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.

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

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

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


