2015 Diagnostic and Prognostic Imaging (MR/CT) of Various Cardiomyopathies: Myocarditis, Dilated, Infiltrative (Amyloid, Sarcoid), Hypertrophic, Noncompaction

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• There are no financial relationships relevant to this presentation.

• Off label use: MRI of patients with implanted devices; Gadolinium for cardiac studies
CMR is the Ideal Tool for Assessment of Myocardial Tissue

- Assessment of left and right ventricular
  - Size; Volume; Wall thickness
  - Mass; Ejection fraction
- Resting and stress perfusion imaging
- Late gadolinium enhancement – for scar
- T2* - for myocardial iron
- Fat suppression black blood – for fatty infiltration
- T2-weighted imaging – for myocardial edema
- T1 mapping – for diffuse fibrosis
Ventricular Scar Imaging

- Gadolinium Contrast
- Extensively validated
- High spatial resolution


Non-Ischemic Cardiomyopathy

Scar can be present and may be the substrate for VT
**Idiopathic Dilated Cardiomyopathy**

- LV (or biventricular) dysfunction after exclusion of other causes of myopathy
  - 46,000 hospitalizations and 10,000 deaths per year
  - Primary indication for cardiac transplantation

- CMR
  - Mid-wall or patchy scar
  - Mid-wall scar is seen in 30% of patients with idiopathic dilated cardiomyopathy

Midwall Fibrosis

- In 26 patients with IDC

- Midwall scar with > 25% scar transmurality was associated with inducible VT
  - OR 9.1, P=0.02 (multivariate analysis)

Midwall Fibrosis Implications

- Wu et al – In 65 DCM patients followed for median 17 months: Midwall scar
  - Was present in 42%
  - Was associated with CHF hospitalization, ICD firing, or cardiac death (HR 8.2, P<0.01)

- Assoumull et al – In 101 DCM patients followed for 658 +/- 355 days: Midwall scar
  - Was present in 35%
  - Was associated with all-cause death and hospitalization (HR 3.4, P=0.01)
  - Was associated with sudden death or VT (HR 5.2, P=0.03)

Midwall Fibrosis can Guide VT Ablation: Success Site Along Scar
T1 Mapping
Correlates with normal and abnormal myocardium

Most important for diffuse fibrosis

Turkbey, Nazarian, et al AHA 2011
Dilated Cardiomyopathy

• Diagnosis:
  – CMR and CT good for functional assessment
  – Patterns of LGE can differentiate dilated non-ischemic from ischemic cardiomyopathy

• Prognosis:
  – Midwall fibrosis is an independent predictor of negative outcomes
  – Abnormal T1 mapping is an independent predictor of mortality and need for transplant
Myocarditis
Myocarditis

- Gradual presentation
  - Difficult to diagnose
  - Can rapidly progress to heart failure and/or SCD

- CMR features
  - Myocardial edema (T2)
    - Higher sensitivity than LGE (84% vs. 44%)
  - Wall motion abnormalities and hyperemia
  - Patchy LGE (95%)
    - PVB19 predominantly lateral wall of LV - epicardial
    - HHV6 predominantly anteroseptal – intramural
    - Decreases with time and can monitor response to therapy

Myocarditis

• Diagnosis: (Pts with functional abnormalities)
  – Increased intensity in T2 weighted images (edema)
  – Increased ratio of LGE in myocardium to skeletal muscle (hyperemia)
  – Focal midwall or epicardial LGE

• Prognosis:
  – Presence of LGE – hazard ratio of 10 for mortality
Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy

- Inherited disorder – 0.2% in US
  - LV hypertrophy – focal or diffuse
  - Phenotypic expression varies
  - Myocardial disarray
  - 1.5% annual risk of SCD
- Other diseases can mimic HCM
- LGE can be observed
  - Typically at RV insertion points
- T2 abnormalities can be seen (33%) in the setting of severe LVH and at sites with LGE

Frenneaux, et al. Heart 2004;90;570-575
CMR for HCM Risk Stratification

Rubinshtein, Gersh, et al Circ Heart Fail. 2010 Jan;3(1):51-8

- 424 pts with HCM (no prior ablation/myectomy; age 55±16); a mean f/u of 43±14 months
  - 56% had LGE, ranging from 0.4% to 65%
  - Gene positive patients were more likely to have LGE (75% vs. 53%, P<0.001)
  - NYHA class was unassociated with LGE
  - LGE-positive patients
    - More likely to have NSVT (27% vs. 8.5%, P<0.001)
    - More episodes NSVT/pt (4.5+/−12 vs. 1.1+/−0.3, P=0.04)
    - ↑ PVCs/24 hours (700±2080 vs. 103±460, P=0.003)
  - SCD and ICD shocks only in LGE + patients
Hypertrophic Cardiomyopathy

• **Diagnosis:**
  – CMR is recommended for patients in whom echocardiography is insufficient for diagnosis

• **Prognosis:**
  – Presence of LGE is an independent predictor of sudden cardiac death (even in patients with less severe hypertrophy)
  – LGE not currently in guidelines as a factor in determining indications for ICD placement
Amyloidosis
Cardiac Amyloidosis

- Manifestation of one of several systemic diseases
- Extracellular protein deposition
- Cardiac involvement
  - Common with senile systemic, and transthyretin amyloidosis
  - Absent to severe (!) in light chain amyloidosis
  - Rare in secondary amyloidosis
- SCD usually due to PEA

Falk, Circulation. 2005 Sep 27;112(13):2047-60
Cardiac Amyloidosis

• CMR
  – Biventricular wall thickening with non-dilated ventricles
  – Atrial dilation
  – LGE
    • Circumferential sub-endocardial
    • Blood pool can appear atypically dark
    • “Zebra-stripe” pattern

Falk, Circulation. 2005 Sep 27;112(13):2047-60
Maceira, Pennell Circulation. 2005 Jan 18;111(2):186-93
In 28 patients with systemic amyloidosis
- LGE was observed in 68%
- During a median follow up of 29 months
  - 18% of patients died
  - LGE did not associate with survival
  - LGE volume was correlated with serum BNP (R=0.64, P<0.001)
    - RV EDV and SV were associated with mortality

In 29 patients with proven cardiac amyloidosis
- During a median follow up of 623 days
  - 58% of patients died
  - LGE did not predict mortality
  - The 2 min post contrast T1 difference between subepicardium and subendocardium predicted mortality
    - 23 ms, 85% accuracy

Amyloidosis

• **Diagnosis:**
  – Endomyocardial biopsy is the current gold standard
  – LGE correlates well with quantity of amyloid
  – LGE in amyloid pattern has a sensitivity of 80% and specificity of 94% for Dx compared with biopsy

• **Prognosis:**
  – LGE did not predict mortality
  – T1 differences between subendocardium and subepicardium did predict mortality
Sarcoidosis
Cardiac Sarcoidosis

- Systemic illness associated with CHF, cor pulmonale, arrhythmias and sudden death
- Japanese Ministry of Health criteria
  - Histologic
    - Endomyocardial biopsy with noncaseating epithelioid granulomas
  - Clinical (in patients with systemic sarcoidosis, must have “a” and one of “b-e”)
    a. RBBB, AV block, VT, PVCs, q waves, or ST-T changes
    b. Abnormal wall motion, regional wall thickening, or LV dilation
    c. Perfusion defect on imaging or abnormal accumulation on scintigraphy
    d. LVEDP, or LV dysfunction
    e. Interstitial fibrosis or >moderate cellular infiltration on biopsy

Cardiac Sarcoidosis

- CMR
  - ↑ T2 signal due to inflammation – can be used to monitor response to therapy
- Regional wall motion abnormalities
- RV dilation, hypertrophy, HK
- LGE - Various patterns:
  - Predilection for basal or midventricular septum
  - Roughly half are endocardial based
  - 2/3 have subendocardial enhancement of the RV side of septum

CMR for Diagnosis and Prognosis of Cardiac Sarcoidosis

• In 81 patients with biopsy proven extracardiac sarcoidosis
  – LGE was observed in 26% (JMH criteria in 12%)
  – During a follow up period of 21±8 months
    • 6 patients died
    • LGE associated with cardiac mortality (11.5 fold increase)
    • LGE associated with the combined endpoint of death, defibrillator shock, or pacemaker requirement (9 fold increase)

Cardiac Sarcoidosis

• **Diagnosis:**
  – No international consensus
  – Cardiac biopsy low yield due to high sampling errors
  – CMR (in pts with extracardiac sarcoid) will likely become the gold standard for cardiac involvement
  – LGE likely indicator of chronic involvement

• **Prognosis:**
  – Presence of LGE is highly predictive of morbidity and mortality.
Non-Compaction
Non-Compaction

- Uncommon genetic cardiomyopathy
  - Rare cause of heart failure
- Characterized by trabeculations and recesses within the ventricular myocardium
  - Most commonly affecting the left ventricle
- Diagnostic criteria inconsistent, but it is an imaging-based diagnosis
Non-compaction: CMR
Non-compaction

- Present with heart failure symptoms
  - Diastolic and systolic dysfunction.
- Risk for thromboembolism and arrhythmias
  - Atrial fibrillation or ventricular tachycardia.
- Early diagnosis may lead to a better prognosis.
- No specific guidelines used for managing.
- Generally managed symptomatically
  - ACEi, beta-blockers
  - Anticoagulants to prevent thromboembolic events.
MRI in Patients with Legacy Devices
Risk is much less than previously thought

- 555 MRI examinations performed in 438 pts
  - 55% with permanent pacemaker
  - 45% with ICD
- Monitoring/programming by device nurse
- No significant adverse events – a few POR
- Trivial changes in pacing parameters
- Diagnostic question answered in 95%


- Now over 2100 scans – similar results
- CMS approved
ICD Artifact Reduction

Visualization of scar with artifact suppression

No ICD

ICD
No Suppression

ICD
Suppression

Courtesy Dr Kolandaivelu
Summary

• The spatial resolution, multi-planar capabilities, and specialized pulse sequences available with CMR provide great utility for
  – Establishing the various etiologies of non-ischemic cardiomyopathy
  – Monitoring of disease progression and response to therapy
  – Risk stratification

• MR Imaging of pts with implanted legacy devices is feasible
Questions?
Apical hypertrophy
Hemochromotosis
Cardiac Hemochromatosis

• Cardiac iron overload
  – Hereditary hemochromatosis
    • Recessive, 1:400 northern Europeans
  – Transfusion dependent anemia

• Iron overload diagnosed by
  – Transferrin saturation > 55%
  – Serum ferritin > 200 and 300 mcg/L in men and women

• Reversible with chelation
  – If started early…

Diagnosis and Risk Stratification in Cardiac Hemochromatosis

- Iron deposits shorten T2*
- In 652 thalassemia major patients
  - Cardiac T2* < 20 ms
    • RR for arrhythmia 4.6
      (95% CI 2.7-8)
  - Cardiac T2* < 10 ms
    • RR for CHF 160
      (95% CI 39-653)
  - Cardiac T2* < 6 ms
    • CHF in 47% within a year
    • RR for CHF 270 (95% CI 64-1129)
    • Arrhythmia in 14% within a year

Images courtesy of S. Zimmerman, JHH
ICD Placement in Sarcoidosis

• Cardiac manifestation – clear presence of scar on Delayed Enhancement MRI
• Some other manifestations
  – Syncope or pre-syncope
  – Arrhythmia
  – Compromised heart function
• Occasionally do EPS
Scar Pattern Predicts Event-Free Survival

- 79 patients
- No ICDs
- EF 25-28%

Event-free survival
Focal, patchy DE: 37.6 ± 2.4 months vs
Diffuse myocardial DE: 24.0 ± 3.5 months

CMR for HCM Risk Stratification


• Of 76 patients with HCM
• 43 were deemed high risk - ≥1 of the following:
  • History of cardiac arrest
  • Family history of sudden cardiac death
  • Unexplained syncope or presyncope
  • Documented non-sustained ventricular tachycardia
  • Presence of severe left ventricular hypertrophy ≥ 30 mm

  – High risk patients had more LGE than low risk patients (67% vs. 47%, P=0.03)
  – High risk patients had higher LGE burden per patient (14% vs. 3%, P<0.001)

• 38 had EP testing (12 had inducible VT)
  – Patients with inducible VT had higher LGE burden (22% vs. 10%, P=0.03)
Non-compaction: Echo
Arrhythmogenic RV Dysplasia

• Inherited cardiomyopathy – AD pattern (*PKP2* – plakophilin-2)
  – Fibro-fatty infiltration of RV >> LV
  – Age 20-40, M > F
  – RV predominance but biventricular reported
• Task force criteria – 2 major; 1 major & 2 minor; or 4 minor criteria
  – Major
    • Family disease confirmed at autopsy or surgery
    • Epsilon waves on ECG
    • Severe segmental or global RV dilation
    • RV aneurysm
    • Fibro-fatty replacement of RV myocardium
  – Minor
    • FH of SCD < 35 or suspected ARVD
    • ECG abnormalities in V1-3
    • Mild global or segmental RV wall motion abnormalities

Al-Mallah, HFR (2011) 16: 369–380
Br Heart J 1994:71
ARVD
Tandri, Bluemke, et al JACC 2005 Jan 4;45(1):98-103

• On CMR
  – RV dyskinesia, sacculation, or aneurysm formation
  – LGE (61%)
    • Correlates with fibro-fatty replacement on biopsy
    • Predicts inducible VT at EP study
Arrhythmogenic right ventricular dysplasia/cardio- 
mymopathy: CMR is indicated for identification of structural 
and functional abnormalities by SSFP in this disease. 
Intrinsic tissue characteristics with fatty infiltration by T1 
imaging and fibrosis by LGE can also be seen.
Arrhythmogenic RV Displasia