LMNA* mutations (EMDM, LGMDB1), lamin A/C cardiomyopathy does not typically produce apparent skeletal muscle weakness. Nevertheless, muscle imaging in these patients may reveal fibrosis of the gastrocnemius muscles (right panel, arrows), suggesting a continuum in the *LMNA* gene disorders between phenotypes with selective cardiac involvement and phenotypes with both cardiac and skeletal muscle abnormalities.

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Muscular Dystrophies – Crib Sheet

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Young man in wheelchair

DMD

Older man with walking difficulties

BMD

Conduction disease

Lamin A/C

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3) Sarcoidosis

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3/ Sarcoidosis

- Patterns of LGE
- Frequency of LGE
- Extracardiac findings in sarcoidosis

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Sarcoidosis

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Inflammatory

Granulomas

Any organ can be affected

- Lungs – pulmonary fibrosis
- Skin – erythema nodosum
- Lymph nodes – enlarged

Cardiac

- Has been under estimated in the past!
- Conduction failure
- Arrhythmias (VT)

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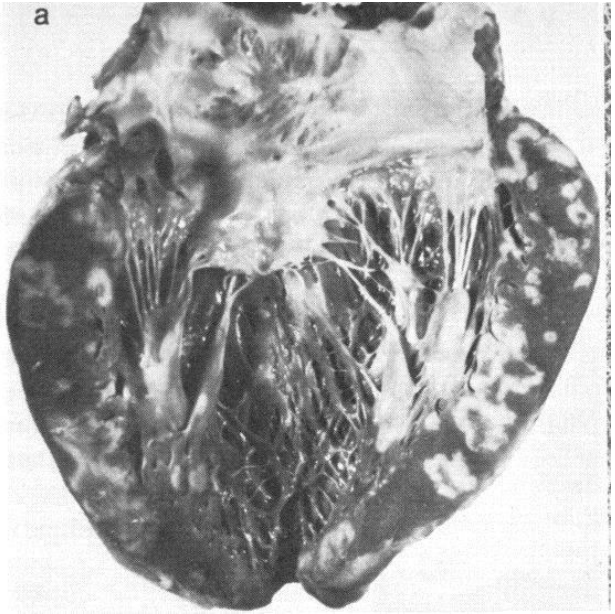
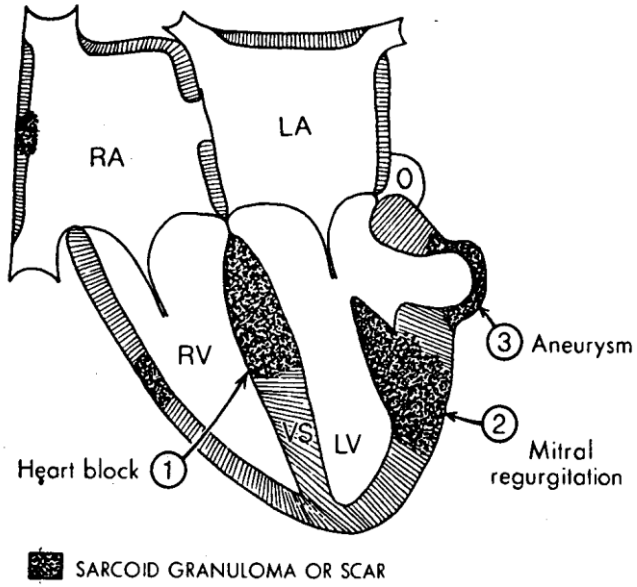
Sarcoidosis





Granulomatous Infiltration & fibrosis of the LV free wall and papillary muscle

Roberts et al. Am J Med 1977; 63: 86

Sarcoidosis – CMR findings

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LV dilatation and dysfunction

tion and dysfunction

is/late gadolinium enhancement

Pericardial

RV abnormalities

Is there lymphadenopathy

Did they give you lung image???

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Sarcoidosis

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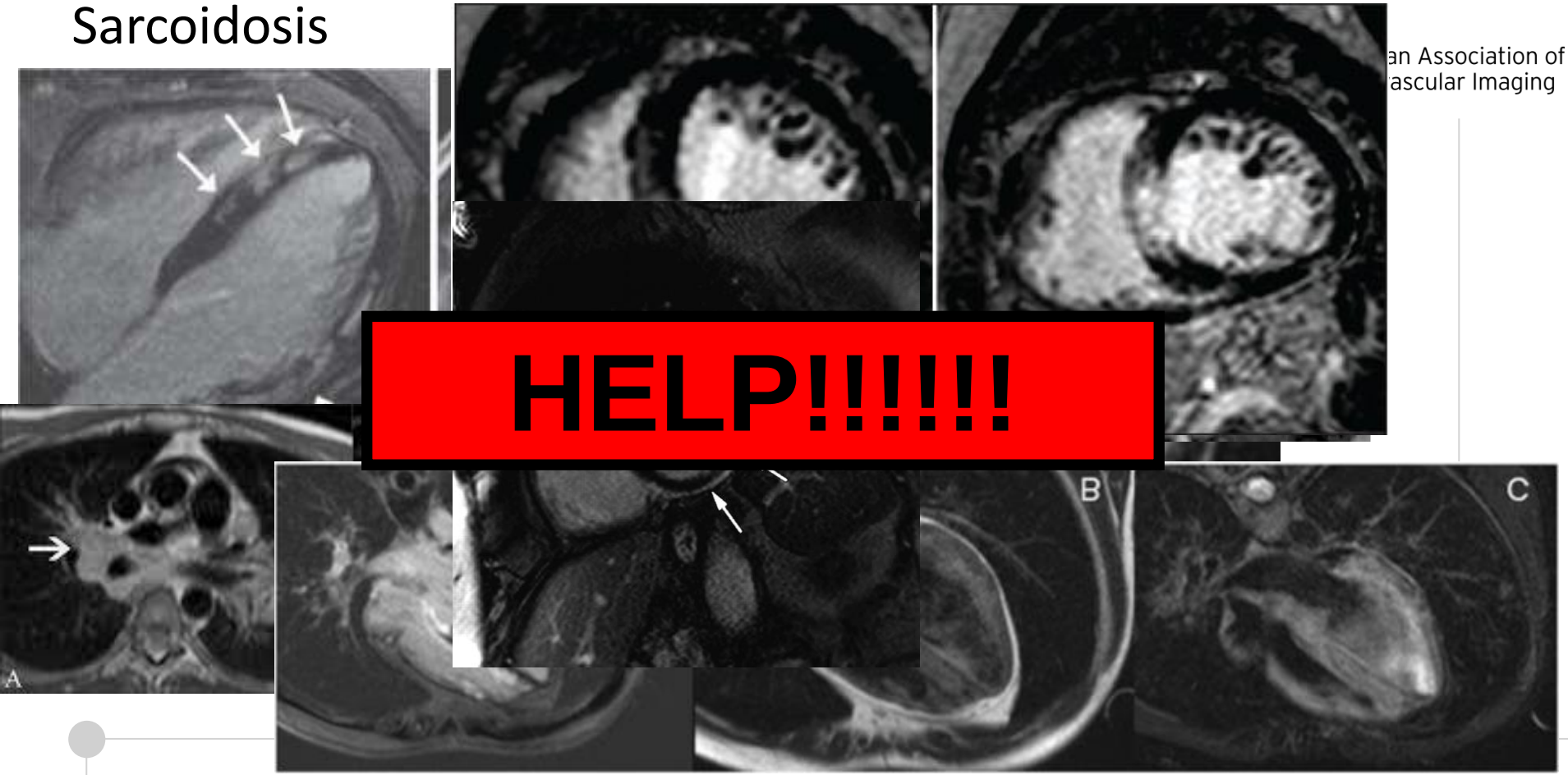

Sarcoidosis

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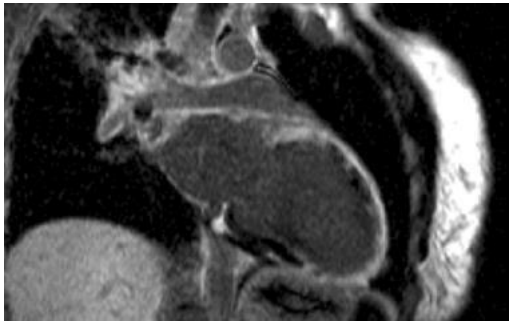

You must know this!

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Sarcoidosis

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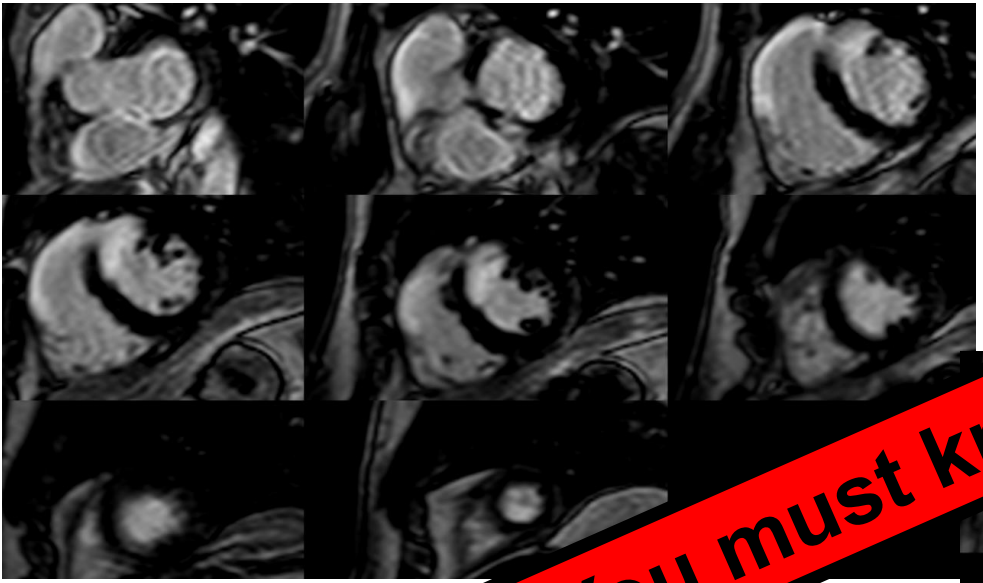
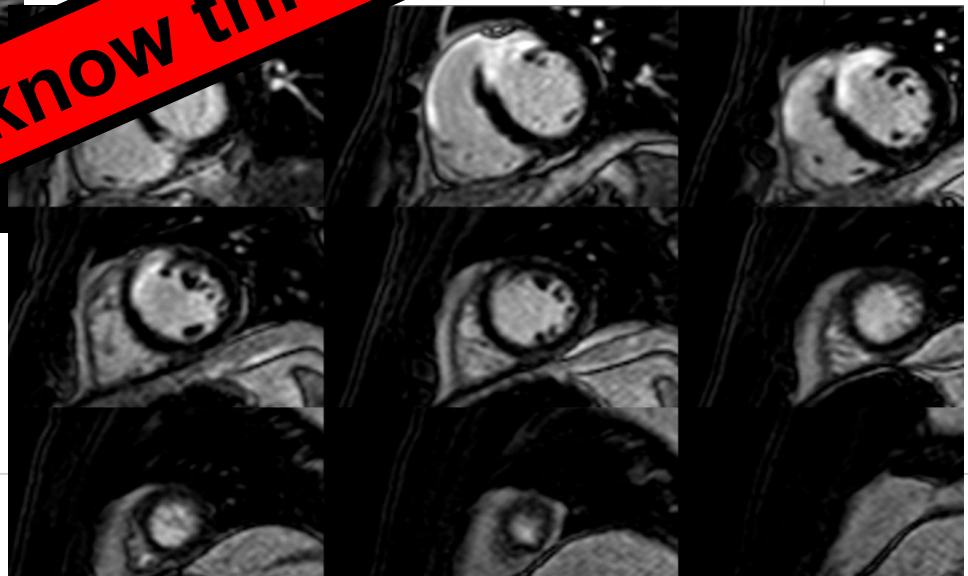


Initial Scan

You must know this!

9 months later



Sarcoidosis – Crib Sheet

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Lymphadenopathy

Lung images

Any old pattern of Late Gadolinium enhancement!

RV ‘Shepherd’s Crook’

PATHOGNONOMIC

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4) Amyloidosis

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4/ Amyloidosis

- LGE-pattern and contrast kinetics
- Typical cardiac morphology and function
- Pericardial and pleural effusions
- Related: Contrast administration in renal insufficiency
- Relative diagnostic yield of echo and CMR

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Amyloidosis

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There is more than 1 type

AL

Extracellular deposition immunoglobulin light chains

Abnormal fibrillar form

Median survivial 6-15 months

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Amyloidosis

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There is more than 1 type

TTR (transthyretin)

- Familial form
- More LVH early on
- Senile type (25% of those >80)

- More indolent course

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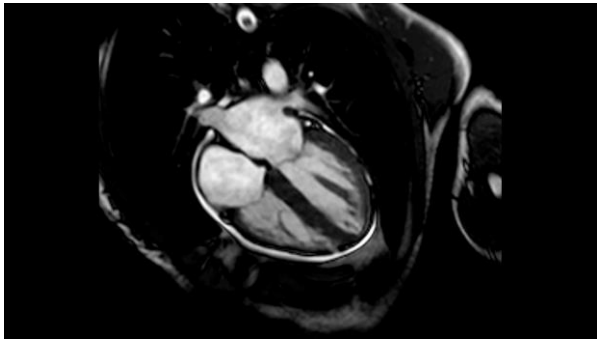
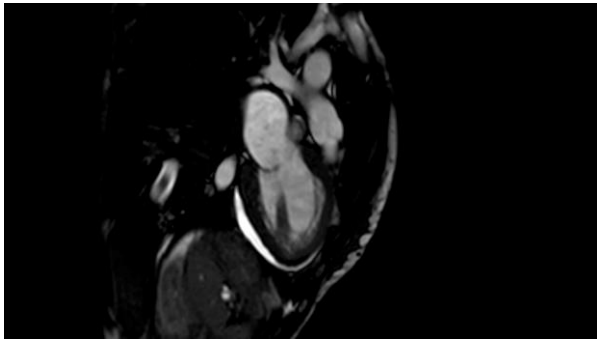
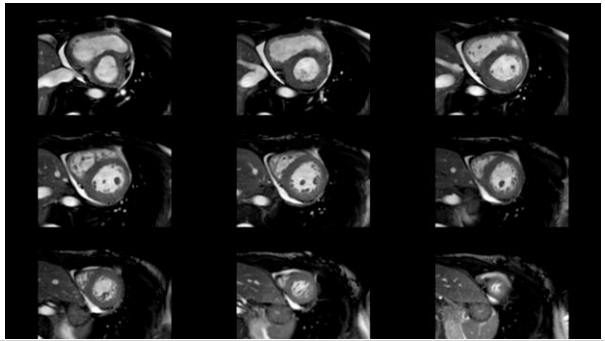
Amyloidosis







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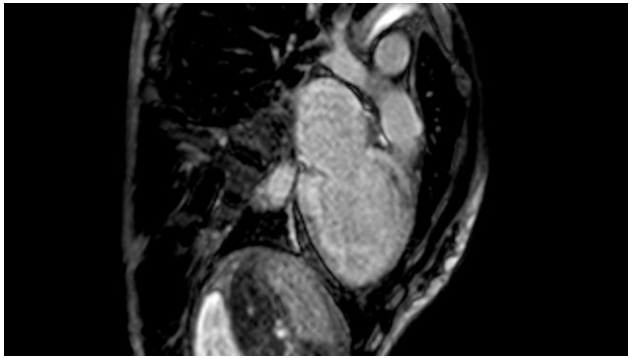

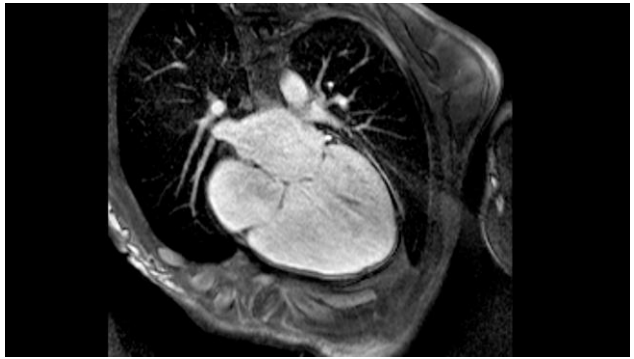

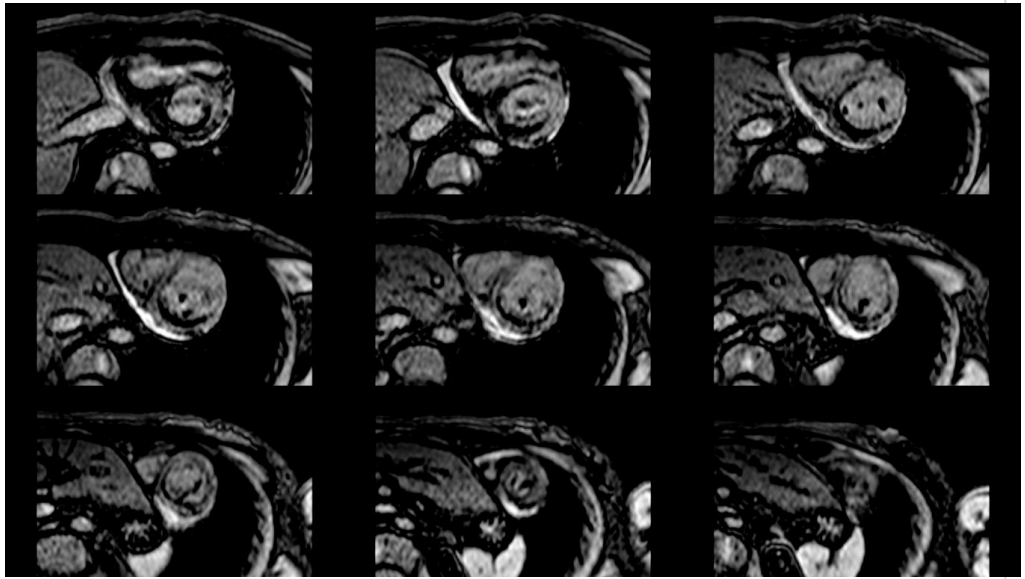

Amyloidosis

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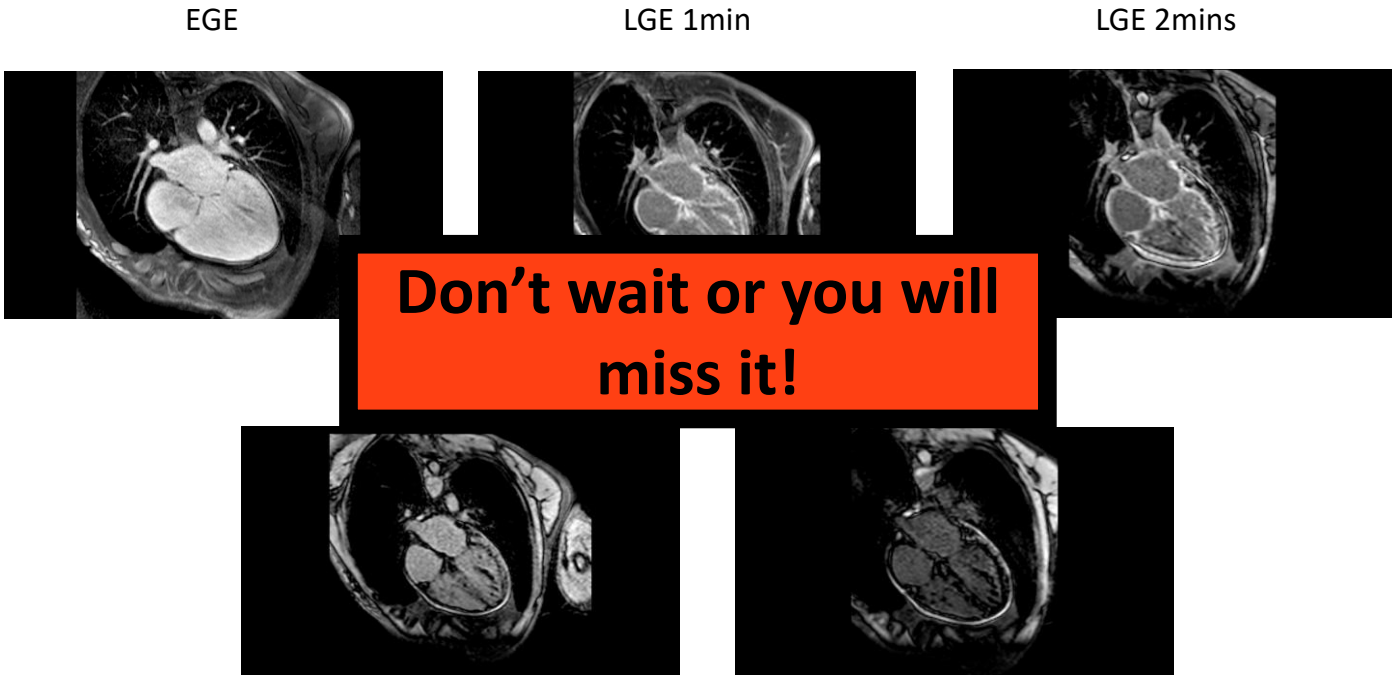

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Amyloidosis

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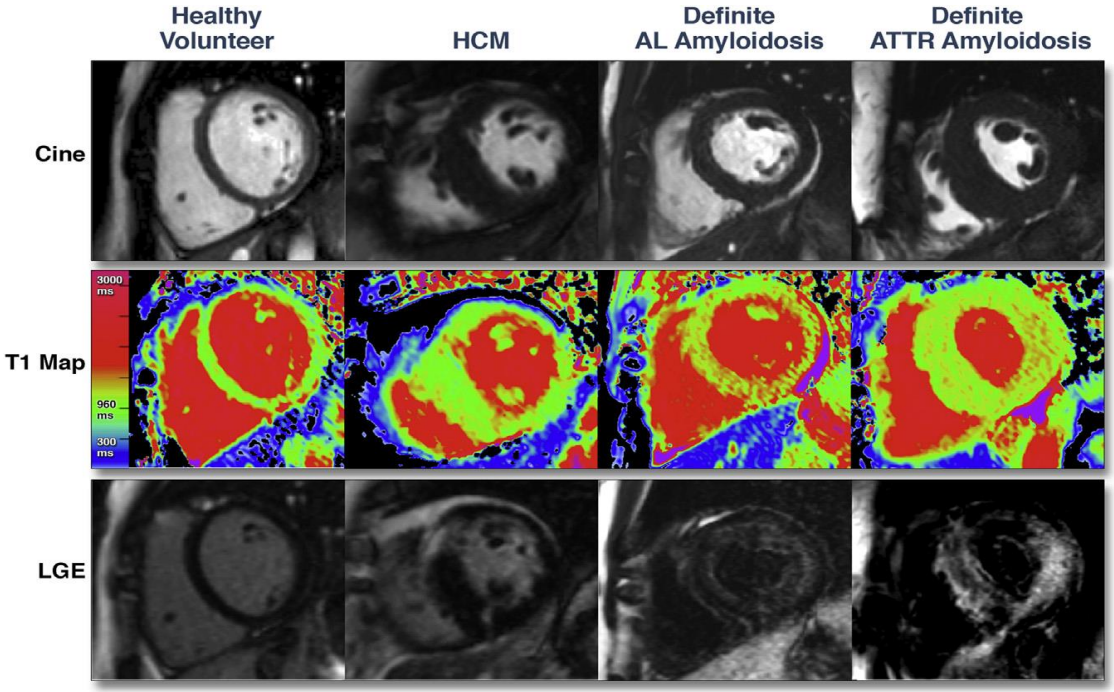

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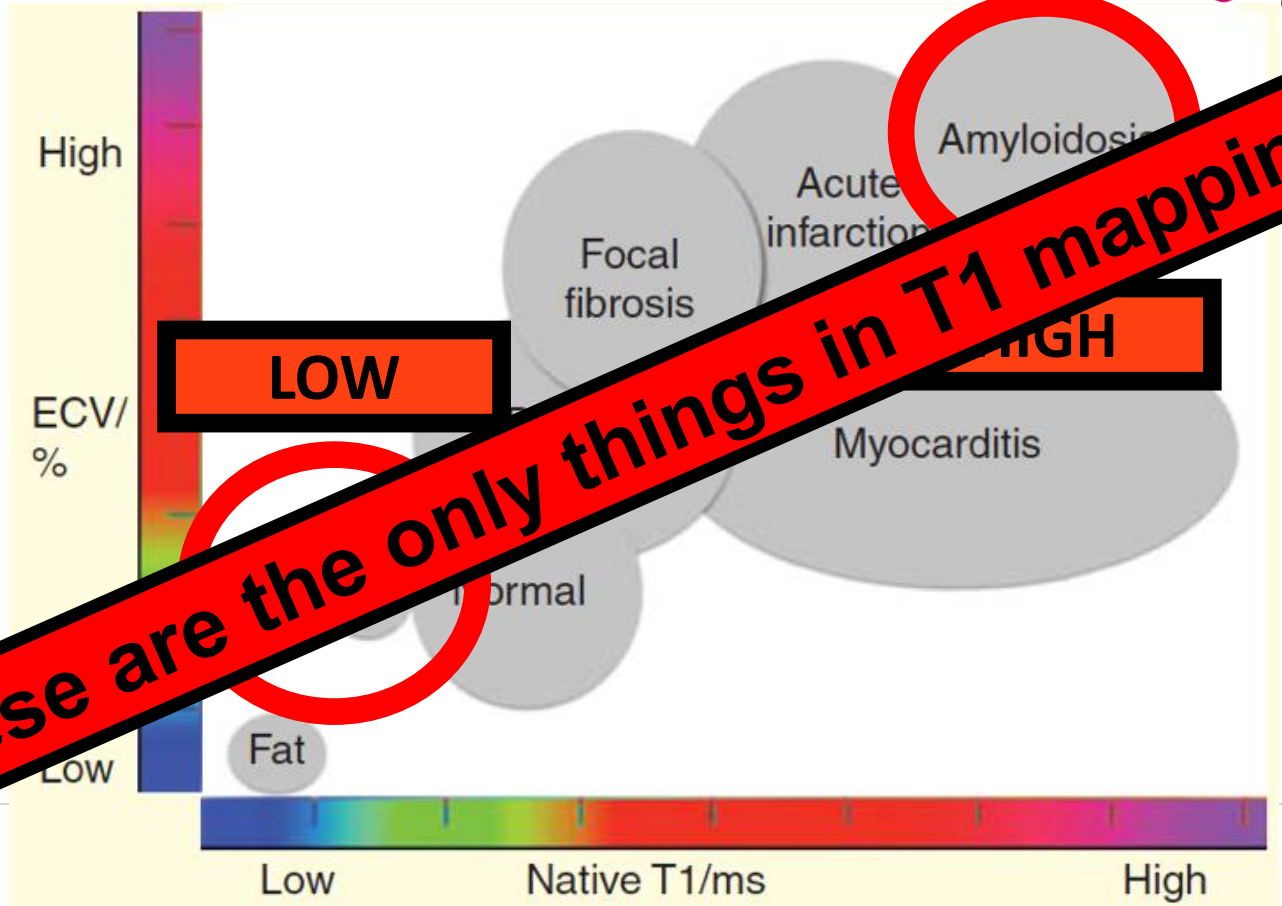

LOW

HIGH

These are the only things in T1 mapping!

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Amyloidosis – Crib Sheet

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LVH

Poor Long Axis Function

Big Atria

Pericardial/Pleural Effusions

Late Gadolinium Images Awful

TI Scout

T1 map

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5) Iron Overload Cardiomyopathies

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5/ Iron overload cardiomyopathies

- Concept and challenges of T2* measurement
- Location of myocardial T2* measurement
- Hepatic involvement

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Iron overload

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Mainly thalassemia

Haemachromatosis – rare cardiac iron loading

Hereditary anaemia of Beta Globin

Chronic anaemia

Transfusions

Death due to cardiac iron loading

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Iron overload – T2*

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Based on magnetic susceptibility

A measure of the exent to which a subst… agnetised when it is placed in an external magnet…

More Iron – Faster Signal Decay!!!!

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Iron overload – T2*

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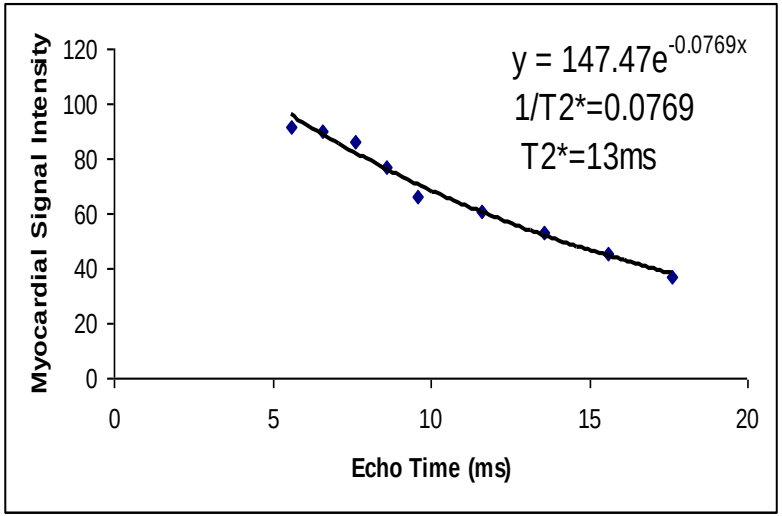

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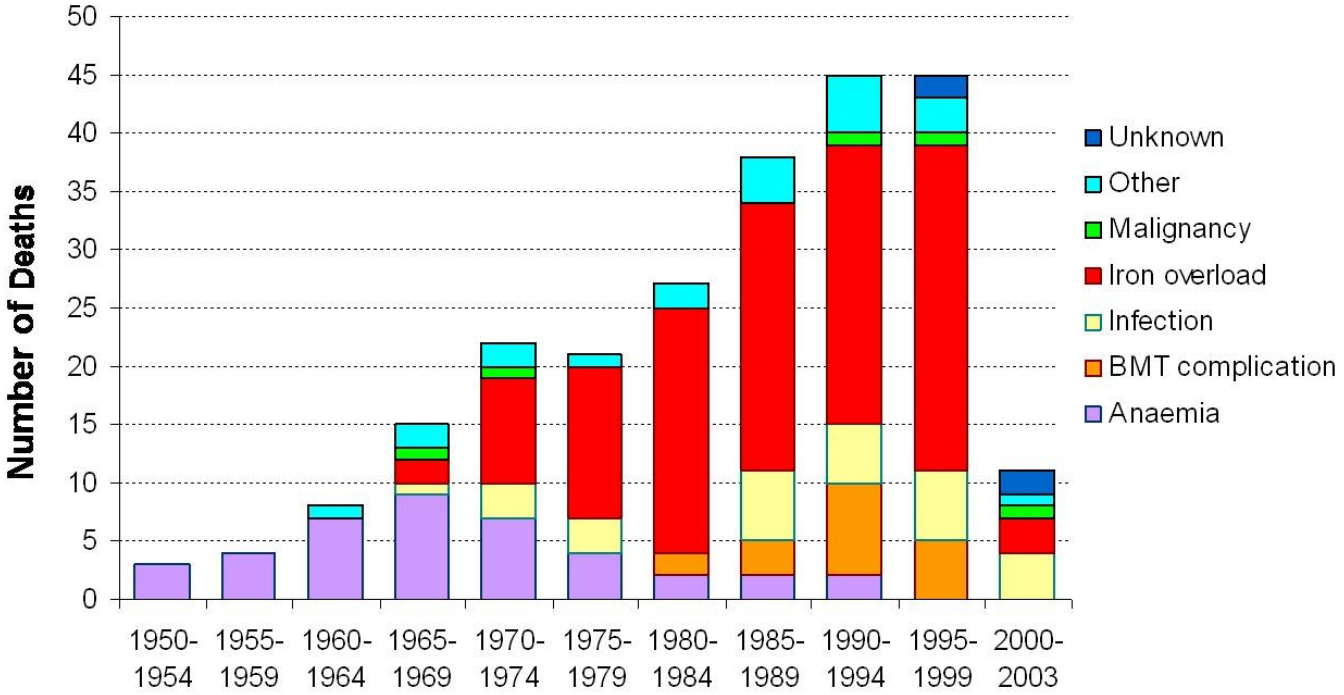
Iron overload – T2*



Modell et al. JCMR, 2008; 10: 42

Iron overload – T2* - liver and heart iron

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Iron loading – Crib Sheet

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Is there a T2* images – it is for iron

Are the images black?

There is no correlation between liver and heart iron

This technique saves lives

Normal >20ms

Severe <10ms

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6) Athlete's Heart

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6/ Athlete's heart

- Ways to differentiate athlete´s heart from cardiomyopathy
- Types of sports typically associated with cardiac changes

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Athlete's Heart

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Changes are well known:

Increased LV end diastolic volume

Increased LV stroke volume

EF is the same

This occurs for the RV

Gadolinium can occur but is pathological

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Athlete's Heart – Crib Sheet

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Clue will be in the history!

They will give you normal ranges as changes are

If pathological changes likely w

In Short – they won't ask this!

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7) Endomyocardial Fibrosis

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7/ Endomyocardial fibrosis

- Restrictive pathophysiology
- LGE
- RV involvement
- Thrombus formation

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Endomyocardial fibrosis

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Hypereosinophilic syndromes

- Persistent eosiniphilia
- Absence of primary cause (parasites, allergic disease)
- Evidence of eosinophil mediated end organ damage

Tropical endomyocardial fibrosis vs Hyereosinophilic syndrome

- Location (tropics for Tropical!)
- Cardiac wise identical

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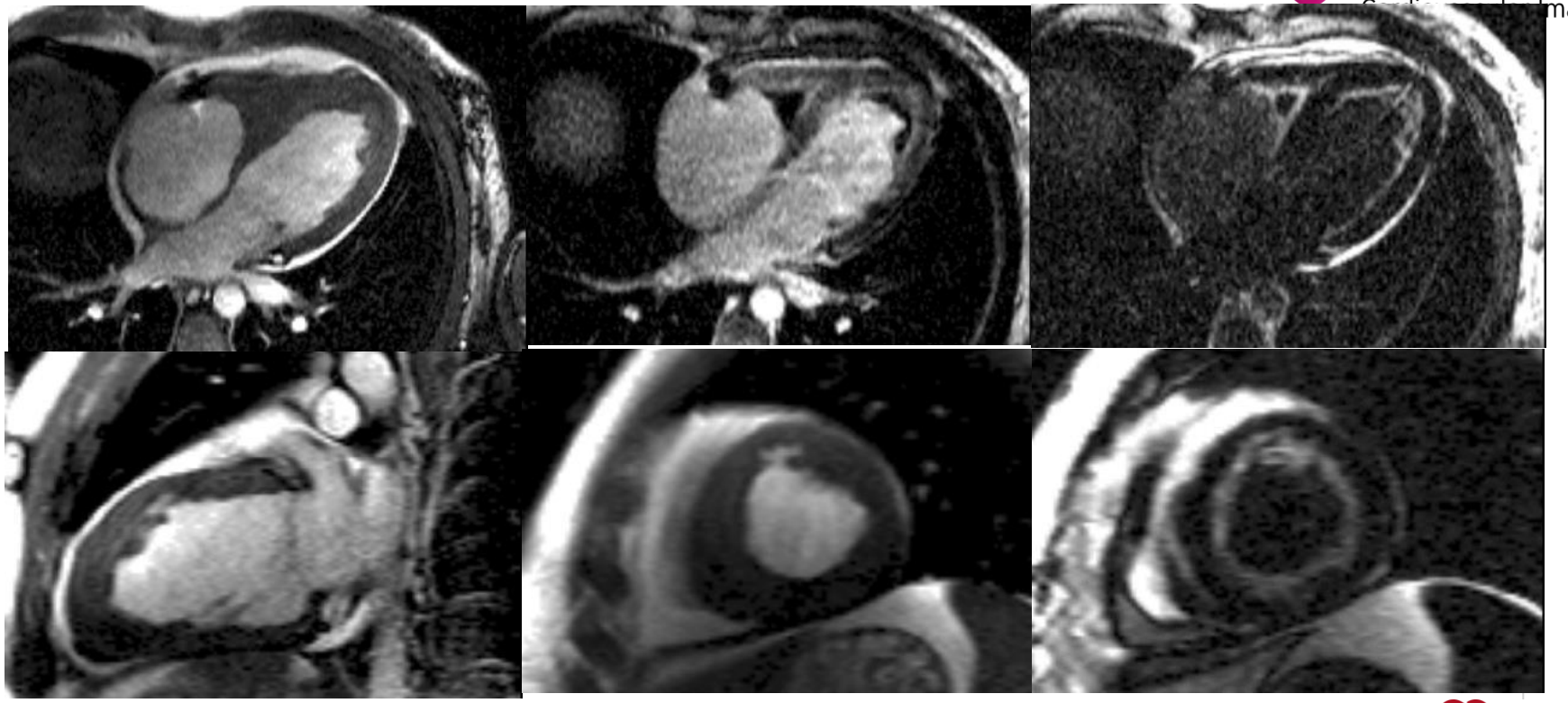
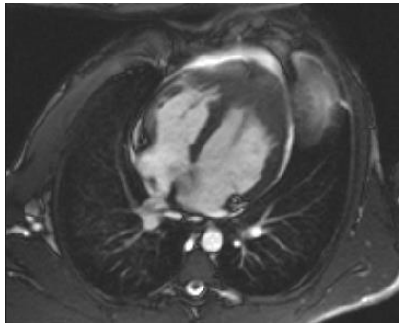
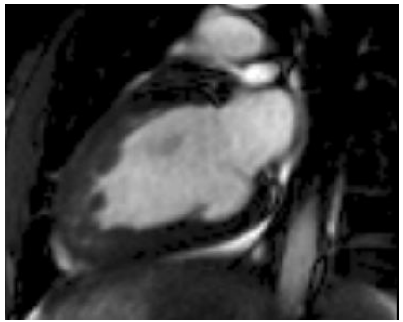
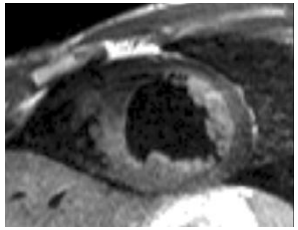
Hypereosninophilic syndromes

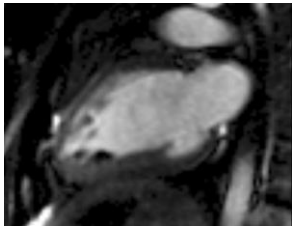


Hypereosninophilic syndromes









Hypereosinophilic syndromes – Crib Sheet

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‘Odd looking

Subendocardial apical late ga

with overlying

Churg-Strauss: Eosinophilia !!

What will be the differential?

8) Chagas Disease

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8/ Chagas disease

- Pathophysiology
- Morphology
- LGE
- Epidemiology
- Key elements of diagnosis

Torreão et al. JCMR 2015 ; 17: 97

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Chagas Disease

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Trypanosoma cruzi infection

- Cardiac involvment in 1/3rd of patients
- Main cause of death from heart failure in Latin America

Has phases

- Long asymptomatic (Indeterminate) phase
- Then unknown trigger leads to
  - Heart Failure
  - Ventricular tachycardia

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Chagas Disease

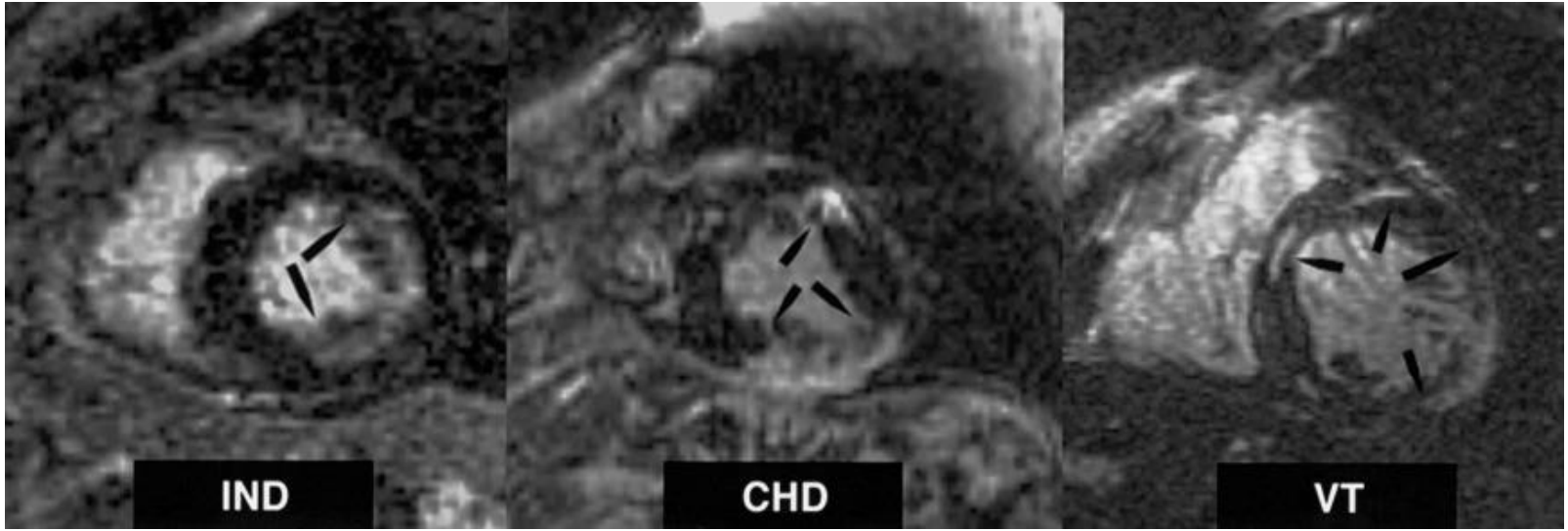

Figure 1. Myocardial delayed enhancement **(arrowheads)**on left ventricular short-axis slices in different stages of Chagas' disease. CHD = Chagas' heart disease group; IND = indeterminate phase group; VT = Chagas' heart disease with ventricular tachycardia group.

Chagas Disease

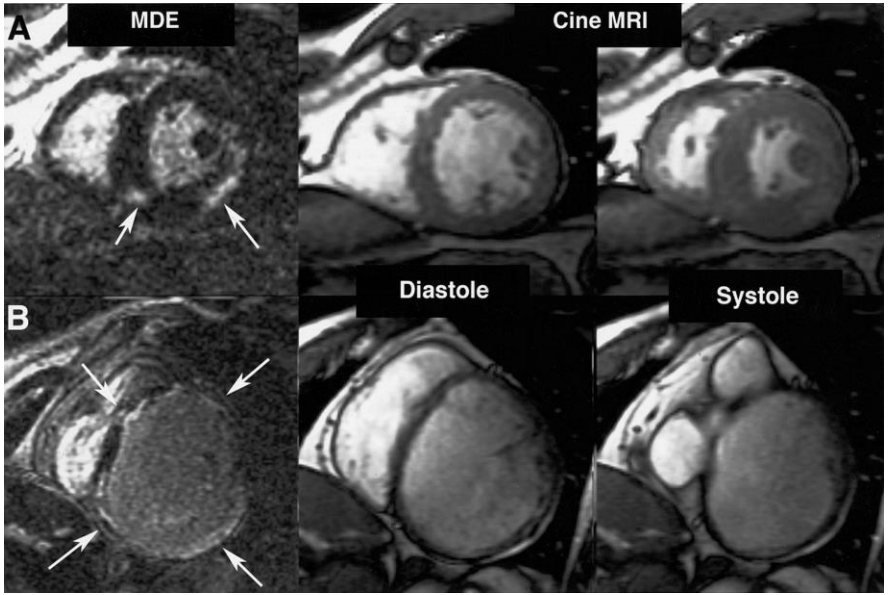

Figure 3. Extent of myocardial fibrosis (MF) **(arrows)**and left ventricular function. **(A)**Patient with small area of MF (8.2%) and normal left ventricular ejection fraction (65.5%). **(B)**Patient with large area of MF (23.8%) and severe left ventricular dysfunction (left ventricular ejection fraction 19.2%). MDE = myocardial delayed enhancement; MRI = magnetic resonance imaging.

Chagas Disease

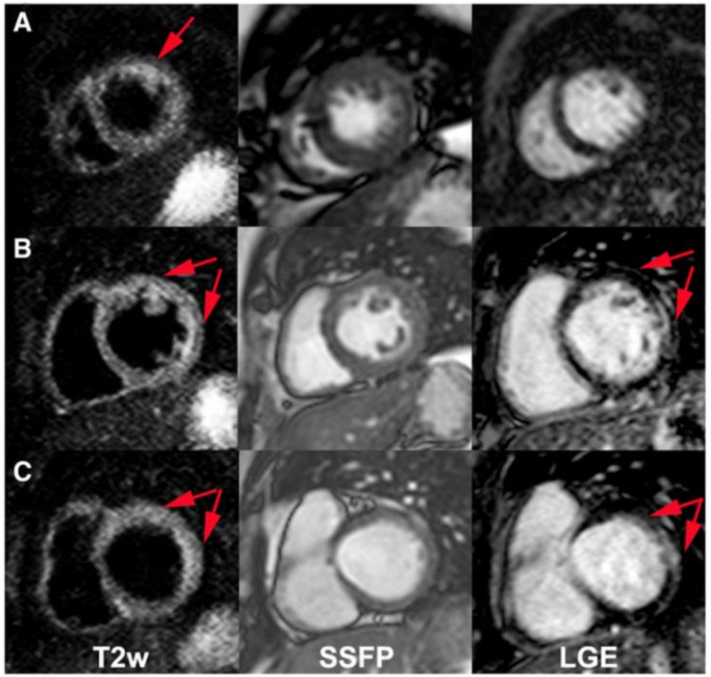

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Fig. 1
Apical (**a**), mid (**b**) and basal (**c**) short-axis images of a Chagas' heart disease patient in the indeterminate phase (patient 54, IND) with T2w (*left column*), cine SSFP for anatomical reference (*mid column*) and LGE (*right column*). *Red arrows* indicate increased myocardial signal intensity (T2W Ratio: 2.5). On the apical short-axis slice one can see a positive T2w image without correspondent LGE

Chagas Disease

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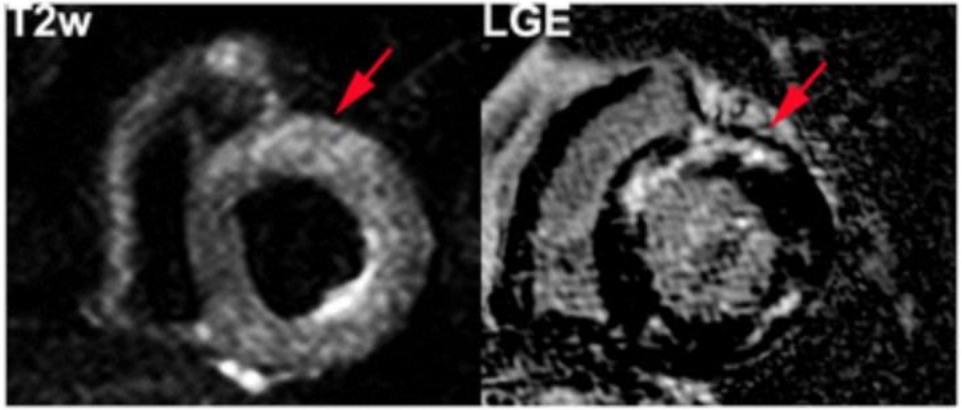

Fig. 2
Short-axis images of a Chagas' heart disease patient in the cardiac phase without LV dysfunction (patient 15, CPND). LGE (*right*) and T2 weighted (*left*) images with increased regional myocardial signal intensity on both techniques (T2W Ratio: 2.6)

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Chagas Disease

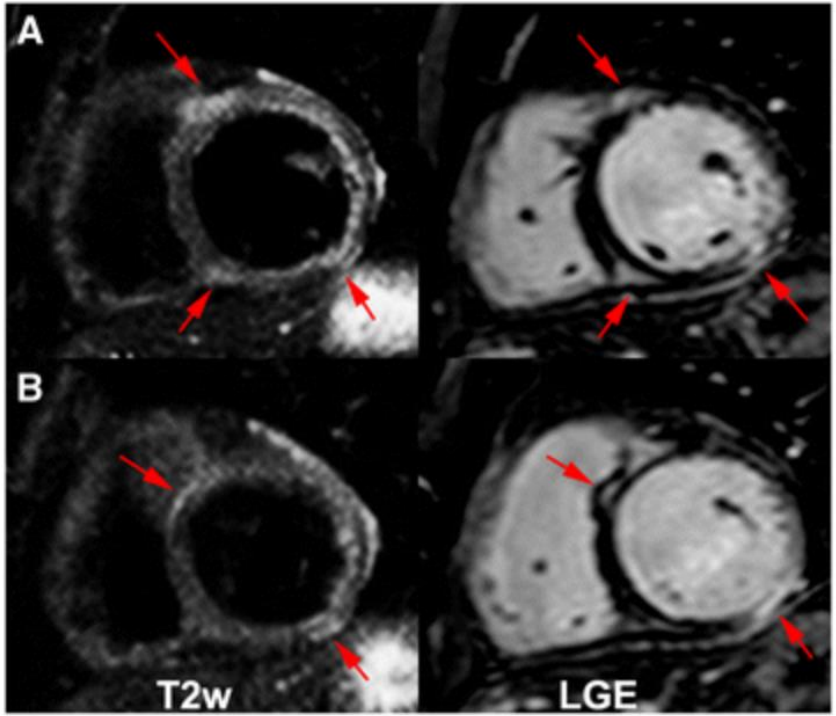

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Fig. 3
Mid (**a**) and basal (**b**) short-axis images of a Chagas' heart disease patient in the cardiac phase with LV dysfunction (patient 24, CPD). LGE (*right*) and T2 weighted (*left*) images with increased regional myocardial signal intensity on both techniques (T2W Ratio: 2.4)

Chagas Disease – Crib Sheet

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Something around South America

- Native
- Travel

Patches of focal fibrosis

Patches of oedema

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9) Fabry's Disease

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9/ Fabry´s disease

- LVH
- LGE, replacement fibrosis, spatial distribution
- Extracardiac findings
- Epidemiology
- Key elements of diagnosis

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Fabry's Disease

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Rare (1:40,000) inherited disorder

Lack of α-galactosidase A,

Leads to accumulation of Globotriaosylceramide(Gb-3)

>50% of patient have cardiac involvement

Leading cause of death

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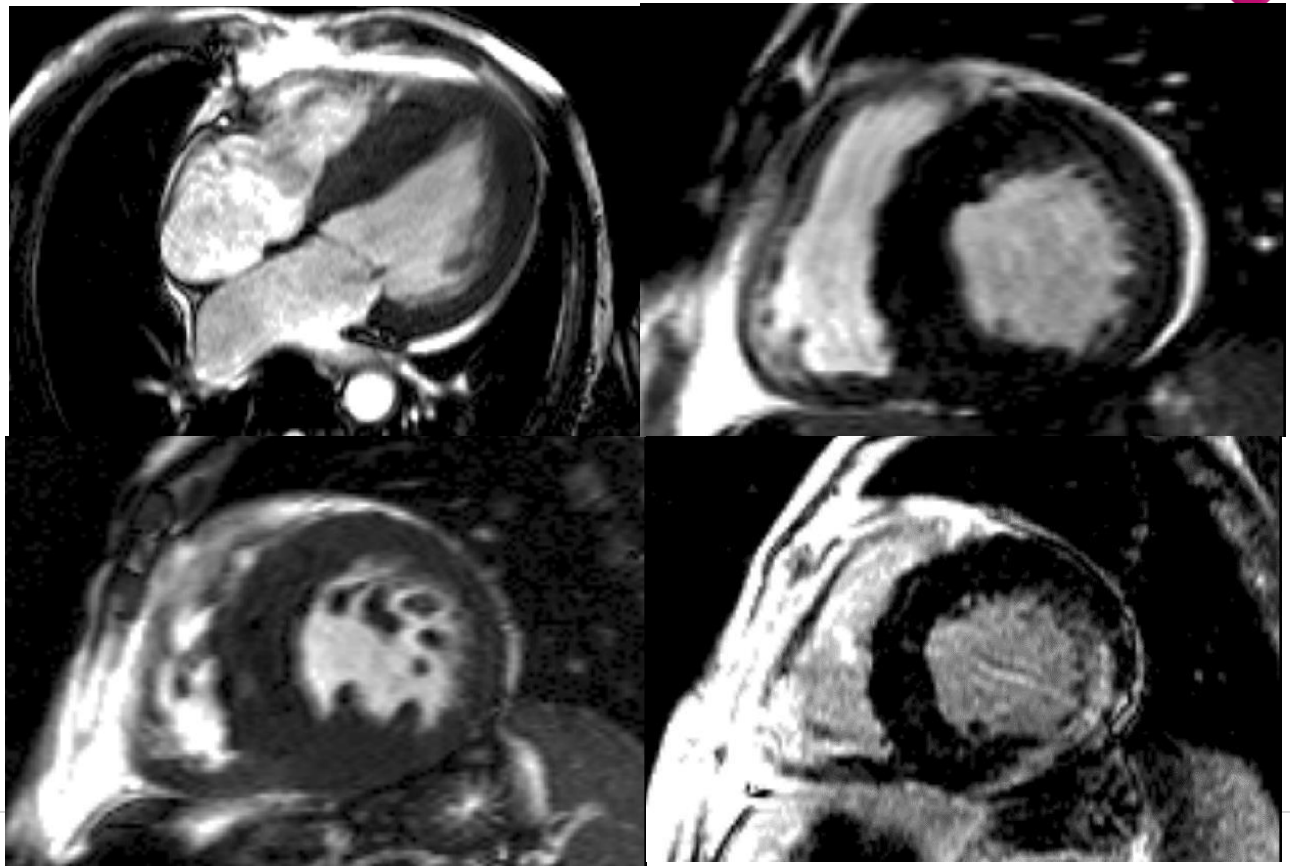
Fabry's Disease



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Fabry's Disease

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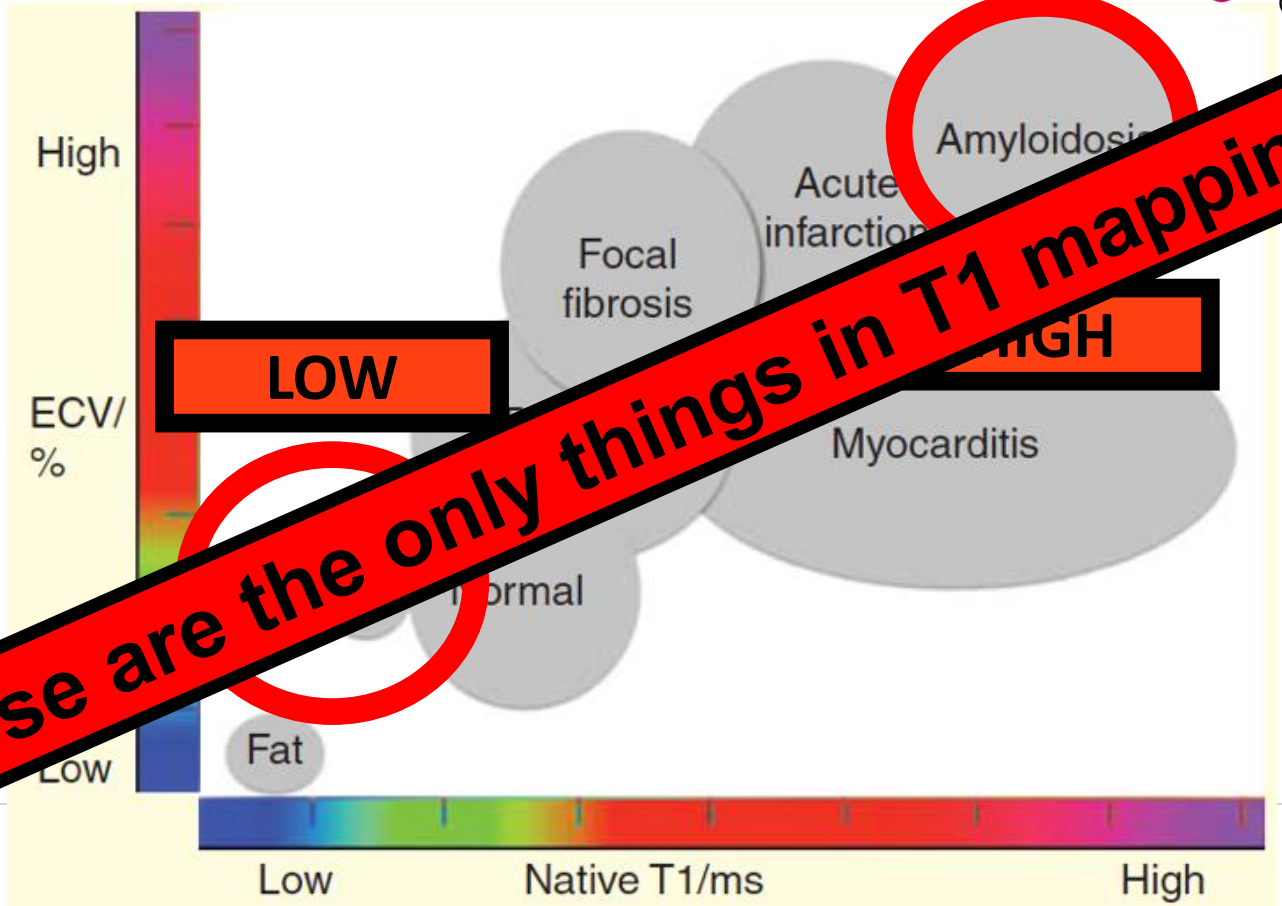

LOW

HIGH

These are the only things in T1 mapping!

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Fabry's Disease

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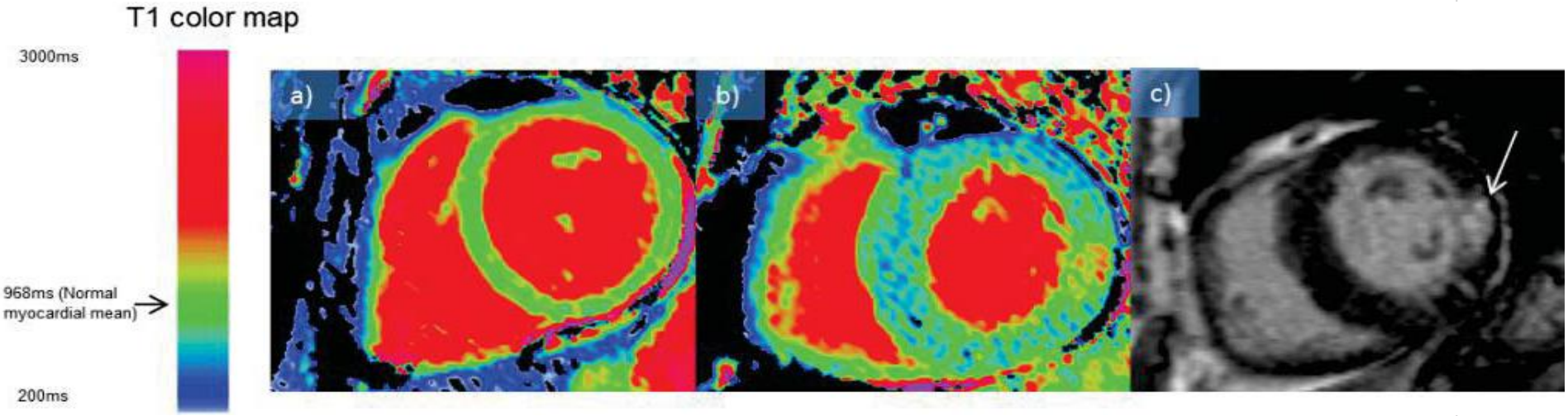

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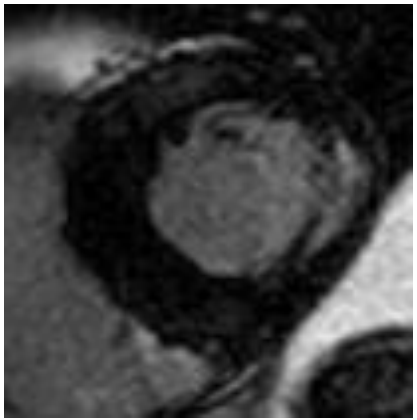
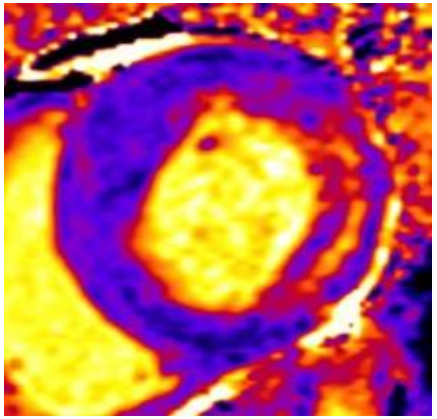


LGE



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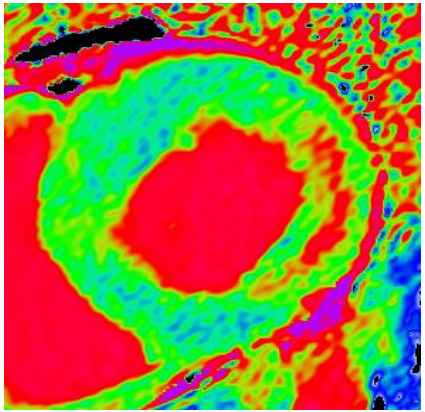

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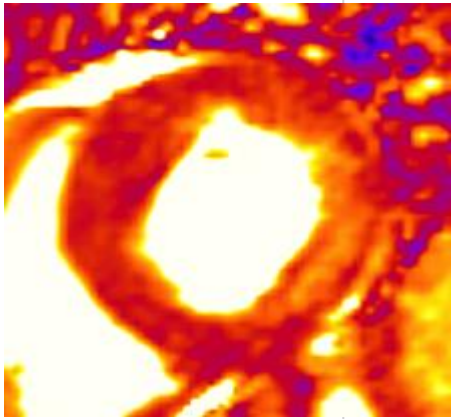

T2 MAP

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Fabry's Disease – Crib Sheet

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LVH

Looks like HCM

Late gadolinium in lateral wall

Though this is possible in HCM

T1 images

Rememb[illegible] of fibrosis T1 will be high

Make sure you can tell Fabry's from HCM!

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Conclusion

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1) Vasculitis

2) Muscular Dystrophies

3) Sarcoidosis

4) Amyloidosis

5) Iron Overload Cardiomyopathies

6) Athlete's Heart

7) Endomyocardial Fibrosis

8) Chagas Disease

9) Fabry's Disease

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