DIFFERENTIATING PHYSIOLOGIC V. PATHOLOGIC LV HYPERTROPHY

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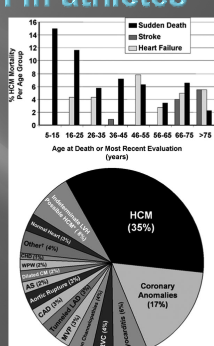
Goals of presentation

- Identify increased LV wall thickness (WT)
- Understand increased WT in athletes
- Understand hypertrophic cardiomyopathy (HCM)
- Enhance understanding of various echo modalities to differentiate increased WT in athlete's heart v. HCM
  - 2D/M-mode
  - Doppler
    - Tissue, color, strain, strain rate imaging, speckle

Sudden death in athletes



Contemporary insights and strategies for risk stratification and prevention of sudden death in hypertrophic cardiomyopathy.

Maron BJ
Circulation. 121(3):445-56, 2010 Jan 26.

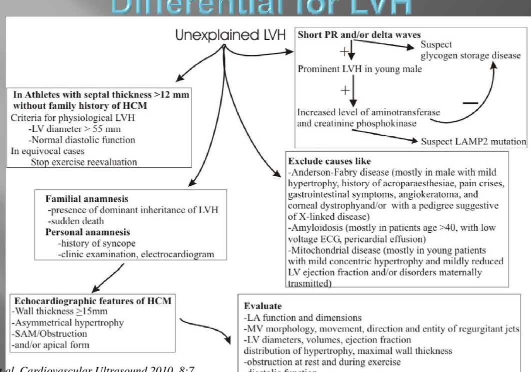
Beaumont Healthy Heart Check

- Cardiac directed history form
- EKG
- Cardiac directed physical with BP
- Algorithm for possible abnormalities
- "Quick look" echo
  - Ventricular function
  - LVH
  - Coronary artery anatomy
- Recommendations for follow-up evaluation and risk stratification to stop sports

EKG criteria for LVH

- Voltage criteria
  - Sokolow-Lyon
    - Sum of S-wave in V1 and R-wave in V5 >3.5 mV
  - Romhilt-Estes Point score
  - R-wave in V5 or V6 > 30 mV
- False (+) EKG
  - Italian study, *Pelliccia et al, JAHA, 102(3), 278-284, 2000*
  - 1005 athletes (24 $\pm$ 6y) in 38 sport disciplines
  - 14% markedly abnormal, 26% mildly abnormal
    - 5% considered "bizarre" with no structural abnormality

Differential for LVH

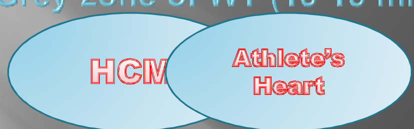


Losi et al. Cardiovascular Ultrasound 2010, 8:7

Athlete's heart

- Morphologic and electrical remodeling
- In Caucasian athletes
  - Males rarely > 12 mm
  - Females all < 11 mm
- In black athletes
  - Small minority >= 15 mm without other pathology
    - *Basavarayaiah,S, Ethnic differences in LV remodeling in highly trained athletes. JACC 2008;51:2256-62*

Grey-zone of WT (13-15 mm)



HCM		Athlete's Heart
+	Unusual pattern of LVH	--
+	LV cavity < 45 mm	--
--	LV cavity > 55mm	+
+	Marked LA enlargement	--
+	Bizarre EKG patterns	--
+	Abnormal LV filling	--
+	Female	--
--	Decrease in thickness after deconditioning	+
+	Family Hx of HCM	--
--	Max VO2 > 45 ml/kg/min	+

Maron Cardiac disease in young trained athletes Circ 1995;p 91:1595-601

LV wall thickness in elite adolescent athletes

- 720 elite adolescent athletes (EAA)
- 14-18 yo, 15.7 $\pm$ 1.4 yr
- 75% male
- 98% Caucasian
- 2D echo at height of competitive season
- 50% national level athletes
- 9.8 $\pm$ 3.6 hr/week training
- 46% soccer and tennis
  - boxing, cycling, field hockey, karate, rowing, rugby, swimming and triathletes

Sharma et al. JACC 2002;40:1431-6

LV wall thickness in elite adolescent athletes

- Control group-age match, gender, BSA
  - Sedentary
  - < 2 hr/week of organized activity
- LVWT measured in parasternal short axis at ED
  - Maximal measurement recorded

Sharma et al. JACC 2002;40:1431-6

LV wall thickness in elite adolescent athletes

- RESULTS:
- Athletes had significantly greater
  - LV Wall thickness (13%)
  - EDD (11%)
  - LV mass (6%)
  - LA diameter (5%)
- Of 720….38 (5%) had LVWT > predicted upper normal limits (11mm)
  - Rowing, soccer, swimming
- All females had wall < 11 mm

Sharma et al. JACC 2002;40:1431-6

LV wall thickness in elite adolescent athletes

- Of 38….
  - All concentric (symmetrical)
  - No change > 2mm in LVWT between 2 contiguous wall segments
  - All had > than predicted LVEDD cavity size
    - 54.4 ± 2.1 mm (52-60 mm)
  - All had normal mitral inflow velocity patterns
- 82% of above 38 had EKG LVH
- In all study, 33% had LVH criteria by EKG but normal LVWT
  - No deep T waves, Q > .04 s, no ST segment depression

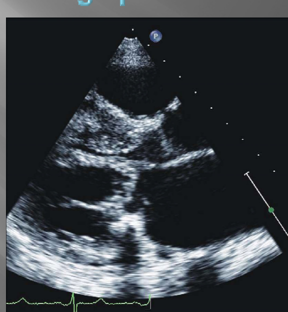
Sharma et al. JACC 2002;40:1431-6

HCM-2D echo

- Asymetrical septal hypertrophy
  - Septal thickness > 15 mm
  - Septal to posterior free wall ratio > 1.3 (IVS/PW)
  - May be seen in RV hypertension, systemic hypertension, aortic stenosis, septal sarcomas, Fabry disease, Freidrich's ataxia, amyloidosis
- SAM
  - 98% specificity
  - May be seen in TGA, hypercontractile states, anomalous papillary m. insertion and anteroapical infarct

Echo HCM in 6 mo

Abnormal insertion of papillary m. causing "pseudo SAM"



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HCM-2D echo

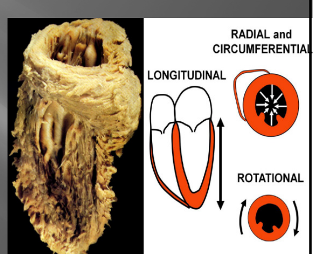
- LVOT
  - Fluttering of aortic valve on m-mode
  - Fibroseptal changes at level of leaflet-septal contact
  - Increased gradient with Valsalva or exercise
- Systolic function
  - Hyperdynamic or normal
  - Shortening fraction
    - Radial contractile shortening
  - EF still preserved despite abnormal longitudinal contractile function as seen d/t attenuation of annular velocities (strain and strain rate)

TDI in geno+/pheno- HCM

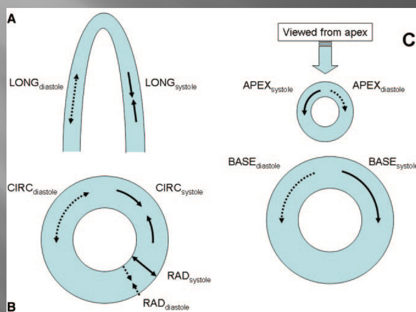
- Systolic (Sa) and diastolic (Ea) myocardial velocities are decreased in genotype+/phenotype-
  - *Nagueh. Circ. 2003;108:395-398*
  - 12 pts (17-51)
  - No LVH by echo
  - Known mutation for HCM
  - F/U echo shows increased mean septal thickness and LV mass
  - Lateral Sa (15$\pm$ 1.2 v. 8.2 $\pm$ 2.1 cm/s) and lateral Ea (16.5 v. 8.1) mutation v. controls
  - 6 pts diagnosed with HCM within 2 years

Tissue Doppler imaging

- Nice review in normal children
  - Eidem, JASE 2004
- High amplitude, low velocity signals
- Real time quantification of myocardial function
  - Longitudinal or axial

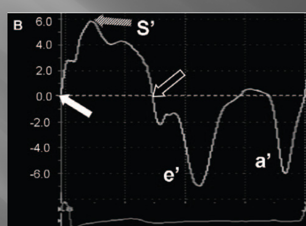


Principles of deformation



Abraham, Circulation 2007, 116:2597-2609

Tissue velocity



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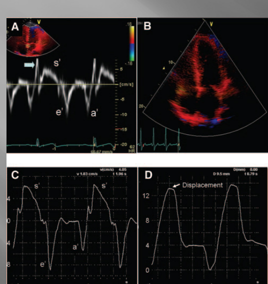
Pulse wave tissue velocity



Tissue velocity tracing tracks myocardial motion at the point of interrogation.

Pulsed Doppler (A) yields myocardial motion only at the region of interest.

In this example, the sample volume is located in the basal septum off an apical 4-chamber view.

Color TDI collects tissue velocity from the entire sector(B).

With color TDI, the region of interest can be placed anywhere in the sector to yield tissue velocities (C).

In the longitudinal orientation (apical views), the base moves toward the apex in systole and depicts a positive polarity (s) and moves away from the apex to yield the early (e) and late (a) negative diastolic velocities.

Integration of the tissue velocity signal yields displacement (D) that indicates the distance moved by that

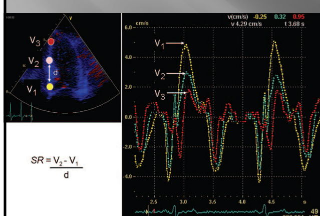
1. point.

Abraham, Circulation 2007, 116:2597-2609

TDI in HCM v. athlete's heart

- Differentiation Between Pathologic and Physiologic Left Ventricular Hypertrophy by Tissue Doppler Assessment of Long-Axis Function in Patients with Hypertrophic Cardiomyopathy or Systemic Hypertension and in Athletes
  - *Vinereanu. Am J Cardiol 2001;88:53–58*
  - Mean systolic annular velocity < 9 cm/sec (87% specificity/97% sensitivity) for HCM
  - Mean early diastolic annular velocity < 9 cm/sec
- Doppler Tissue Imaging: Regional Myopcardial Function in Hypertrophic Cardiomyopathy and in Athlete's Heart
  - *Cardim et al. JASE. 2003. 16(3);223-232*
  - No athlete's heart with regional *e/a* < than 1

Tissue velocity apex to base



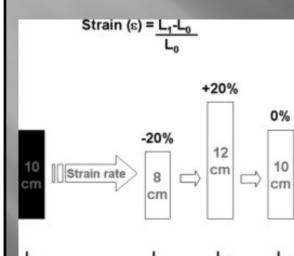
In the longitudinal orientation, normal heart motion is such that the base moves toward the apex, which moves little or not at all. Thus, tissue velocity is maximum at the base (V1), lower in the mid heart (V2) and least at the apex (V3). This gradient in velocities is used to calculate strain rates.

Strain rate is calculated with tissue Doppler as the difference between 2 tissue velocities along the ultrasound beam (V2V1) normalized to the intervening distance between these 2 velocities (d).

Colored circles indicate the positions of the region of interest in the myocardium (left) for the corresponding tissue velocity traces in the right panel

Abraham, Circulation 2007, 116:2597-2609

Strain and Strain rate



Strain measures tissue deformation and is defined as the change in dimension or length (L1L0) normalized to the initial length (L0) of the region of interest.

For example, if the initial length of a myocardial segment is 10 cm, then shortening it by 2 to 8 cm indicates a strain of 20%.

Likewise, a lengthening of the segment to 12 cm indicates a strain of 20%.

No change in length would suggest 0% strain.

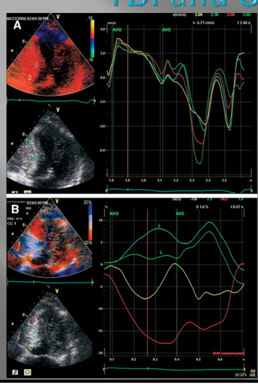
The rate at which any of these length (dimension) changes occur is strain rate.

Abraham, Circulation 2007, 116:2597-2609

Strain/Strain Rate

- Identify heterogeneity in contractile function
- Improves understanding of mechanic in HCM
- Normal in Athlete's Heart
  - *Saghir,M. Strain Rate Imaging Differentiates Hypertensive Cardiac Hypertrophy from Physiologic Cardiac Hypertrophy (Athlete's Heart). JAmSoc Echocardiogr 2007; 20:151-157*
  - No significant difference in strain, SR_S, SR_E or peak late diastolic strain rate

TDI and Strain in HCM



Myocardial tissue Doppler velocity tracings from 4 representative regions of interest (ROIs) in a patient with HCM.

Note significantly attenuated systolic and early diastolic velocities from disparate ROIs in the septum.

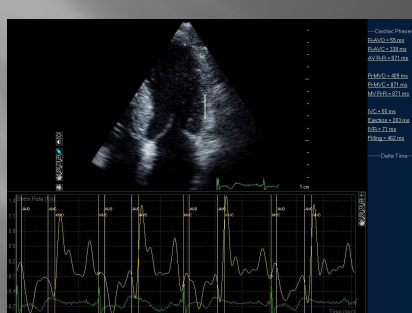
(B) Tissue Doppler-derived longitudinal strain curves in the same areas shown in (A) and corresponding parametric color strain map. Note positive longitudinal strain (systolic lengthening) or "paradoxical strain" (blue and green tracings) in 2 of the 4 depicted ROIs (basal septum) and attenuated longitudinal strain elsewhere (yellow tracing).

Note striking heterogeneity of strain tracings in contrast to tissue Doppler data.

AVO aortic valve opening, AVC aortic valve closure

Afonso et al. Echocardiography Evaluation of HCM. JACC. 2008;1 (6) 787-800

Post-systolic strain in HCM



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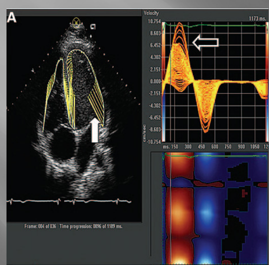
2D speckle tracking in healthy heart



- Arrow direction is directions and length is amplitude of motion
- One systolic peak
- Synchronous color m-mode

Afonso et al. Echocardiography Evaluation of HCM. JACC. 2008;1 (6) 787-800