Diastolic Dysfunction: In Myocardial Diseases: Hypertrophic Cardiomyopathy

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Diastolic Dysfunction: HCM

Objective:

Review clinical utility of diastolic function assessment in daily practice:

Clinical Manifestations

- Sudden Death
- Symptoms
- Genetics
- Arrhythmia
- MR
- Obstruction
- ischemia
- Fibrosis
- Baroreceptor dysf.
- Diastolic dysf.
- Microvasc. Dym.

Diastolic Dysf.
Pathophysiology of Diastolic Dysfunction: HCM

- Prolonged Relaxation
- Fibrosis
- Chamber Stiffness
- Loss of Suction
- Twist / Untwist

Diastolic function: HCM

- Screen for preclinical disease
- Distinguish HCM from athlete’s heart
- Determine left atrial pressure
- Discuss what variables affect exercise
- Discuss HCM subtype - “restrictive filling”
Preclinical Diagnosis HCM

26 yr. old woman with a FH of HCM (father has HCM with ICD - diagnosed age 35). She is asymptomatic and has a normal EKG.

Screening for Preclinical HCM

- Screening Family Members
- EKG
- Echo
- Gene Testing

Screening for Preclinical HCM

- Familion/ PGxHealth
- Gene Dx
- Correleagen
Screening for Preclinical HCM

Limitations Genetic Testing in 2012:
- Only ~60% (+) tests (with suspected HCM) - uncertain significance of (-) test
- Testing may be expensive / not covered by insurance
- Uncertain implications on jobs, insurance, etc

Preclinical Diagnosis HCM

Preclinical Diagnosis HCM
Preclinical Diagnosis HCM

Lat Eann – 8 cm/sec
Lat Ann – 5 cm/sec
Lat Sann – 9 cm/sec

What does this mean?

Findings suggest:
Phenotype (-) and
Genotype (+) HCM

Screening for Preclinical HCM

TDI:
100% sensitive
90% specific
for mutation (+) pts.
with HCM but no LVH
(Ea < 14 cm/ sec)

Circulation 2001;104:128
Screening for Preclinical HCM

**TAKE HOME POINTS**

- Important to assess diastolic function and TDI annular velocities when screening for HCM
- Abnormal TDI annular velocities may be the earliest marker of preclinical disease
- If preclinical disease detected - more frequent surveillance should be undertaken

Diastolic function: HCM

- Screen for preclinical disease
- *Distinguish HCM from athlete’s heart*
- Determine left atrial pressure
- Discuss what variables affect exercise
- Discuss HCM subtype - “restrictive filling”

Distinguish HCM from Athlete’s Heart

**What do these 2 athletes have in common?**

- a) Play hockey with Dr. Klein
- b) Same training program
- c) Eat the same diet
- d) None of the above

Distinguish HCM from Athlete’s Heart

What do these 2 athletes have in common?

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Maron, NEJM 2003349:1064

Distinguish HCM from Athlete’s Heart

19 yr. old football player. One episode of nonexertional chest pain. No FH SCD or HCM
Distinguish HCM from Athlete’s Heart

LVIDd-5.6, LVIDs-3.4, IVS-1.3cm, PW-1.2cm
Distinguish HCM from Athlete’s Heart

Pt. exercised 19 METS
No sxs.
No EKG changes / arrhythmias
No regional abnormalities
No LVOT obstruction
No hypotension

Distinguish HCM from Athlete’s Heart

Diastolic Function

E/A ratio - 1.5
Color M-mode – Vp - 80 cm/ sec
E ann – 16 cm/ sec
S ann – 11 cm/ sec
E/E ann - 3.5

What does this mean?

Distinguish HCM from Athlete’s Heart

Diastolic Function

E/A ratio - 1.5
Color M-mode – Vp - 80 cm/ sec
E ann – 16 cm/ sec
S ann – 11 cm/ sec
E/E ann - 3.5

Findings c/w Athlete’s Heart
**Distinguish HCM from Athlete's Heart**

### Performance of echocardiographic criteria in differentiating between pathologic left ventricular hypertrophy and control

<table>
<thead>
<tr>
<th>Sensitivity</th>
<th>Specificity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Longitudinal strain cutoff &lt; 20.3%</td>
<td>93%</td>
</tr>
<tr>
<td>Longitudinal SRS cutoff &lt; 1.17s⁻¹</td>
<td>97%</td>
</tr>
<tr>
<td>Early diastolic annular velocity &lt; 9 cm/s</td>
<td>85%</td>
</tr>
<tr>
<td>Systolic annular velocity &lt; 9 cm/s</td>
<td>66%</td>
</tr>
</tbody>
</table>

Saghir M, JASE 2007;20:151-157

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**Athletes** | **HCM**
---|---
Longitudinal Base (%) | 15.0 ± 3.6 | 11.5 ± 5.7 |
Mid-LV (%) | 16.4 ± 3.0 | 13.5 ± 5.1 |
Apex (%) | 18 ± 4.0 | 21.5 ± 8.1 |

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**Transverse**

| Base (%) | 35.4 ± 22.2 | 25.7 ± 19.8 |
| Mid-LV (%) | 32.8 ± 13.4 | 21.8 ± 15.0 |
| Apex (%) | 10.5 ± 19.9 | 21.3 ± 15.5 |

Richard V, Am J Cardiol 2007;100:128

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Asher CR, "Echocardiographic Profiles of Disease Associated with SCD in Young Athletes", The Athlete and Heart Disease, Lippincott 1999
Distinguish HCM from Athlete’s Heart

**TAKE HOME POINTS**

- The remodeling of athlete’s heart may mimic HCM
- Several criteria aid to distinguish the 2 conditions
- Any abnormality of diastolic function is not consistent with athlete’s heart

Diastolic function: HCM

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- Determine left atrial pressure
- Discuss what variables affect exercise
- Discuss HCM subtype - “restrictive filling”

Determine left atrial pressure in HCM

Nagueh, JACC 1997;30:1527
Determine left atrial pressure in HCM

Echo:
IVS-2.1 cm; PW-1.5 cm
LVOT resting gradient = 70 mmHg with SAM; 2+ MR
Determine left atrial pressure in HCM

Diastolic Function:
- E/e' = 10 (equivocal range)
- AR - A = 35 msec
- PASP = 40 mmHg
- LA volume = 39 ml/m2

↑ LAP

100 patients with HCM:
- Age: 58 ± 13
- Male: 49%
- FC III-IV: 82%
- Wall thickness: 20 ± 5.5 mm
- LVOT grad (rest): 53 ± 47 mmHg
- LA size: 48 ± 18 cc/m²

42% simultaneous hemodynamics including direct LAP — remainder of studies within 48 hrs

Circulation 2007:116:2702
Determine left atrial pressure in HCM

ALL PATIENTS

HCM

R=0.87

R=0.28

Low E/e’ is very uncommon

25% of HCM pts with E/e’ > 15 had LAP < 15 mmHg

Poor Correlation between E/e’ and BNP in HCM pts

Circulation 2007:116:2702

JASE 2011:24:1020
Determine left atrial pressure in HCM

**TAKE HOME POINTS**

- E/e' has a relatively weak correlation with left atrial pressure in pts with HCM

Diastolic function: HCM

- Screen for preclinical disease
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- Determine left atrial pressure
- **Discuss what variables affect exercise**
- Discuss HCM subtype - “restrictive filling”

What variables affect exercise in HCM

38 yr old woman with HCM
Asymptomatic with good exercise capacity!

ECHO:
- IVS-2.1 cm; PW-1.5 cm
- LVOT resting gradient = 70 mmHg with SAM, 2+ MR
What variables affect exercise in HCM

Exercise Capacity

- MR
- Obstruction
- Ischemia
- Arrhythmia
- Diastolic
  dysf
- Inappropriate
  Vasodilation

Exercise SV and LAP

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63 pts HCM:
Exercise echo / PeakVO$_2$

Predictors of exercise capacity:
- Shape / degree hypertrophy
- LVOT gradient
- Diastolic function

Finding:
LA volume index – best predicts exercise capacity

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93 pts HCM
Exercise metabolic EKG
Resting Echo

Predictors of peakVO$_2$
assessed:
Clinical; GXT; Echo

Finding:
Multivariate model:
- LA volume index
- Resting HR; BMI
- LVIDs

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Am Heart J 2009; 158:e27

JASE 2005;18:1373
What variables affect exercise in HCM

85 pts with HCM:
Relation of symptoms and exercise capacity to diastolic function

Finding:
E'/e' predicts NYHA class and PVO₂

Heart 2002;87:247

What variables affect exercise in HCM

High E'/e' and LAV index Predicts Cardiac Events in HCM

AJP 2011;108:1614

What variables affect exercise in HCM

**Take Home Points**

- E'/e' has a good correlation with symptoms and exercise capacity
- LA volume index as a reflection of chronic adverse loading conditions due to diastolic dysfunction and MR predicts exercise capacity
- Still, much is unknown about the variable that affect exercise capacity
**Diastolic function: HCM**

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**HCM Subtype of Restrictive Filling Pattern**

62 yr old man with dyspnea
1,226 patients with HCM
19 (1.5%) with Restrictive Phenotype
Age: 42 ± 19 yrs; Female: 58%
Symptoms at presentation: 84%
History AF: 74%

JACC 2007;49:2419
HCM Subtype of Restrictive Filling Pattern

• Restrictive phenotype associated with minimal hypertrophy without LVOT obstruction and restrictive filling pattern
• Poor prognosis with high prevalence of CHF, AF, stroke, death and need for transplant
• Common finding of β-MYH and Troponin I mutations

Diastolic dysfunction is ubiquitous in HCM
• Comprehensive diastolic function assessment is essential to screen for HCM, distinguish HCM from AH and predict exercise capacity
• Determination of LAP with E/ e' has a very modest utility in HCM
• The “Restrictive phenotype” is important to recognize since the prognosis is poor