

AMSER Case of the Month

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33 year-old male with weakness

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Patient Presentation

- 33 year-old male with cardiomyopathy and weakness
- Medical history: withheld
- Surgical, Family and Social History: No family history of cardiac disease; otherwise noncontributory
- Physical Exam: Normal vital signs, lungs clear to auscultation, unremarkable cardiac exam
- Labs: No current significant labs
- Echo: Normal cardiac chamber size, normal ejection fraction, no valvular abnormalities

What Imaging Should We Order?

ACR Appropriateness Criteria: Suspected nonischemic cardiomyopathy. Initial imaging.

Variant 3. Suspected nonischemic dilated and unclassified cardiomyopathy. Ischemic cardiomyopathy already excluded. Initial imaging.

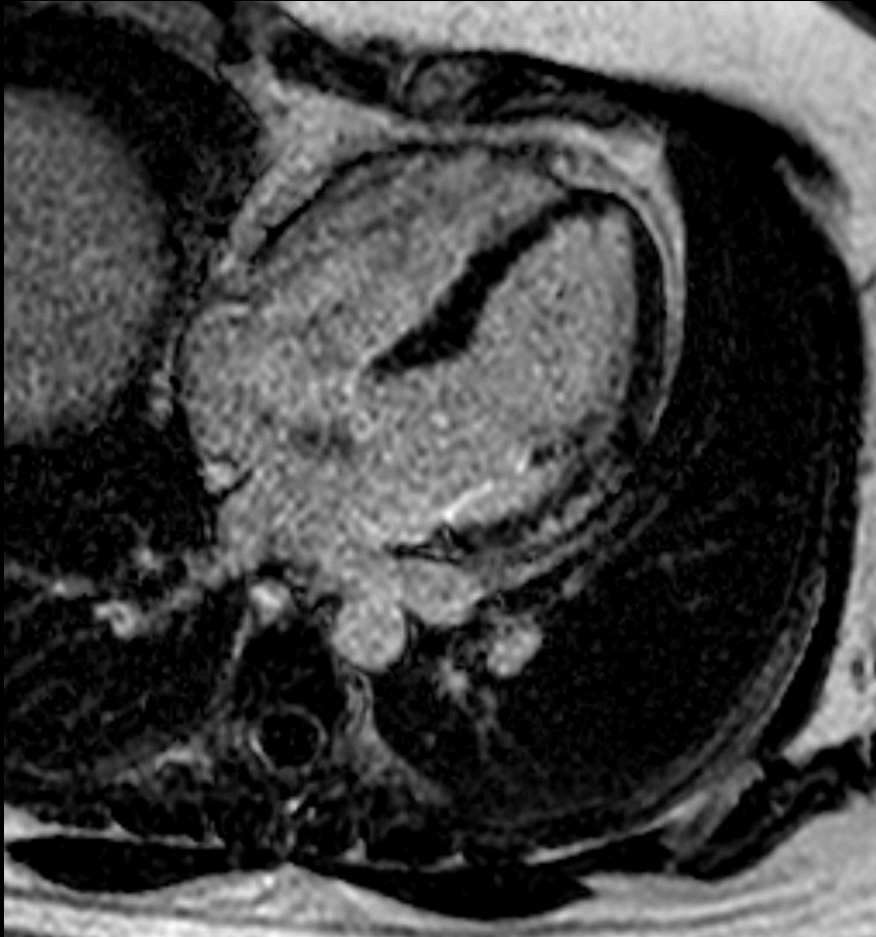
Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	0
MRI heart function and morphology without and with IV contrast	Usually Appropriate	0
MRI heart function and morphology without IV contrast	Usually Appropriate	0
CT heart function and morphology with IV contrast	May Be Appropriate	⊕⊕⊕⊕
CTA coronary arteries with IV contrast	Usually Not Appropriate	⊕⊕⊕
Arteriography coronary	Usually Not Appropriate	⊕⊕⊕
Arteriography coronary with ventriculography	Usually Not Appropriate	⊕⊕⊕
CT chest with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT chest without and with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT chest without IV contrast	Usually Not Appropriate	⊕⊕⊕
CT coronary calcium	Usually Not Appropriate	⊕⊕⊕
FDG-PET/CT heart	Usually Not Appropriate	⊕⊕⊕⊕
MRI chest without and with IV contrast	Usually Not Appropriate	0
MRI heart with function and inotropic stress without and with IV contrast	Usually Not Appropriate	0
MRI heart with function and inotropic stress without IV contrast	Usually Not Appropriate	0
MRI heart with function and vasodilator stress perfusion without and with IV contrast	Usually Not Appropriate	0
US echocardiography transesophageal	Usually Not Appropriate	0
US echocardiography transthoracic stress	Usually Not Appropriate	0
MRI chest without IV contrast	Usually Not Appropriate	0

Ordered by cardiologist for workup

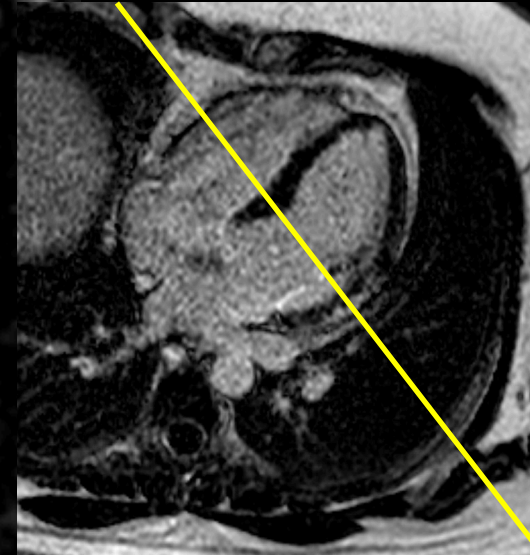
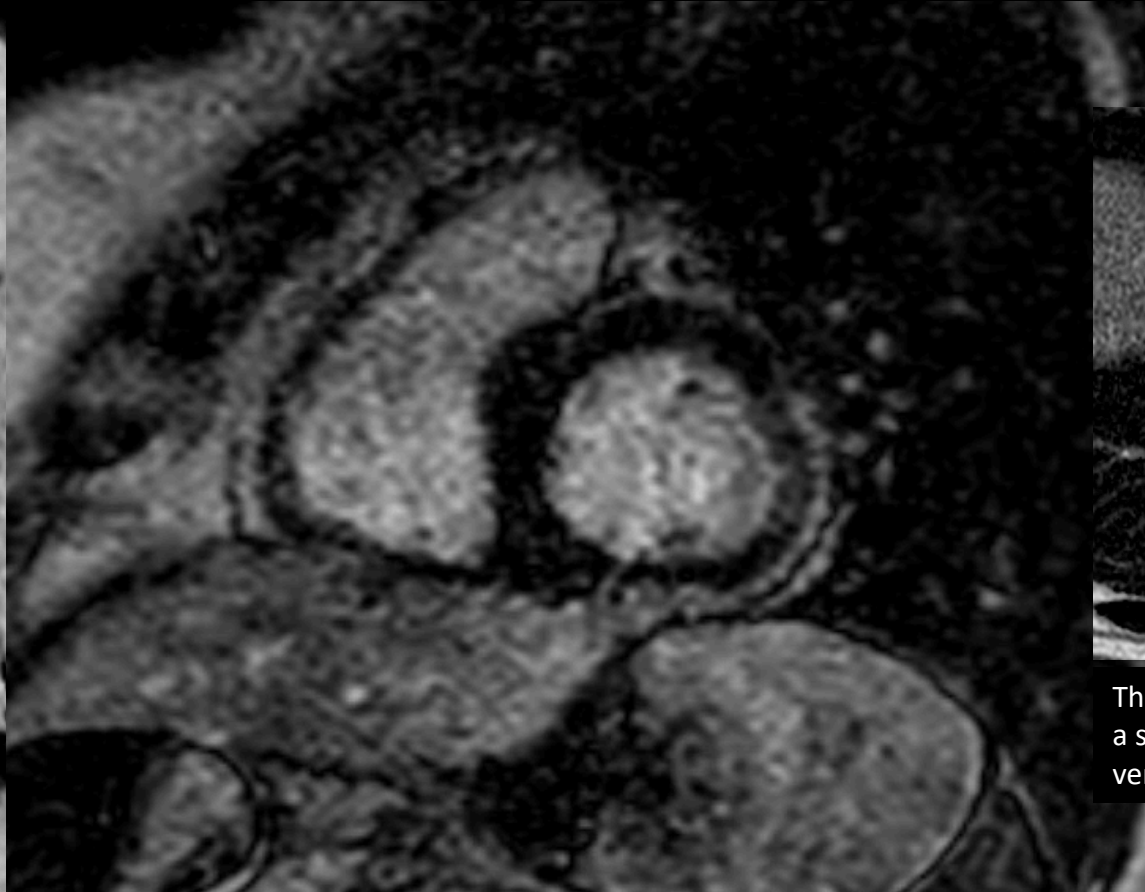


Findings (unlabeled)

4 Chamber



Short Axis

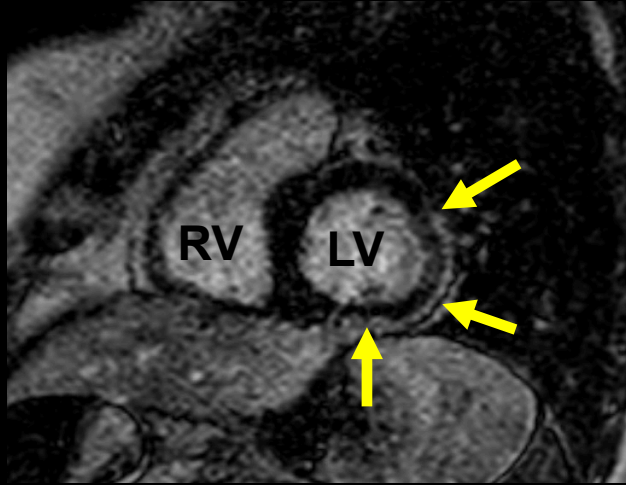


The yellow line is a reference for how a short axis slice through the ventricles is obtained.

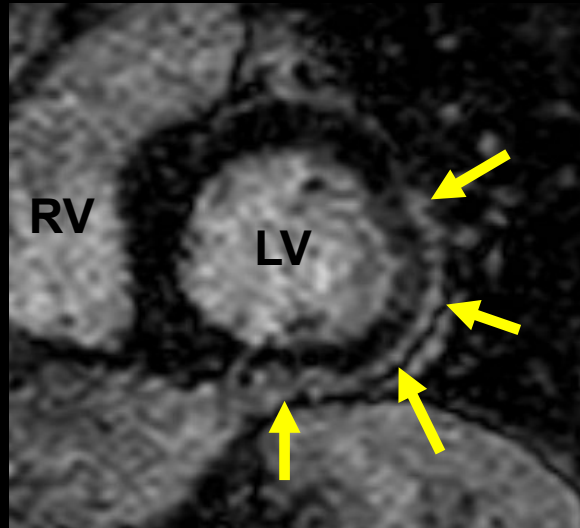
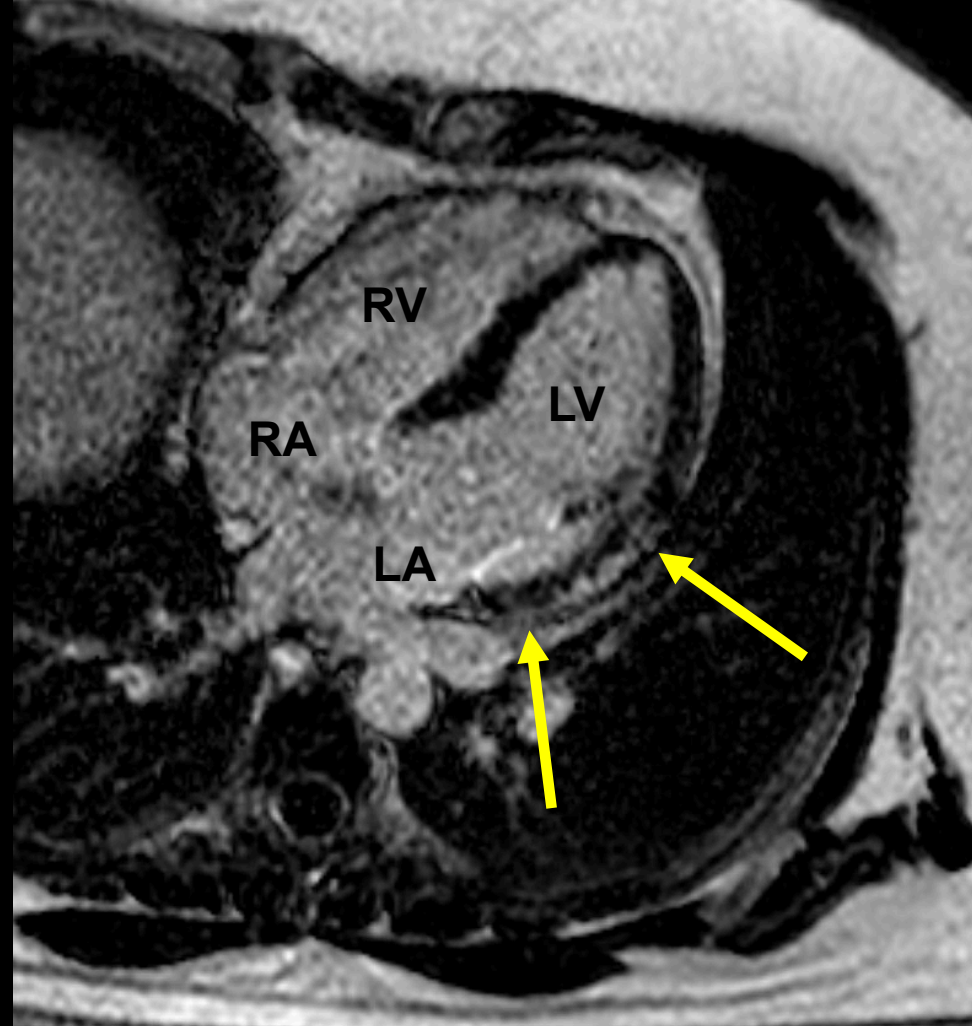
Post contrast phase sensitive inversion recovery/late gadolinium enhancement imaging

Findings (labeled)

Short Axis



4 Chamber



ADDITIONAL CLUE TO DIAGNOSIS: What do you think about the size of the chest wall muscles?

Post contrast inversion recovery: Late gadolinium enhancement of the sub-epicardium of the anterolateral to inferior walls (arrows).

Final Dx:

Cardiomyopathy related to Becker Muscular
Dystrophy

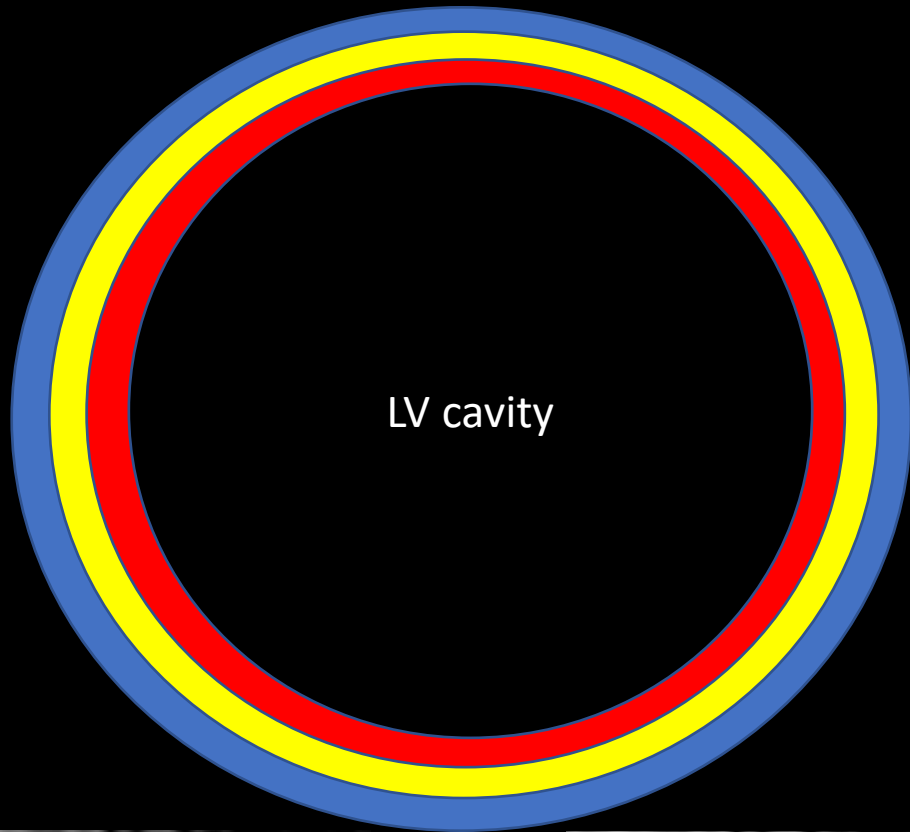
Diagnosis: Becker Muscular Dystrophy

- Epidemiology: ~0.26 per 10000 individuals. Less common, less severe than Duchenne muscular dystrophy.
- Etiology: X-linked recessive mutation in dystrophin gene
 - *Dystrophin*: protein in muscle fiber cell membrane. Loss of dystrophin causes decreased cell membrane integrity and degeneration of myofibers.
- Typical clinical presentation: Onset typically ages 2-11
 - Common symptoms: Difficulty climbing stairs/walking, myalgias, exercise intolerance due to progressive muscular degeneration
 - Progression: Falls, **cardiomyopathy**, respiratory weakness
 - Prognosis: Death typically occurs by age 40-50
 - Most common cause of death: **Cardiomyopathy or arrhythmias**

Why Choose Cardiac MR to Evaluate Cardiomyopathy in Becker Muscular Dystrophy?

- **Cardiac MR** allows for myocardial tissue characterization and includes **late gadolinium enhancement imaging**
 - Contrast is retained in the myocardium when fibrosis, necrosis, edema or infiltrative substances are present
 - Gold standard for quantification of function and assessment for fibrosis
 - The presence of fibrosis is a poor prognostic findings in cardiomyopathy and can be seen with a normal ejection fraction
 - Becker patients with enhancement, even when ejection fraction is preserved, may be treated with cardioprotective medications to slow progression of cardiomyopathy
- Late gadolinium enhancement in Becker has most commonly been described as subepicardial or midmyocardial and involving the lateral wall

Describing Enhancement on Cardiac MR



The myocardium of the left ventricle can be divided into 3 layer

Subepicardium

Midmyocardium

Subendocardium

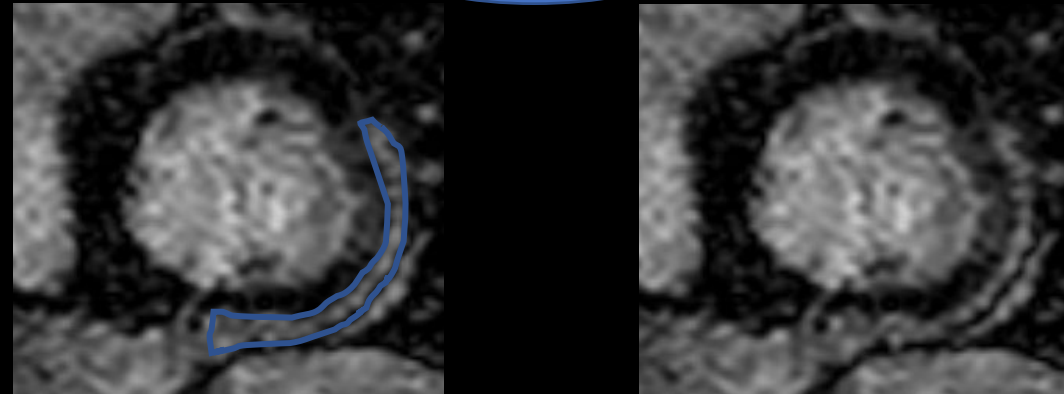
The distribution of enhancement can allow for diagnosis and prognosis. For example...

Subepicardium: Becker, Duchene, Myocarditis

Midmyocardium: Hypertrophic cardiomyopathy

Subendocardium: Ischemia/Infarct, Amyloid

Transmural (all 3 layers): Multiple causes



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