



May 2011

MRI as a Lifesaving Tool in Hypertrophic Cardiomyopathy

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Outline

- Patient presentation
- Overview of hypertrophic cardiomyopathy (HCM)
 - clinical manifestations
 - histology
 - complications
- Echocardiographic criteria for diagnosis
- Advantages of cardiac MRI in evaluating HCM



Our patient: presentation

- 42M presents after
 - 10 minutes of chest pain that resolved with rest
 - Syncope without warning symptoms
- No significant PMH
- Meds
 - Vicodin PRN back pain

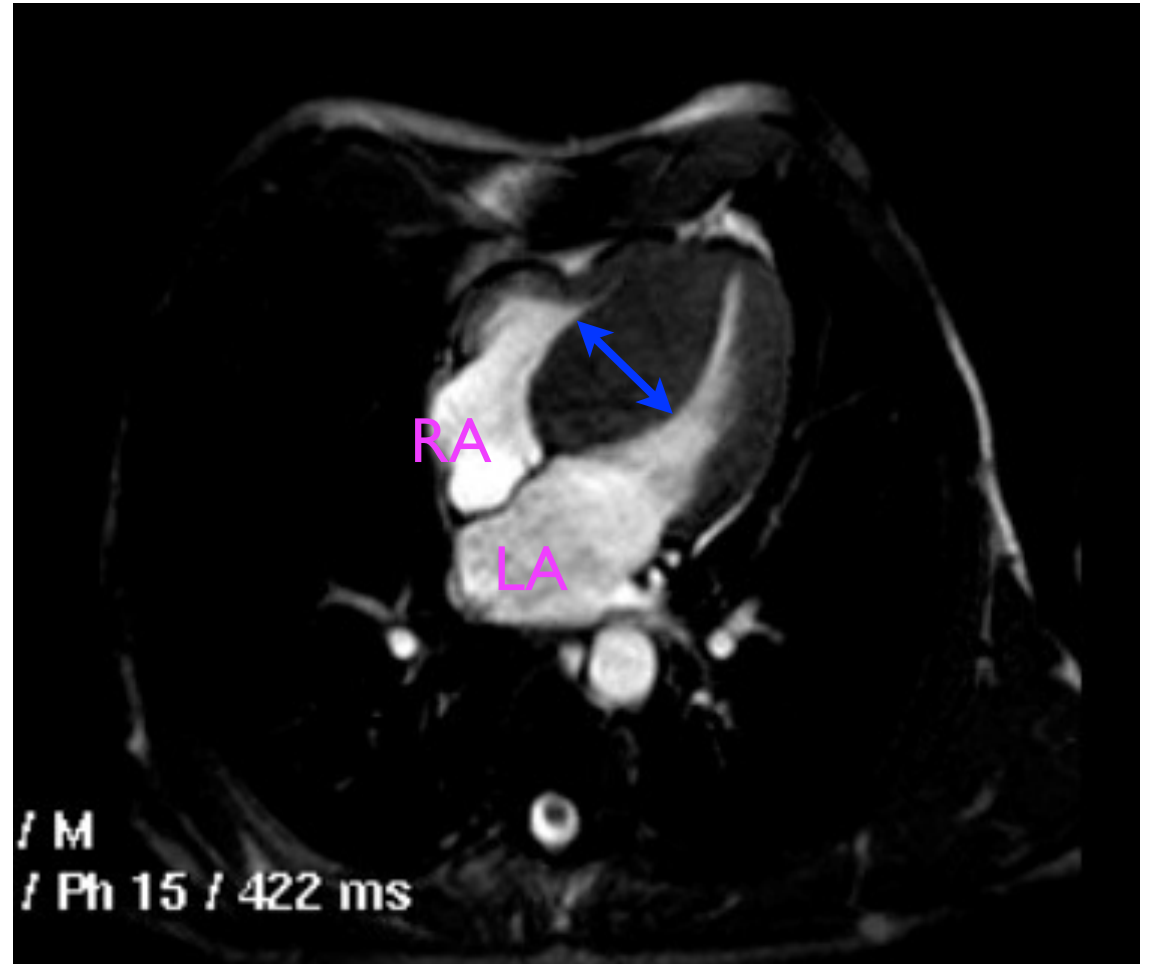


Our patient: evaluation

- ECG reveals left ventricular hypertrophy
- Stress echo reveals 2.5cm septal thickness

Our patient: HCM on cardiac MR

Bright-blood CMR
shows **markedly
thickened septum** to
2.5cm

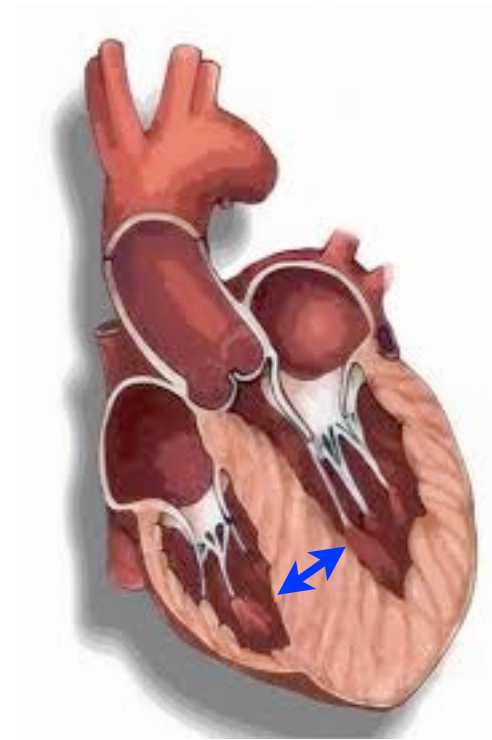


axial, bright blood, C-,T2 weighted cardiac MRI
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Hypertrophic cardiomyopathy: overview

- HCM manifests as severe **LV wall thickening**, generally involving the interventricular septum. Patients may be asymptomatic or may present with a number of cardiac complications, discussed below.
- It is autosomal dominant and many potential causative genes have been identified. Many of these are components of the actin-myosin complex, though there is some thought that connective tissue genes may be involved as well.

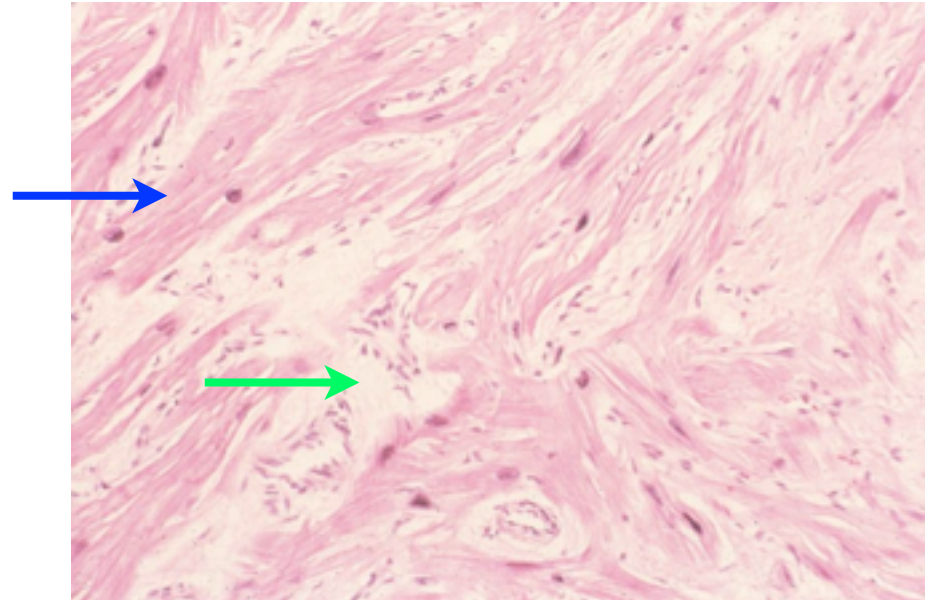


<http://www.metrohealth.org/body.cfm?id=1509&oTopID=C>



Hypertrophic cardiomyopathy: histology

- The myocardium is made up of **hypertrophied myocytes** arranged in a **disordered pattern** and is infiltrated with **fibrotic tissue**.

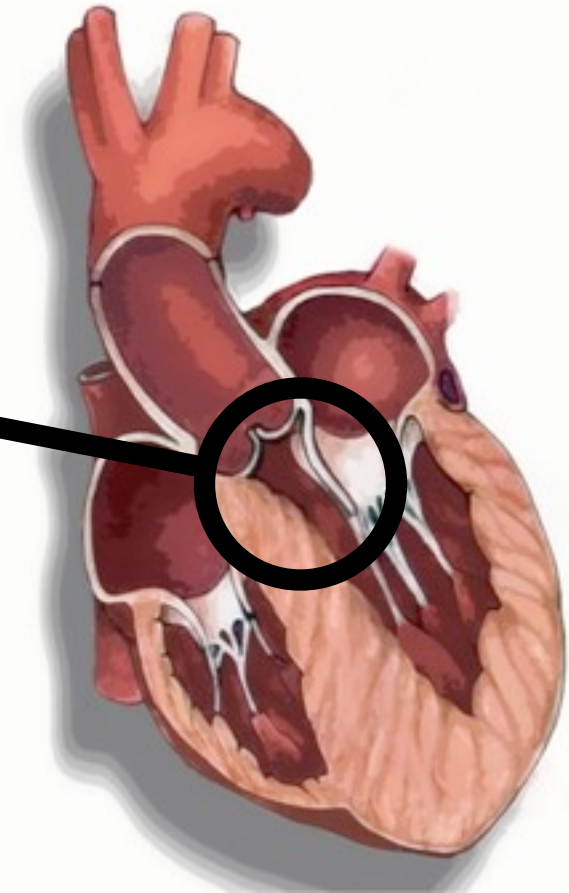


Adnan MM. <http://medicinembbs.blogspot.com/2011/03/hypertrophic-obstructive-cardiomyopathy.html>



Complications: LV outflow tract

The thickened interventricular septum reduces the cross-sectional area of the LV outflow tract. This reduces cardiac output and can lead to symptoms of fatigue, dyspnea, and syncope.

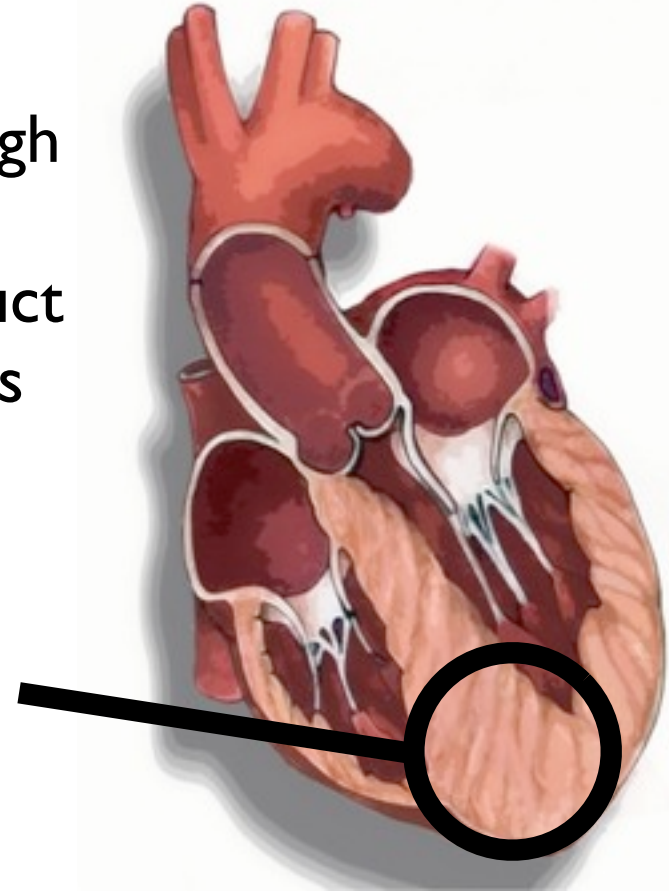


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Complications: myocardium

The hypertrophied myocardium has a high oxygen demand, leading to angina with minimal exertion. It also does not conduct electricity properly, predisposing patients to life-threatening arrhythmias like ventricular tachycardia or ventricular fibrillation. Patients who die from complications of HCM generally die of fatal arrhythmias.

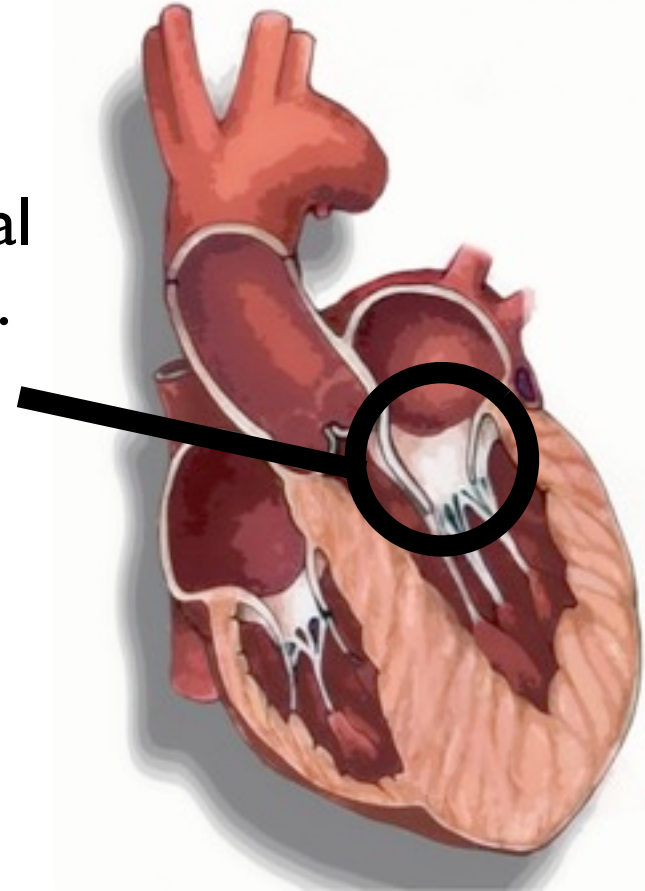


<http://www.metrohealth.org/body.cfm?id=1509&oTopID=C>



Complications: mitral valve

Blood rushing through the narrow LV outflow tract causes a pressure gradient that draws the anterior leaflet of the mitral valve towards the interventricular septum. This is known as systolic anterior motion of the mitral valve (SAM). SAM causes mitral regurgitation, leading to left atrial dilation and pulmonary edema.



<http://www.metrohealth.org/body.cfm?id=1509&oTopID=C>



Echocardiographic diagnostic criteria

- Echocardiography is the gold-standard first line diagnostic test. Major and minor diagnostic criteria have been established. Diagnosis requires 1 major or 2 minor criteria

Major	Minor
LV wall thickness $> 13\text{mm}$ severe systolic anterior motion	LV wall thickness $> 12\text{mm}$ moderate systolic anterior motion redundant mitral valve leaflets

McKenna WJ, P Spirito, M Desnos, et al. Experience from clinical genetics in hypertrophic cardiomyopathy: proposal for new diagnostic criteria in adult members of affected families. *Heart*. 1997; 77(2): 130-132



Utility of cardiac MRI

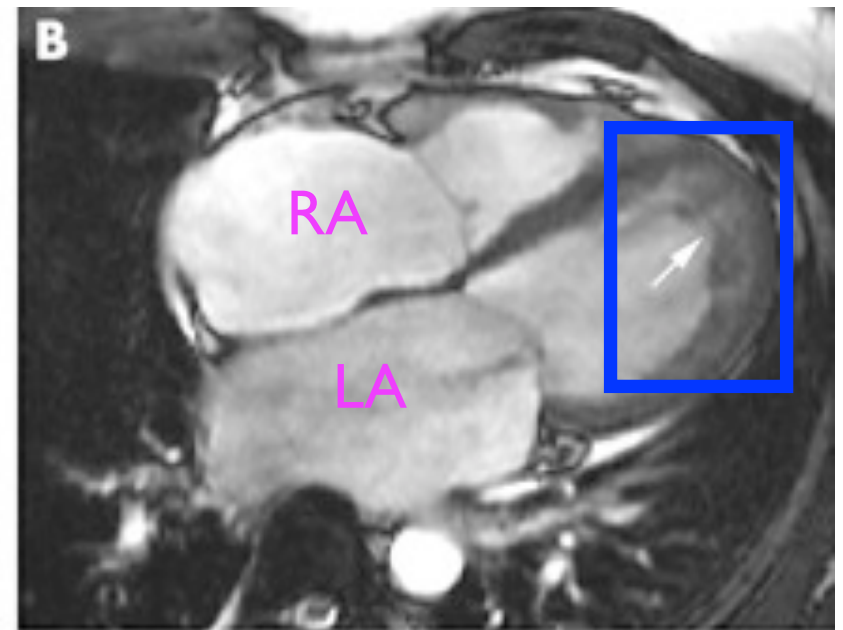
- While echocardiography is the gold-standard imaging modality to diagnose HCM, cardiac MR offers several advantages. Continue to see examples of information that can be obtained only with cardiac MR.



Companion Patient #1: Apical HCM on MRI

There is a rare form of HCM that primarily involves the LV apex. The LV apex is not well visualized with standard echocardiography. Because cardiac MR is a cross-sectional imaging modality with good spatial resolution, it can image the LV apex well.

In this example, we see a patient with HCM presenting only with **apical myocardial hypertrophy**.



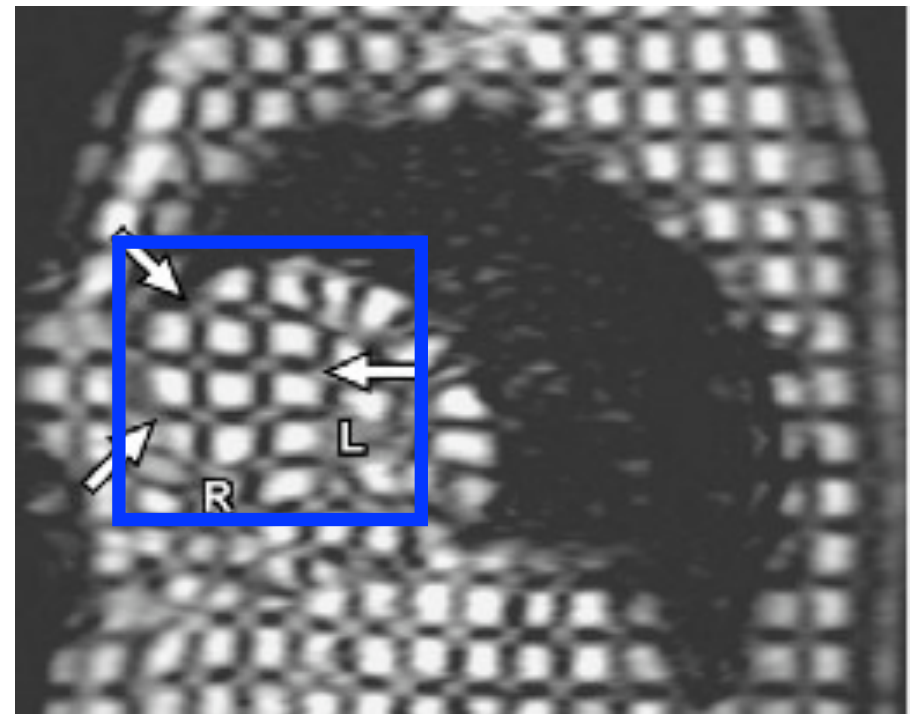
axial, bright-blood, C- cardiac MR

Varghese A, NG Fisher, DJ Pennell. Late recognition of left ventricular non-compaction by cardiovascular magnetic resonance. *Heart*. 2005; 91(3): 282

Companion patient #2: akinesia on MRI

- Small perturbations in the magnetic field can disrupt proton spins in a grid-like distribution. The protons affected will relax more slowly and appear dark on the resulting image.
- This grid can be set up during diastole and will persist through the cardiac cycle
- The grid will deform in areas that are contracting and will remain unchanged in akinetic regions of myocardium.

A region of **akinesia** is identified by the undeformed gridlines surrounded by deformed gridlines



Dillman JR, GC Mueller, AK Attili, et al. Case 153: Atypical Tumefactive Hypertrophic Cardiomyopathy. *Radiology*. 2010; 254(1): 310-313



Other benefits of cardiac MR

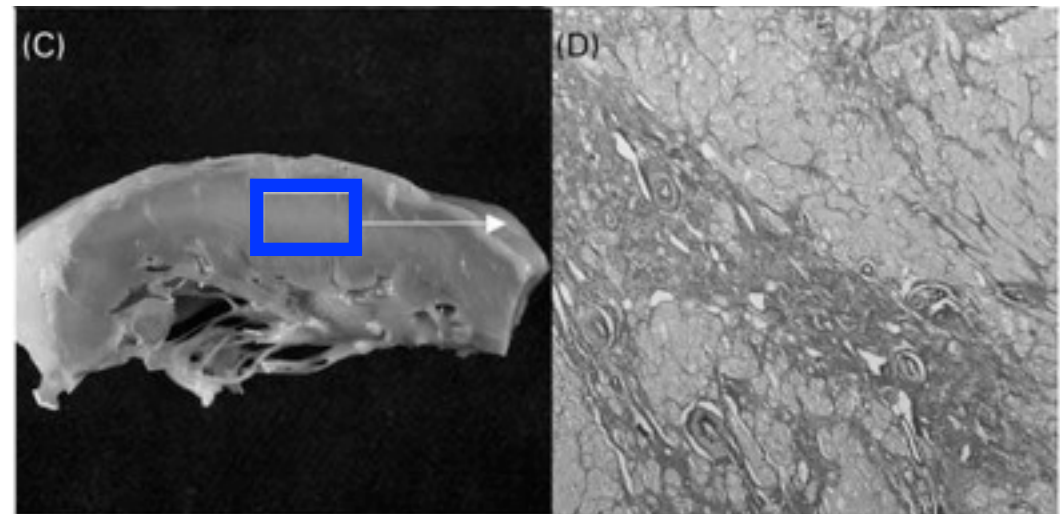
- Quantify SAM
- Follow up septal resection/ablation
- Identify areas of myocardial fibrosis

Pennell DJ, UP Sechtem, CB Higgins, et al. Clinical indications for cardiovascular magnetic resonance (CMR): Consensus Panel report. *European Heart Journal*. 2004; 25(21): 1940-1965

Rikcers C, NM Wilke, M Jerosch-Herold, et al. Utility of cardiac magnetic resonance imaging in the diagnosis of hypertrophic cardiomyopathy. *Circulation*. 2005; 112(6): 855-861

Myocardial fibrosis: pathology

- Along with pathological hypertrophy, HCM patients have myocardium that is infiltrated by **scar**
- People with HCM have up to 8 times more myocardial collagen than normal controls
- Scar tissue and fibrosis leads to arrhythmias and sudden cardiac death



Papavassiliu T, P Schnabel, M Schroder, et al. CMR scarring in a patient with hypertrophic cardiomyopathy correlates well with histological findings of fibrosis. *European Heart Journal*. 2005; 26(22): 2395-2395

Shirani J, R Pick, WC Roberts, et al. Morphology and significance of the left ventricular collagen network in young patients with hypertrophic cardiomyopathy and sudden cardiac death. *J Am Coll Cardiol*. 2000; 35(1): 36-44



Can imaging help identify myocardial scar? (Yes!)



Kinetics of extracellular fluid in scar tissue

- Scar tissue is non-viable and has lower capillary density compared to surrounding tissue
- Extracellular fluid is slow to enter scar tissue because of this decreased capillary density
- Extracellular fluid is also slow to leave areas of scar



Delayed gadolinium enhancement (DGE)

- Gadolinium is a marker for extracellular fluid in MRI studies
- Based on kinetics of extracellular fluid in scar tissue, gadolinium should be slower to wash-in and slower to wash-out from areas of fibrosis
- To find areas of DGE, image 10 minutes after gadolinium infusion. Areas that enhance should correspond to areas of scar

Kim R, E Chen, J Lima, et al. Myocardial Gd-DTPA kinetics determine MRI contrast enhancement and reflect the extent and severity of myocardial injury after acute reperfused infarction. *Circulation*. 1996; 94: 3318-3326



Predicting complications with DGE

- Not every HCM patient has areas with DGE
- Patients with DGE are more likely to
 - test positive on current genetic screens
 - have ventricular ectopy
 - have non-sustained ventricular tachycardia
 - require ICD implantation
 - have sudden cardiac death

Rubinshtein R, JF Glockner, SR Ommen, et al. Characteristics and clinical significance of late gadolinium enhancement by contrast-enhanced magnetic resonance imaging in patients with hypertrophic cardiomyopathy. *Circulation: Heart Failure*. 2010 3(1): 51-58

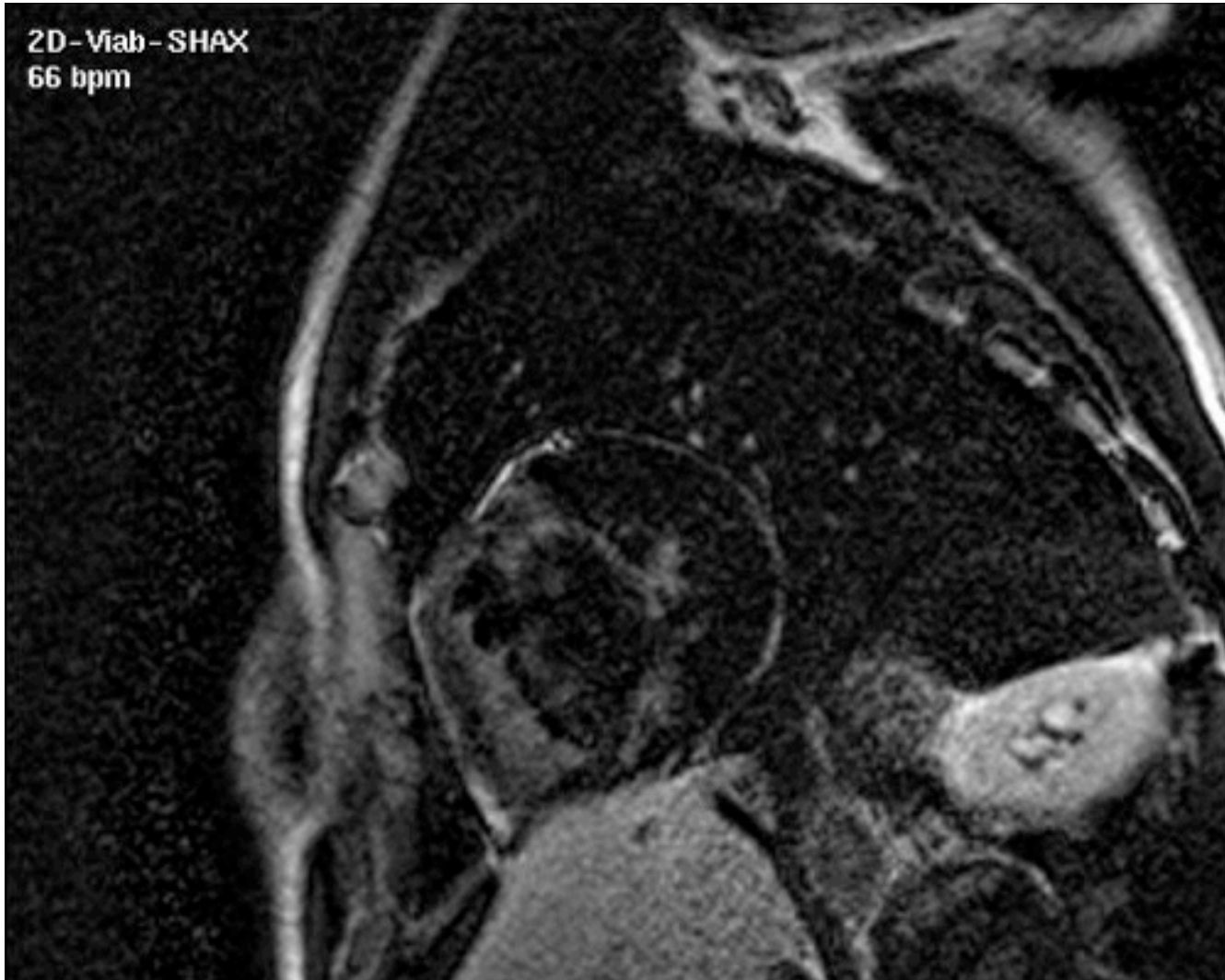


What about our patient?



Our patient: Cardiac MR

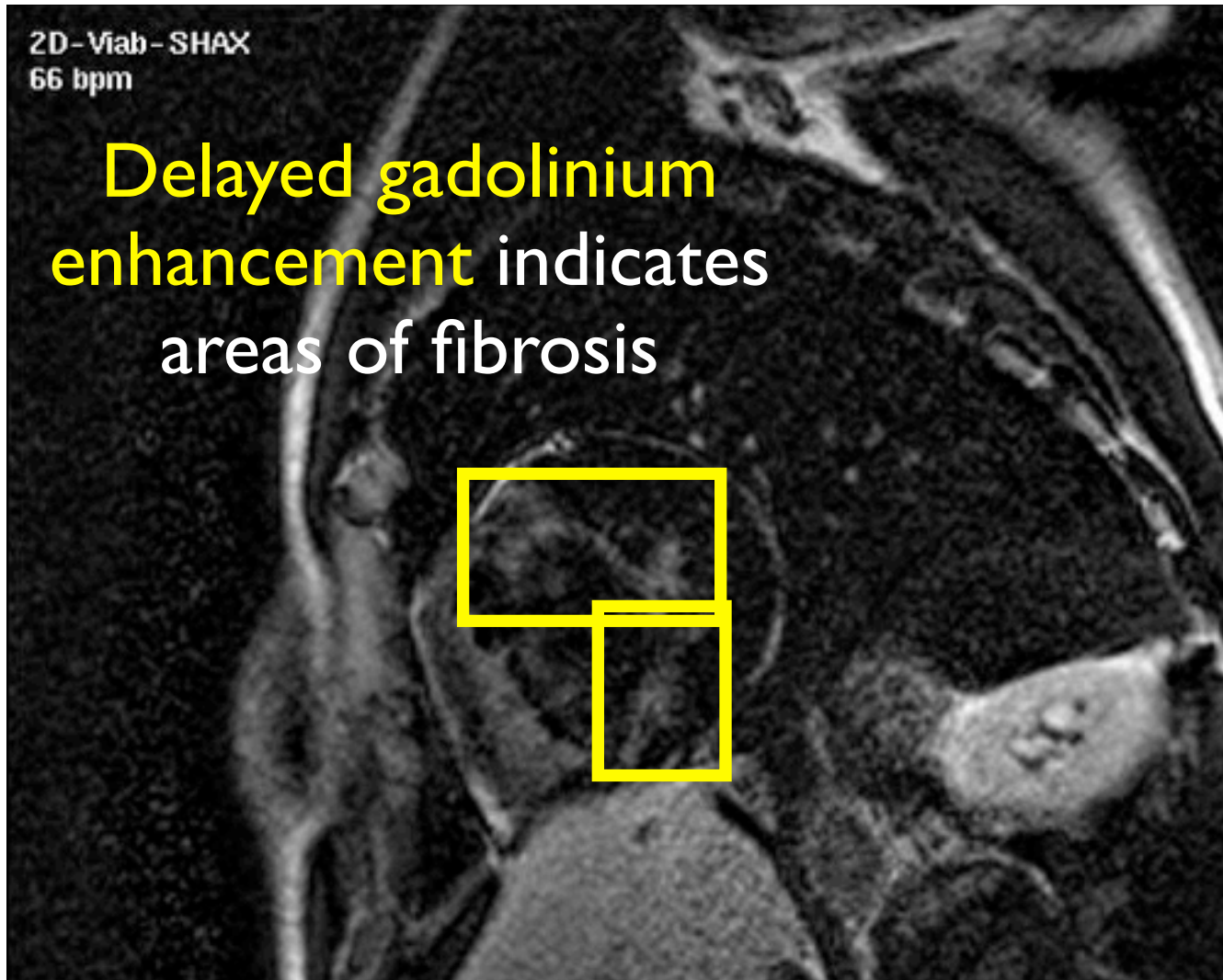
Please evaluate the image and continue to view findings



multiplanar reformat, dark-blood, C+ cardiac MRI

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Our patient: delayed gadolinium enhancement on cardiac MR



multiplanar reformat, dark-blood, C+ cardiac MRI

What does this mean for our patient?

- Interpreting our patient's DGE findings is difficult. Correlating the amount and distribution of DGE in patients with HCM and their actual risk for complications is an area of active research
- In the future, it may be possible to use DGE to identify patients who need prophylactic ICD implantation to prevent sudden cardiac death



Summary

- Echocardiography is the first line diagnostic method for HCM
- MR can help with
 - anatomy: visualizing the entire LV wall
 - function: visualizing areas of akinesis
 - prognosis: identifying areas of fibrosis



References

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Acknowledgements

- Dr. Gillian Lieberman
- Dr. Martin Smith
- Dr. Raymond Chan