Basal Septal Hypertrophy: Normal Variant or HCM?

Dr Lynne Williams
Consultant Cardiologist
Echocardiography and Inherited Cardiovascular Conditions

INTRODUCTION

Localized thickening of the basal portion of the ventricular septum has been identified by imaging studies for decades

No consensus on its significance

Remarkable lack of consistency in terminology

INTRODUCTION

- Early studies - BSH was a non-pathologic result of aging or of uncoiling of the aorta
- Associated ageing, aortic stenosis, and hypertension
- Recent echo- Doppler studies show that BSH can be associated with dynamic outflow tract obstruction
- Asymmetric thickening of the ventricular septum compared to the posterior wall of the LV has been recognized as a marker for HCM
- The distinction between isolated BSH and HCM can be challenging - important implications for family screening

ANATOMICAL CHANGES WITH AGEING

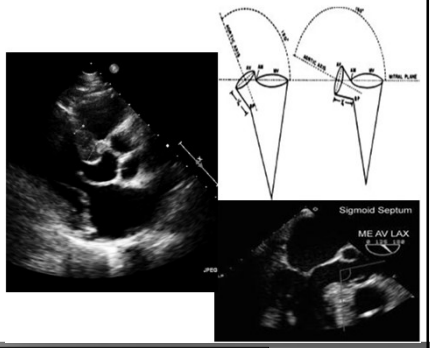
Age-related dilatation + lengthening of the aorta

Rightward shift of dilated ascending aorta

Basal portion of ventricular septum bends leftward and bulges into the LVOT

↑ angle aortic: mitral
↓angle aorta: septal

Small left ventricular cavity



Goor D et al. Am J Roentg Radium Ther Nucl Med 1969;107:366
Waller BF et al. Mayo Clin Proc 1988;63:625–627

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DEFINITION OF BASAL SEPTAL HYPERTROPHY

Upper interventricular septum thickness ≥1.4 cm

Upper septum thickness/mid-septum thickness ratio ≥1.3

Posterior wall thickness <11mm

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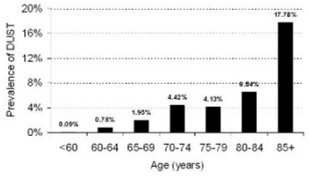
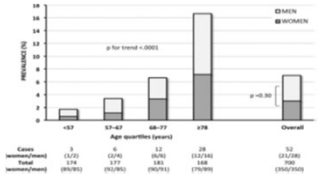
PREVALENCE OF BASAL SEPTAL HYPERTROPHY





Framingham Heart Study (n=3562;age 58yrs)

Prevalence 1.5%

Clinical correlates:

- ↑age (OR /10 year increment 2.59)
- systolic BP (OR 1.55)

Not associated with CVD or mortality risk

Baltimore Longitudinal Study of Ageing (n=700;age 64yrs)

Prevalence 7.0%

Clinical correlates:

- ↑age (OR /year increment 1.06)
- ↑aortic root dimension

Diaz T et al. Echocardiography 2009;26:247–253
Canepa M et al. Am J Cardiol 2014;114:796-802

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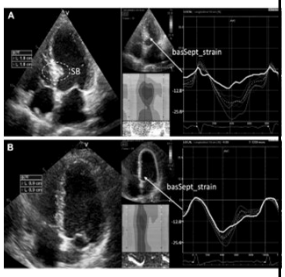
BASAL SEPTAL HYPERTROPHY - EARLY MARKER OF HYPERTENSIVE HEART DISEASE?

- N=110 – no history of and no Rx for HTN
- BSH in 43.6% (basal-septal WT ≥2 mm more than mid-septal WT)
- BSH group - 79.2% showed HTN by either by CET or ABPM
- No BSH group - 95.2% no HTN by CET or ABPM
- Regional myocardial deformation of the basal-septum in BSH group significantly lower than in no-BSH group (14 ± 4% vs. 17 ± 4%; *P* < .001)



Gaudron P et al. J Am Soc Hypertens 2016;10:70-80

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BSH AND LVOT OBSTRUCTION

- LVOT obstruction has been classically observed in HCM
- Basal septal hypertrophy may result in dynamic LVOTO
- LVOTO in HTN Initially described by Brock in 1957*
- Provocable LVOT gradients were shown in a group of elderly with protruding angulated septum similar to that in BSH with HHD**
- Dynamic LVOTO can be provoked in conditions similar to HCM

ENHANCED CONTRACTILITY
Upright exercise
Dobutamine

REDUCED PRELOAD
Upright posture
Sublingual Nitroglycerin
Valsalva maneuver

VASODILATOR STRESS
Amyl nitrite
Sublingual Nitroglycerin

PROVOCABLE OBSTRUCTION

**Brock et al. Guy's Hosp Rep 1957;106:221-238*
***Henein MY et al. JACC 1997;30:1301-1307*

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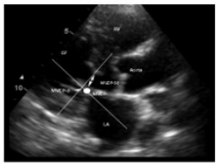
BSH AND LVOT OBSTRUCTION

Ranasinghe et al n=4104
5.8% had BSH; 3.3% resting LVOTO
Not related to basal septal thickness
Related to hyperdynamic LV
Displacement of MV coaptation (apical/septal)



Cha et al n=168
8% sigmoid septum/51% sigmoid septum + BSH
20% resting LVOTO; 80% dynamic LVOTO (Valsalva)

Ranasinghe I et al. Int J Cardiol 2014;172:487-493
Cha JJ et al. Cardiovasc Ultrasound 2014:12:23

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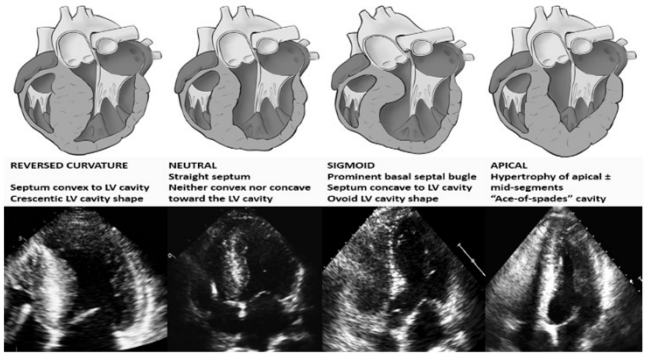
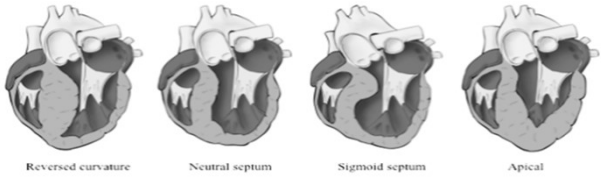
HCM AND SEPTAL MORPHOLOGY



HCM AND SIGMOID SEPTUM



Mayo clinic study n=400pts
47% had sigmoid septum
10% had +ve genetic test
80% of reverse curvature HCM patients had a +ve genetic test

Young vs old HCM cohort
BSH in 54% of elderly HCM /4% reverse curvature
Prevalence of SAM/LVOTO similar between groups

Binder J et al. Mayo Clin Proc. 2006;81:459-467
Lever H et al. Circulation. 1989;79:580–589

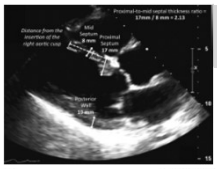
DISTINGUISHING BSH FROM HCM

BSH common with advanced age, often with a history of HTN and mitral annular calcification
Some HCM variants present at an advanced age
Importance of diagnosis – implications for family screening

Accurate diagnosis involves integration of multiple parameters:
Family history (helpful if +ve for HCM/SCD)
ECG appearances (LVH 60-70% HCM; ?12% in BSH)
History of symptoms
Imaging appearances (echo + CMR are complimentary)

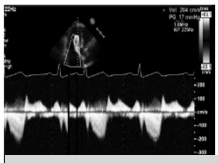
2014 ESC Guidelines on diagnosis and management of HCM

DISTINGUISHING BSH FROM HCM

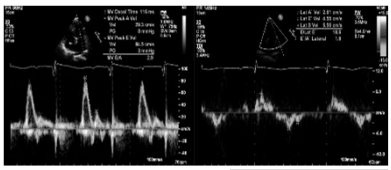


BSH ≥14mm; HCM ≥15mm
Basal-mid-septal ratio ≥ 1.3
PW < 11mm

AORTO-SEPTAL ANGLE
<110° in BSH; >110° in HCM
<100° - 91% specificity for BSH

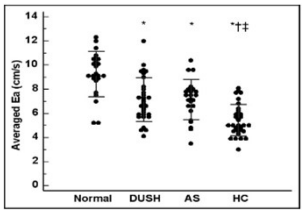


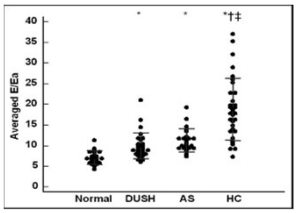
LVOTO
<30mmHg at rest in BSH
>30mmHg at rest in 1/3 of HCM



DIASTOLOGY
E/A ratio <1.0 in BSH
E/E' ≥13 in HCM

DIASTOLIC FUNCTION AND LEFT ATRIAL SIZE





Aging is associated with worsening LV diastolic function:
- ↑ventricular mass/collagen infiltration/altered calcium handling

Diastolic dysfunction is hallmark of HCM
E' velocities are ↓in HCM > than in BSH; E/E' ratio in HCM > than in BSH
E/E' ratio ≥ 13 has 78% sens/90% spec for distinguishing HCM from BSH

Chen-Tournoux A. Am J Cardiol 2008;101:1498-1503

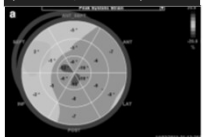
DISTINGUISHING BSH FROM HCM

Measurements of subtle LV systolic dysfunction by STE may help differentiating between HCM and other forms of LVH

Patients with HCM have abnormalities in regional and global longitudinal strain - related to the site and degree of hypertrophy

Patients with sigmoidal HCM have more abnormal strain in the proximal septum than in other LV segments

BSH with normal LV mass
(BS 13mm, PW 9mm)



HCM with ASH
(BS 18mm, PW 12mm)

Liu et al. Eur J Med Res (2016) 21:21

CONCLUSIONS

Sigmoid septal morphology and basal septal hypertrophy are common with advancing age

Represent a diagnostic challenge due to overlap with appearances of HCM

A integrative approach utilising family history, ECG, and imaging appearances is required

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