Imaging the Cardiomyopathies
A SAM PRESENTATION

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During this Presentation We will discuss........

• Definition and Classification of the Cardiomyopathies

Cardiomyopathy and DME: How it is performed and patterns of disease

• Illustrative examples for audience response
Classification of the Cardiomyopathies (WHO 1996 AHA 2006)

Primary Cardiomyopathies
Genetic Etiology: HCM, ARVD, LVNC
Mixed Etiology: DCM, RCM
Acquired: Inflammatory
Acquired: Infiltrative
Misc: Tukatsubo, Peripartum

Maron, B Circulation 2006
Imaging Cardiomyopathy Protocol

Cine SSFP SAX, 4C Horizontal and Vertical LAX, 3C LVOT, Phase Contrast imaging of valve flow and function
Delayed Myocardial Enhancement
A DIAGNOSTIC BREAKTHROUGH!

In 1999, Kim et al demonstrated a match between the enhanced zone of tissue on GAD enhanced viability MR sequences and infarct size on morphometry studies in canines. He concluded that the extent of infarction with GRE-IR images was identical to the extent of myocardial necrosis.
What is the Mechanism of Delayed Myocardial Enhancement?

Paramagnetic contrast resulting in TI shortening diffuses rapidly into myocardium (< 5 minutes)
Higher concentration of GAD in necrotic / fibrous tissue with faster TI recovery allows increased signal with TI-weighted pulse sequences
Delayed Enhancement Mechanism

Viable Myocardium

- Intact cell membranes
- $[\text{Gd}] = \text{Low}$

Acute MI

- Ruptured cell membranes
- $[\text{Gd}] = \text{High}$

Scar or edema

- Collagen matrix
- $[\text{Gd}] = \text{High}$

Kim R et al 2002
Double dose (0.2 mmol/kg) Gad (T1 shortening paramagnetic).
Scan 10 – 15 minutes post-contrast
T1 weighted GRE with a nonselective 180 degree IR pulse. TI adjusted to null normal myocardium

ECG

Trigger

Necrotic

Viable

180° IR

data

250 - 350 ms

180° IR

data

250 - 350 ms

Delayed Myocardial Enhancement

Siemens Medical System
Phases of Myocardial Enhancement following GAD injection

- **Viable Myocardium**
- **Infarcted or fibrotic Myocardium**
- **Ischemic but viable Myocardium**

**First-Pass**

- Time: < 1 min

**Delayed Enhancement**

- Time: > 5 min

Courtesy of Siemens Medical Systems
Patterns of DME
Transmural, subepicardial, mid-myocardial, subendocardial, diffuse
ROLE OF DELAYED ENHANCEMENT

Helps to estimate functional impairment
Separate ischemic from nonischemic etiologies
Narrow the DDx based on patterns of DME
Help to guide biopsy or obviate need for biopsy
Arrhythmogenic RV Dysplasia

- Fatty replacement in RV free wall on SE black blood images
- RV dilatation
- Autosomal dominant  Familial in 30%
- Systolic and/or diastolic dysfunction, sacculation or aneurysms
- DME found in diffuse or segmental RV scar and correlates with decreased RVEF

AHA  ARVD Task Force 2010
28 year old athlete with syncope. Arrhythmia and angina on exertion.

Courtesy of Mark Fogel MD
Hypertrophic Cardiomyopathy

- Mainly genetic disorder (autosomal dominant) resulting in LV hypertrophy
- Occurs in 1/500 with preferential septal involvement

Maron BJ JAMA 2002
Hypertrophic Cardiomyopathy

- Myofibril disarray due to contractile protein mutations
- ASH is most common together with symmetrical, apical, mid-ventricular and misc subtypes
- Systolic function normal to increased, LV volume normal to reduced, diastolic dysfunction due to altered compliance
- SAM with secondary mitral regurgitation is characteristic
Hypertrophic Cardiomyopathy

• DME directly correlates with extent of fibrosis within hypertrophic myocardium

Patchy small particle pattern mid and basal anteroseptum and RV insertion points

Sites of maximum hypertrophy in other types

Moon JC et al JACC 2004
Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy

- DME is useful to assess the results of transcatheter septal ablation where decrease in the extent of hypertrophy plus hyperenhancement at the ablation site may indicate success of therapy.
Hypertrophic Cardiomyopathy
Status Post Alcohol Ablation

First Pass study

Music of the Heart

PSIR DME study

Courtesy of Gilbert Raff MD
Acute Myocarditis

• A cause of acute heart failure usually secondary to viral infections in young adults

• Most have DME (84%) usually beginning in the epicardium and extending to mid-myocardium localized to the inferolateral and lateral LV walls initially focal or patchy later diffuse
Subepicardial Delayed Enhancement
16 year old with acute chest pain and URI
.Elevated cardiac enzymes.ST segment changes
LV Noncompaction Cardiomyopathy

• An autosomal dominant condition
• Prevalence is 0.05% in the general population
• Heart Failure is most common presentation
• Onset of symptoms and/or sudden cardiac death seen in infants to young adulthood

Weiford BC et al. Circulation 2004
Noncompaction Cardiomyopathy

• Arrest of normal embryogenesis of the myocardium
• Persistence of the fetal intertrabecular recesses leading to development of “spongy” myocardium
• Mainly involves the trabecular layer of the developing LV and sometimes RV walls.
• Ratio of 2.3 between the width of the noncompacted and compacted layers in LV systole
Restrictive Cardiomyopathy

• Idiopathic or secondary to radiation or infiltrative diseases such as sarcoidosis and amyloidosis,

• Reduced ventricular filling and diastolic volume leading to ..........

• Biatrial enlargement ,normal sized ventricles and patchy or diffuse myocardial fibrosis

Lim et al AJR 2007
Sarcoidosis

• 5% will have cardiac symptoms but cardiac involvement in up to 50%

• Associated with ventricular arrhythmias, LV dysfunction and conduction abnormalities

• Lymphocyte infiltration edema and damaged myocytes with interstitial fibrosis and scarring
• Found most commonly in mid or subepicardial myocardium but may be transmural

• Septal anterior, lateral and inferior walls

• In 319 cardiac segments, transmural in 71, subepicardial in 89, mid-myocardial in 17, subendocardial in 21.

Matoh F et al J cardiol 2008
Ichinose A et al AJR 2008
56 year old Female with Pulmonary Sarcoidosis

58 year old man with known sarcoidosis and ventricular tachycardia

56 year old Female with Pulmonary Sarcoidosis

Belloni et al AJR 2008

Lim et al AJR 2007
Amyloidosis

- Extracellular deposition of abnormal fibrillary proteins
- Primary-common cardiac involvement
- Familial-autosomal dominant rarely cardiac
- Systemic-most frequently cardiac
Cardiac Amyloidosis

- Ventricular involvement with diffuse mural thickening and DME in affected areas
- Global or subendocardial DME due to increased extracellular space with amyloid deposition

76 year old female with proven amyloidosis   Maceira AM et al Circ 2005
42 year old female with arrythmias, LV thickening and liver disease
Amyloidosis

• IMPORTANT!!!!

• Abnormal myocardial and blood pool GAD kinetics: Myocardium has shorter TI than normals and washout from myocardium and blood is faster than in normals into the virtual amyloid “compartment”
Magnitude Image of Normal Myocardium:

Blood nulls at earlier TI than myocardium

- Blood (shorter T1)
- Normal myocardium (longer T1)

- Positive signal
- Negative signal

earlier TI  
later TI
Magnitude Image of Amyloid Myocardium:

Myocardium nulls at earlier TI than blood

positive signal

negative signal

IR

time

earlier TI

later TI

Courtesy DR Tom Flohr
Audience Response Cases
Question One

- The following is least likely to be a feature of Idiopathic Hypertrophic Cardiomyopathy:
  - 1) Systolic anterior displacement of the mitral valve
  - 2) Selective LV apical hypertrophy
  - 3) Transmural delayed enhancement
  - 4) Asymmetrical septal hypertrophy
Question One

• Answer 3
• Rationale: DME in HOCM is usually found in mid-myocardium rather than transmural DME usually found in ACS. Answers 1, 2 and 4 are classic patterns of HOCM
• Reference: Cummings KW et al Radiographics 2009 29:89-103
Question Two

- In Dilated Cardiomyopathy, a vascular DME distribution suggests
- 1) Ischemic etiology
- 2) Nonischemic etiology
- 3) Valvular disease
- 4) Can’t tell which etiology is most likely
- 5) none of the above
Question Two

• Answer: 1

• Rationale: dilated cardiomyopathy may be found in ischemic disease, nonischemic disease and multivalvular disease but vascular distribution suggests an ischemic etiology due to coronary artery disease.

• Reference: O’Donnell et al Radiology 2012;262: 403
Question Three

• In cardiac amyloidosis the following statements are correct
• 1) DME is due to myocardial fibrosis
• 2) Slower wash out of GAD than in the normal myocardium
• 3) Faster washout of GAD than in the normal myocardium
• 4) DME is related to deposition of amyloid in a virtual interstitial compartment
• 5) Answer 1 & 2
• 6) Answers 3 & 4
Rationale: DME in amyloidosis is due to deposition of amyloid in the interstitium including the myocardium rather than scarring, edema or fibrosis.

Deposited amyloid in the myocardium has a shorter T1 than normal myocardium therefore washes out faster.

Reference: Cummings KW et al Radiographics 2009;29:89
Question Four

AN 18 YO WITH A HISTORY OF CHEST PAIN, SYNCOPE AND AN ARRHYTHMIA.

RVEF of 23%
What is your Diagnosis?

1. Hypertrophic Cardiomyopathy
2. Constrictive Pericarditis
3. Arrhythmogenic Right Ventricular Dysplasia
4. Acute Myocarditis
Answer: 3  Arrhythmogenic Right Ventricular Dysplasia

- Disorder of young adulthood
- Primarily a right sided cardiomyopathy
- Characteristic MRI findings contribute to diagnostic criteria include
  - RV wall thinning and fatty infiltration
  - RV dilation dysfunction and saccular aneurysm
CASE FIVE
A Young African- American Athlete
with Syncope and Occasional Chest Pain. His father died at an early age due to “heart disease”
YOUR DIAGNOSIS?

• 1. HYPERTROPHIC CARDIOMYOPATHY
• 2. CONSTRUCTIVE PERICARDITIS
• 3. ARRHYTHMOGENIC RV DYSPLASIA
• 4. ACUTE MYOCARDITIS
What is your Diagnosis?

1. Hypertrophic Cardiomyopathy
2. Constrictive Pericarditis
3. Arrhythmogenic RV Dysplasia
4. Acute Myocarditis
Heterogeneous Myocardial involvement most often involves the interventricular septum.
Measurement of wall thickening to establish diagnosis and for follow-up.
Impaired diastolic function.
MRI is preferred to ECHO as it evaluates all the myocardial segments with equal accuracy for distribution and severity of hypertrophy.
CASE SIX

• A 28 year old with a history of CVA and Tachyarrhythmia
Diagnosis?

1. Hypertrophic Cardiomyopathy
2. ARVD
3. Acute Myocarditis
4. Noncompaction Cardiomyopathy
DIAGNOSIS?

1. Hypertrophic Cardiomyopathy
2. ARVD
3. Acute Myocarditis
4. Noncompaction Cardiomyopathy
Isolated Ventricular Noncompaction

- Distinct form of cardiomyopathy due to intrauterine arrest of myocardial compaction
- Two layers of thickened LV wall
  - Thin, compact epicardial layer
  - Thick endocardial layer with prominent fine trabeculations and deep recesses
- High morbidity and mortality in young to middle aged adults due to heart failure, thromboembolic events and ventricular arrhythmia

Oechslin et al. JACC 2000;36:493-499
Summary: Role of MR in Cardiomyopathy

- Characterize morphology
- Measure cardiac function
- Delayed enhancement will detect inflammatory or infiltrative processes and fibrosis or scarring
- Serial follow-up has high accuracy and reproducibility to monitor the efficacy of treatment
31 year old with chest pain, fever, and SOB
Arrhythmogenic RV Dysplasia

- Fatty type without wall thinning
- Fibrofatty type with wall thinning
- May involve the LV
- Task force Criteria (2010)
- Use Segmented for cooperative patients with regular cardiac rhythm
- Use Single-Shot for uncooperative or arrhythmic patients

Segmented IR TurboFLASH
- patient had poor breath-hold
- poor image quality

Single-Shot IR TrueFISP
- patient had poor breath-hold
- acceptable image quality
Sarcoidosis

- Clinically evident in 5% of those with sarcoid 20-58% at autopsy
- Inflammatory cells, interstitial edema and damaged myocytes
- Fatal ventricular tachyarrhythmias, conduction disturbances and LV dysfunction
- Dyspnea, chest pain, syncope, palpitations
19 year old African-American man with exercised-induced chest pain
Acute Myocarditis

- Myocardial inflammation - viral
- Clinical presentation – CP, ischemic pattern on ECG, elevated serum troponin
- Major dif dx – acute coronary syndrome
- Difficult diagnosis in the past –
  - Low sensitivity of detection
  - Endomyocardial biopsies positive 30%
  - Focal not diffuse myocardial involvement at presentation
A 32 year old female long distant runner with aortic regurgitation
Amyloidosis

Prognostic Implications of DME
Perugini et al suggested that extent of DME has no correlation with clinical or functional characteristics of the disease

Perugini et al Heart 92:343 2006
Amyloidosis

• As a result, similar TI values for blood and myocardium
• The myocardium null point is earlier than the blood pool
• Equalization of inversion time between subendocardium and epicardium earlier than in others
DME correlates with decline in function, worse prognosis and sudden death.
Delayed Gadolinium Enhanced MRI

While the differential enhancement of abnormal myocardium is widely used and popularized during the last decade
PSIR Image of Normal Myocardium:

Blood is relatively brighter than myocardium at all TI’s
Myocardial Amyloidosis: Difficult to recognize

KEY POINTS

Diffuse contrast uptake and equalization of T1-shortening throughout entire myocardium

Thus ...Finding the optimal TI for images is very difficult

TI Scout and PSIR are very useful diagnostic tools
Delayed Myocardial Enhancement in the Nonischemic Cardiomyopathies

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WHO Classification (1995)
Hypertrophic
Dilated
Restrictive
Arrhythmogenic
****
Valvular
Ischemic
Hypertensive

Richardson P et al Circulation 1996; 93:841
Dilated Cardiomyopathy

• Nonischemic DCM characterized by enlarged chamber volumes and impaired contraction not explained by coronary artery disease.
• Caused by familial, genetic, autoimmune, alcoholic or toxic etiologies
• Arrhythmias, thrombi and sudden death are frequent

Mc Crohan JA et al Circulation 2003
Classification of the Cardiomyopathies (WHO)

Cardiomyopathies are myocardial diseases associated with cardiac dysfunction. Cardiomyopathies were classified as hypertrophic, inflammatory, dilated, ischemic, restrictive, ARVD, and infiltrative.

Richardson PJ et al. Circulation 1996
Recognition of the The Differential Enhancement of Normal and Abnormal Myocardial Tissue with CT goes back to the 1970’s. The original MRI work was done in situ in canines with EKG gated T1 WT MR images.

Ordovas K and Higgins C. Radiology 2011 261:358
Ischemic Cardiomyopathy
Later studies showed that reperfused and nonperfused myocardial infarction could be distinguished.
No delayed enhancement was found when myocardium was reversibly injured but most often occurred with irreversible injury.

Performing Delayed Myocardial Enhancement Sequence

Intravenous injection of 0.1-0.2 mmol/kg Gadolinium chelate at 1-2 ml/sec
First pass images in multiple planes
At 7-8 min, a variable inversion time sequence is selected that optimally nulls the myocardial signal
Each of the approximately 20 images is selected with a different IR time. Imaging is initiated at 10-15 minutes with the SAX signal-nulled TIW sequence from cardiac base to apex, 2-chamber and 4 chamber LAX Images are also obtained using phase sensitive or magnitude imaging. Myocardium will appear dark except for the abnormal myocardium.

Cummings KW et al Radiographic 2009
Flohr, Thomas Personal Communication
Delayed Enhancement Acquisition Methods

2-D DE-MRI (segmented k-space GRE-IR) images with a breath hold across a range of inversion times

Scout low resolution SSFP cine imaging with selected inversion times

Phase sensitive Inversion recovery (PSIR) can be performed without concern for optimal inversion times
Prediction of Recovery

Less than 25% DME resulted in 82% recovery
>75% DME resulted in an 8% chance of recovery
Less than 50% DME yielded a recovery of 33%

Kim R et al NEJM 343:1445 2000
Delayed Enhancement without a History of Myocardial Infarction

In 105 patients with suspected CAD but no history of MI 23% had DME—the strongest indication of an acute event in 16 month followup.

In 240 middle aged patients DME in 25% of whom most were endocardial.

Barbier et al JACC 48:705 2006
Hypertrophic Cardiomyopathy

Patterns of hyperenhancement

Moon et al. JACC 2003 41;1561-1572
Acute myocarditis: DME is due to edema rather than fibrosis or infarction.
Chronic myocarditis: DME is due to fibrosis seen in 70%.
Midwall (68%) and subepicardial (38%) zones are most common.

Laissy JP et al Radiology 237:75 2005
Left Ventricular Noncompaction

- DME in the deep trabecular recesses of the abnormal myocardium
- DME relates inversely to global LV function

Petersen S JACC 2005
31 year old with chest pain fever and SOB
PSIR Image of Amyloid Myocardium:
Myocardium is brighter than blood at all TI’s

positive signal

amyloid myocardium (shorter T1)

negative signal

blood (longer T1)

all TI’s

Courtesy of DR Tom Flohr
Disclosures

• Consultant: Johnson and Johnson

• Consultant: Educational Symposia
• 101 adult patients followed for 1-2 years
• 35% had midwall fibrosis detected by MDE
• Midwall fibrosis is a significant predictor of hospitalization or sudden cardiac death
• May be used to predict the need for AICD

Assomull et al. *JACC* 2006;48:1977
CASE SIX

• A 28-year-old with a history of CVA and Tachyarrhythmia
Arrhythmogenic Right Ventricular Dysplasia

14 yrs, Nonsustained Vtach RVEF 38%
What happens to the DME zone over time?

In one study, the zone of DME decreased 40% over 5 months.

In a study following ablation of the IVS in HOCM, there was no change over 180 days.

In a third study, substantial decrease in the zone of DME from acute to chronic infarct occurred over months.

Ingkanisorn WP et al JACC 43:2253 2004
“Cardiomyopathies are either limited to the heart or are part of a generalized disorder often leading to CV death and/or progressive HF”

Maron B et al. Circulation 2006;113:1807
PREDICTING RECOVERY
Correlating the extent of the zone of DME with acute infarct or with chronic ischemic dysfunction
Choi found that the extent of DME 7 days after the acute event predicted recovery of EF and wall thickening in 8-12 weeks. The best chance of recovery was in those with < 25% of DME or no DME.

Choi et al Circulation 2004
Myocardial Amyloidosis: Use both Magnitude and PSIR images as tools

Magnitude Images:
- Myocardium nulls at earlier TI
- Blood nulls at later TI

PSIR Images:
- Myocardium is brighter at all TI’s
- Blood is darker at all TI’s

Courtesy of DR Tom Flohr
Myocardial Amyloidosis:
Use TI Scout as a tool

If blood nulls before myocardium, then it’s probably normal myocardium.

If myocardium nulls before blood, then it’s probably amyloidosis.
Arrhythmogenic Right Ventricular Dysplasia

- Cardiomyopathy → fibro-fatty infiltration of RV
- Presentation: arrhythmias or sudden cardiac death
- AICD may save life
- Biopsies unreliable
  - patchy distribution → false negatives
- Diagnosis based on major and minor criteria
Amyloidosis

- DME: global diffuse or patchy subendocardial to mid-myocardial enhancement due to interstitial compartment expansion with amyloid infiltration without fibrosis.

Involvement of atrial walls, septum and ventricles
Histologic Basis of Delayed Myocardial Enhancement

Moon et al. JACC 2004; 43:2260
Introduction

- Cardiac MRI is an essential modality for the evaluation of congenital and acquired cardiac disorders.
- Although important for the evaluation of morphology and function, a unique role for MRI is to determine the presence or absence of both ischemic and nonischemic cardiomyopathies characterized by myocardial infarction, fibrosis or edema using viability imaging methods.
Dilated Cardiomyopathy

DME useful to differentiate ischemic from nonischemic cardiomyopathy. 50% were free of DME and 28% had midwall distribution in patients with normal coronary arteries suggesting nonischemic etiology.

Tigen K et al J Am Soc Echo 23:416 2010
Definition of Cardiomyopathy
“Cardiomyopathies are a heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that may exhibit ventricular hypertrophy or dilatation due to a variety of causes”

Maron B et al. Circulation 2006;113:1807
Arrhythmogenic Right Ventricular Dysplasia

- Progressive fibrofatty replacement of the myocardium
- Familial in >30% autosomal dominant
- Ventricular arrhythmia with LBB pattern is most common causing sudden cardiac death in 20% of adults under 35 years

Mahrholdt H et al Eur Heart J 2005
AHA task force 2010
Idiopathic Dilated Cardiomyopathy

- Dilatation and impaired contraction of both ventricles in the absence of volume loading conditions such as valvular regurgitation or ischemic heart disease
- Unclear etiology – genetic, viral, metabolic or toxic
- Progressive myocardial necrosis with interstitial fibrosis and wall thinning
- MRI considered the most accurate imaging technique to estimate ventricular size, myocardial function and wall thickness
DME Pattern and Diagnosis?
Myocardial Delayed Enhancement

Figure 1
Ischemic cardiomyopathy

Figure 2
Dilated cardiomyopathy

Figures 3 & 4
Hypertrophic cardiomyopathy

Figures 5 & 6
Myocarditis

Courtesy of Dr. Okan Ekinci, Deutsche Klinik für Diagnostik, Wiesbaden, Germany
HCM vs Athlete’s Heart

Jan 2006
LV Mass 285g/m²

March 2006
LV Mass 227g/m²
Dilated Cardiomyopathy

Transmural and vascular distribution of delayed enhancement pattern indicates probable ischemic etiology