Cardiac MRI Evaluation of Cardiomyopathy and Myocarditis

Laureen Sena
Children’s Hospital
Boston, MA

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Cardiomyopathy

<table>
<thead>
<tr>
<th>Primary</th>
<th>Secondary</th>
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<tbody>
<tr>
<td>Hypertrophic</td>
<td>Ischemic</td>
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<tr>
<td>ARVD</td>
<td>Valvular</td>
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<tr>
<td>Dilated</td>
<td>Hypertensive</td>
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<tr>
<td>Restrictive</td>
<td>Infectious/inflammatory</td>
</tr>
<tr>
<td>Unclassified</td>
<td>Metabolic</td>
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<td></td>
<td>Muscular Dystrophy</td>
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Hypertrophic Cardiomyopathy

- LVOT dynamic outflow obstruction with SAM and endocarditis

Primary Hypertrophic Cardiomyopathy

- Autosomal dominant with variable penetrance and expression
- Myocardial hypertrophy in any region of the left ventricle in the absence of hypertrophic stimulus (valvular aortic stenosis or systemic hypertension)
- Exertional angina, fatigue, syncope, arrhythmia, sudden death
- Most common cause of sudden death in active, apparently healthy adolescents and young adults

Primary Hypertrophic Cardiomyopathy

- Heterogeneous involvement—often asymmetrical and involves the interventricular septum
- Detection and measurement of wall thickening to establish diagnosis and for follow-up
- Impaired diastolic function
- Echo can have limited evaluation of the RV and the inferior and apical wall of LV
- MRI evaluates all of the myocardial segments with equal accuracy for the distribution and severity of hypertrophy (ventricular mass)
Conventional risk factors for sudden death in HCM

- Unexplained syncope
- Nonsustained ventricular tachycardia
- Abnormal BP response to exercise
- Wall thickness greater than 30 mm

Myocardial delayed enhancement correlates with progressive decline in function, worse prognosis and sudden death in patients with 2 or more conventional risk factors.

Moon et al. JACC 2003 41;1561

Bogaert et al. AJR 2003;180

Hypertrophic Cardiomyopathy

Patterns of hyperenhancement

Moon et al. JACC 2003 41;1561-1572

Myocardial Delayed Enhancement

Normal Myocardium

Acute MI

Scar

Intact cell membrane

Ruptured cell membrane

Collagen matrix

[Gad] = Small

[Gad] = Large

[Gad] = Large

Histologic Basis of Myocardial Delayed Enhancement

MDE = Increased myocardial collagen

Moon et al. JACC 2004; 43:2260
**HCM vs Athlete’s Heart**

<table>
<thead>
<tr>
<th>Jan 2006</th>
<th>March 2006</th>
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<tr>
<td>LV Mass 285g/m²</td>
<td>LV Mass 227g/m²</td>
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</table>

**Hypertrophic Cardiomyopathy**

**Cardiac Fibroma**

- SSFP
- FSET1
- FSE T2 fat sat
- Perfusion
- Post GAD T1 Fat Sat
- Del Enh

**Dilated Cardiomyopathy**

- Decreased ejection fraction, increased EDV and ESV and wall motion abnormalities.
- Can be difficult to distinguish between dilated form of ischemic CM and nonischemic dilated CM
- Segmental wall thinning = old infarction
- Uniform wall thinning and enlarged LV trabeculae = nonischemic dilated CM
- Impaired RV function associated with LV dysfunction is associated with reduced exercise capacity and survival

*(Brieke and DeNofrio CAD 2005;16:5-11)*

**Dilated Cardiomyopathy**

- Delayed hyperenhancement can be absent in 60%
- Patchy midwall hyperenhancement in a noncoronary pattern
Dilated Cardiomyopathy

Subendocardial delayed enhancement indicates probable ischemic CM

LV Apical Aneurysm

- Becker and Duchenne
- X-linked recessive neuromuscular disorders with progressive cardiac and skeletal muscle weakness
- Mutations in the dystrophin gene cause dystrophin deficiency that leads to LV midwall necrosis and fibrosis at path – see diffuse midwall delayed enhancement (Varghese and Pennell, Heart 2004;90)
- Often followed for ventricular size and function by MRI – prior to spinal fusion and for experimental drug protocols to improve and prevent further ventricular dysfunction (Carvediol)

Beckers Muscular Dystrophy
5-y-old girl, s/p coarctation repair with subaortic stenosis

Left Ventricular Noncompaction

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

Ventricular Noncompaction - Pathogenesis

- Arrested myocardial compaction
- Microcirculatory dysfunction?
  - Subendocardial fibrosis (EFE)
  - Systolic and diastolic dysfunction
- Familial occurrence in 44%
  - 50% of infantile cases, 18% of adult cases
- 3 Gene loci for X-linked and AD transmission
- Neuromuscular disorders in 82% of adults
- Facial dysmorphism in 38% of children
- Higher incidence of WPW in Japanese children

Ichida et al. JACC 1999;34:233-40
### Left Ventricular Noncompaction

- Noncompaction can be seen in normal individuals
  - Majority at apex
  - Variation of normal?
- Pathological noncompaction
  - Involves more segments than just the apex
  - Ratio of noncompacted to compacted layers >2.3 in diastole
  - Associated with dilated and hypertrophic CM
  - Associated left sided obstructive lesions

*Peterson et al JACC 2005*
*Biagini et al AJC 2006*

### Cardiomyopathy Related to Myocardial Siderosis or Iron Overload

- Cardiac complications cause 70% of deaths in patients with thalassemia major
- Extremely poor prognosis once heart failure develops (worse than idiopathic dilated CM)

### Myocardial Iron Overload in Thalassemia

- Iron induced cardiomyopathy shown to be reversible with improved LV volume and function during chelation therapy – correlated with improvement in myocardial T2* measurement *Anderson et al Br J Haem 2004;127:348-355*
- Myocardial T2* normalizes more slowly in the heart than the liver due to different rates of clearance *Anderson et al. Eur Heart J 2001;22:2171*
- No correlation between myocardial iron and liver iron and serum ferritin
- Direct T2* measurement of the myocardium needs to be performed to follow-up patients on chelation therapy

### Arrhythmogenic Right Ventricular Dysplasia/ Cardiomyopathy

- Disorder of young adults
- Reentrant ventricular arrhythmias
- Right sided cardiomyopathy
- High familial incidence
- Characteristic MRI findings contribute to diagnostic criteria
  - RV wall thinning and fatty infiltration
  - RV dilation and dysfunction
- Assessment of regional wall motion is now an essential part of the examination
**Arrhythmogenic Right Ventricular Dysplasia/ Cardiomyopathy**

- Clinical presentation is variable
  - syncope and fatigue with minimal exertion
  - arrhythmia with exercise
  - sudden death
- Usually present under age 40 – young men
  - Increased recognition – has been diagnosed from fetal age to 80 yrs old

**Arrhythmogenic Right Ventricular Dysplasia/ Cardiomyopathy**

| 14 yrs, Nonsustained Vtach | RVEF 38% |

**Regional Dysfunction - ARVD**

**Constrictive Pericarditis**

**Diastolic Dysfunction**
Constrictive Pericarditis
Diastolic Dysfunction
Elongated narrow RV with sigmoid shaped intraventricular septum, dilated atria and impaired ventricular filling

Restrictive Cardiomyopathy
Idiopathic or infiltrative (amyloid, sarcoid)
Ventricular hypertrophy with impaired filling
Dilated atria with relatively small ventricles

Restrictive Cardiomyopathy-
Endocardial Fibroelastosis (EFE)
10 yr old with BAV and critical AS, s/p balloon valvuloplasty, pulmonary htn

Viral Myocarditis
Subepicardial Delayed Enhancement

Viral Myocarditis

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
CMR for Acute Myocarditis

Diagnostic Performance of CMR in Suspected Acute Myocarditis. Abdel et al. JACC 2005

- 25 pts – Dx by clinical criteria (only 2 biopsies)
- T2 TIR – myocardial edema
- T1 spin echo pre and post Gad – hyperemia compared to skeletal muscle
- Inversion recovery GRE (Del Enh) – fibrosis
- Best detection when “any two” of the 3 sequences were positive

Acute Myocarditis

TSE-BB T2  TSE-BB PD post Gad  MDE

18 yr old with chest pain, nonsustained VTach, and elevated troponins

Acknowledgements

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Summary: Role of CMR in Cardiomyopathy

- Characterize morphology
- Measure cardiac function – systolic and diastolic
- Delayed enhancement to detect inflammatory or infiltrative processes and fibrosis or scarring (prognosis)
- Add T2 and early post Imaging for suspected myocarditis
- T2* for iron overload
- Serial follow-up has high accuracy and reproducibility to monitor the efficacy of treatment

Thank you for your attention!!!