Echocardiographic Evaluation of the Cardiomyopathies

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Cardiomyopathies (CMP)

“primary disease intrinsic to cardiac muscle”

- Dilated CMP
- Hypertrophic CMP
- Infiltrative CMP
- Left Ventricular Non-compaction
- Arrhythmogenic Right Ventricular CMP
- Takotsubo Cardiomyopathy
Utility of Echo in CMP

- CMP presentation is **clinically indistinct**
  - New onset CHF
  - Arrhythmias

- Echo is the easiest and most utilized diagnostic test in the evaluation of CHF
  - Etiology
    - Valvular, congenital defects, ischemia
  - Prognosis
    - Systolic function
    - Diastolic function
    - Filling pressures
  - Complications
    - MR
    - Thrombus
Dilated Cardiomyopathy

- Global decrease in systolic contractile function with eventual dilatation of the ventricles

- Echocardiographic features
  - Dilated chambers, LV, LA, RV, RA
  - Usually normal LV wall thickness
Dilated Non-Ischemic CMP - end stage
Apical 4 C Non-ischemic CMP
Mitral Regurgitation

- MV tethering
- MV annulus
- LV
- LA
Mitral Regurgitation

- Altered LV geometry
- Apical tethering of leaflets
- Annular size increases
- Decreased LV closing force
- Decreases survival
Dynamic Mitral Regurgitation
Apical Thrombus
Contrast Enhances Apical Thrombus
Echocardiographic Predictors in CMP

- LV size and function
- RV involvement
- Diastolic indices
  - DT
    - Best predictor of death in symptomatic patients
    - EF< 25%: DT < 130ms mortality in 2 yrs of 65% vs 26% DT >125
  - Filling pressures
  - LA size

- CRT another lecture
Hypertrophic Cardiomyopathy

- *Unexplained* hypertrophy (no HTN, AS…)
  - Any degree or pattern: diffuse, septal, apical

- Abnormalities of diastolic function
  - Always present
    - Not a consequence of hypertrophy
    - Precedes hypertrophy!!
  - Correlates with symptoms
  - Early diagnosis in genetically at risk

- Increased risk of sudden death
  - Athletes
  - Marked septal hypertrophy
  - Family history of sudden death
  - LVOT obstruction with mild or no symptoms
Hypertrophic CMP Screening

- Autosomal dominant
- Phenotypic expression variable
- 1/500
- First degree relatives
  - Ages 12-18 yrs annual clinical, ecg, echo
  - > 18 yrs ASX every 5 years
  - <12 yrs evaluate if symptomatic or intense sports
  - Affected family members need repeat screening every 12-18 months
Echocardiographic use in HCM

- **Best screening test**
- Defines magnitude and location of hypertrophy
- Excludes other causes of hypertrophy (AS, VSD)
- LV wall thickness >13 mm
  - Does not require asymmetry (not specific)
  - ASH—leads to earlier diagnosis---SAM—murmur and symptoms
Hypertrophic Cardiomyopathy
LVOT Obstruction

- **Systolic Anterior Motion (SAM)** most common cause of LVOT obstruction in HCM
- Only 25% of HCM at rest have LVOTo > 30 mmHg
- **Dynamic**
  - Recent study found 70% of patients without LVOTo at rest developed LVOTo with exercise
  - Increases with increases in contractility and HR
  - Decreases with increases in LV volume
LVOT Obstruction

- Independently predicts
  - More severe diastolic dysfunction and symptoms
  - Stroke
  - Atrial fibrillation
  - Arrhythmia-related death

- Most common in
  - Females
  - Elderly
  - Greater degree of hypertrophy
Systolic Anterior Motion

- Abnormal independent anterior motion of the mitral leaflets into the LVOT during systole
- Either leaflet; anterior, posterior or both
- Requires
  - Narrow LVOT
  - Hypercontractile LV
  - Redundant MV leaflets
- Severity of LVOT o is related to the duration and degree of SAM and to the reduction in LVOT size
- Decreases stroke volume
- Raises ventricular pressures creating more hypertrophy
Systolic Anterior Motion by M mode
HCM and SAM
Color alias pattern of SAM, suggests area of LVOTo
HCM and SAM

Lossy compression - not intended for diagnosis
HCM, SAM and MR
Doppler Echocardiography
Doppler Profiles in HCM

LVOTo from SAM
- Later-peaking than MR jet
- Delayed onset
- Estimate LVOTo
  - Peak LVOTo = 4 (MR systolic jet velocity)^2 + LA p - SBP

Mid-cavity obstruction
- Steep, dagger shaped,
- Latest peak
- Short duration

Mitral Regurgitation
- Posteriorly directed
- Earlier onset
- Longer duration
- Greater magnitude than SAM jet
- Earlier peak (first third of systole)
Doppler Trio in HCM
Intracoronary Coronary Contrast Alcohol Septal Ablation
Septal Motion Following ASA

Lossy compression - not intended for diagnosis

POST ABLATION
Restrictive CMP: Infiltrative or Endomyocardial Diseases
Restrictive CMP
Echo Findings

- Hypertrophy

- Enlarged atria
  - Consequence of poor ventricular compliance
  - Atrial fibrillation common

- Restricted ventricular filling
  - Remember the myocardium is involved primarily
  - Annular E’ is markedly reduced (always < 8)

- Elevated filling pressures
Echocardiographic Findings in Amyloid

- Normal to small LV cavity size
- Thickened LV and often RV walls
- Speckling not specific
- Infiltration into valve structures
- Enlarged atria
- Pericardial effusion in up to a third
- High EF but low stroke volume
- Elevated filling pressures
- Restrictive filling
Amyloid Most Common
Amyloid
Amyloid
Significant TR / MR
LV Non-Compaction

- Not yet in WHO as a form of CMP
- Abnormality of ventricular morphogenesis
- Unique, rare cause of CHF, lethal arrythmias, and systemic embolization
- Identifiable non-invasively
Echocardiographic diagnostic Criteria in LVNC

- Absence of other coexisting cardiac abnormalities
- Thin, compacted, epicardial tissue layer and a thick spongy, endocardial, non compacted layer with extensive trabeculations and sinusoids
  - End-systolic ratio of compacted to non-compacted layers of > 2 discriminates LVNC from HCM, HTN, etc
- Deep intratrabecular recesses that communicate with LV cavity
- Localization in the apex, mid lateral, or mid-inferior walls
- Local or global LV dysfunction
- RV can be involved but difficult to diagnose (normally trabeculated)
LV Non-Compaction
Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

- Genetic CMP: desmosomes fail with mechanical stress
  - M:F 1:3
  - Familial occurrence 30-50%
  - Dominant: desmoplakin, ryanodine
  - Recessive: Naxos-keratosis
- Pathologic fibrofatty infiltration of the RV free wall

- Lethal arrhythmias in young patients

- Clinical Criteria for diagnosis
  - No gold standard
  - Clinical symptoms, ecg, structural changes
Echocardiographic Findings in ARVC

- **Regional RV dilatation (89%)**
  - PSLA view RVOT > 3.2 cm
  - PSAX RVOT > 3.6 cm

- **Morphologic RV abnormalities**
  - Trabecular derangement (54%)
  - Moderator band hyperreflectivity (34%)
  - Focal RV saculations or aneurysms (17%)

- **Abnormalities in Regional RV function (62%)**
  - 60% of patients with normal RV function had identifiable structural abnormalities
ARVC

A4C view (modified)

Subcostal view

PLAX

PSAX
ARVC

22 yr. male ballet dancer with syncope

normal subject
22 yr. male ballet dancer with syncope

[Images of echocardiograms with labeled anatomical structures: RA, LA, LV]
ARVC

Lossy compression - not intended for diagnosis
<table>
<thead>
<tr>
<th>Echo Features</th>
<th>Hypertrophic cardiomyopathy</th>
<th>Dilated cardiomyopathy</th>
<th>Restrictive cardiomyopathy</th>
<th>Arrhythmogenic right ventricular cardiomyopathy</th>
<th>Isolated ventricular non-compaction</th>
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<tbody>
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<td>↑</td>
<td>N</td>
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LV, left ventricular; LVEDD, left ventricular end diastolic diameter; N, normal; RV, right ventricular