Sports Medicine

A Look at Echo and an Update on Hypertrophy

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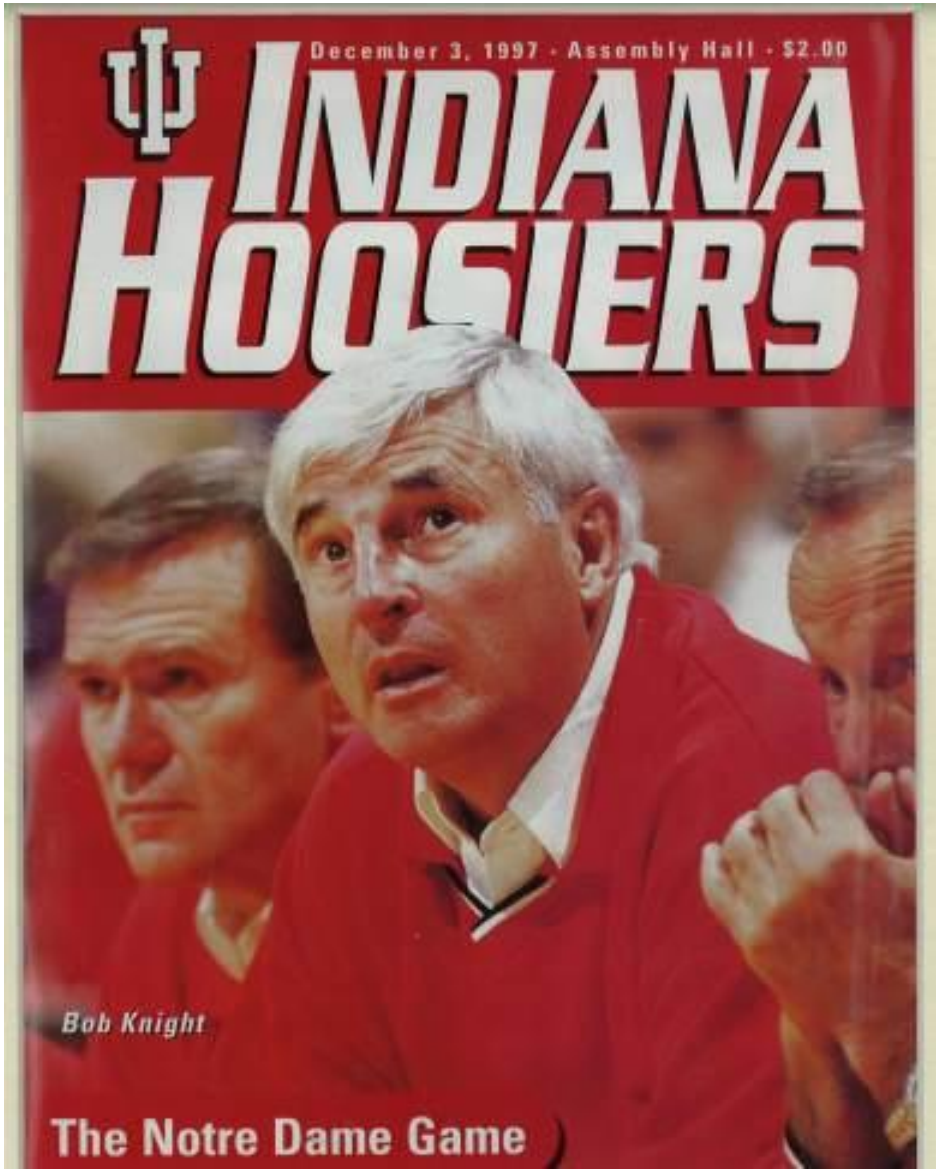

December 3, 1997 · Assembly Hall · $2.00
INDIANA HOOSIERS
Bob Knight
The Notre Dame Game

Sudden Death in Athletes

- Incidence of sudden death 1:200,000 – underestimated
- Risk is very low
- Tragedy rate very high
- Males dominate
- Basketball and soccer dominate
- Loss of life within one hour of symptoms

50% have symptoms before death

Causes of Sudden Death in 387 Young Athletes

Cause	# of Athletes	Percent
Hypertrophic cardiomyopathy	102	26.4
Commotio cordis	77	19.9
Coronary artery anomalies	53	13.7
Left ventricular hypertrophy	29	7.5
Myocarditis	20	5.2
Ruptured aortic aneurysm (Marfan syndrome)	12	3.1
Arrhythmogenic right ventricular cardiomyopathy	11	2.8
Tunneled (bridged) coronary artery	11	2.8
Aortic valve stenosis	10	2.6
Artherosclerotic coronary artery disease	10	2.6

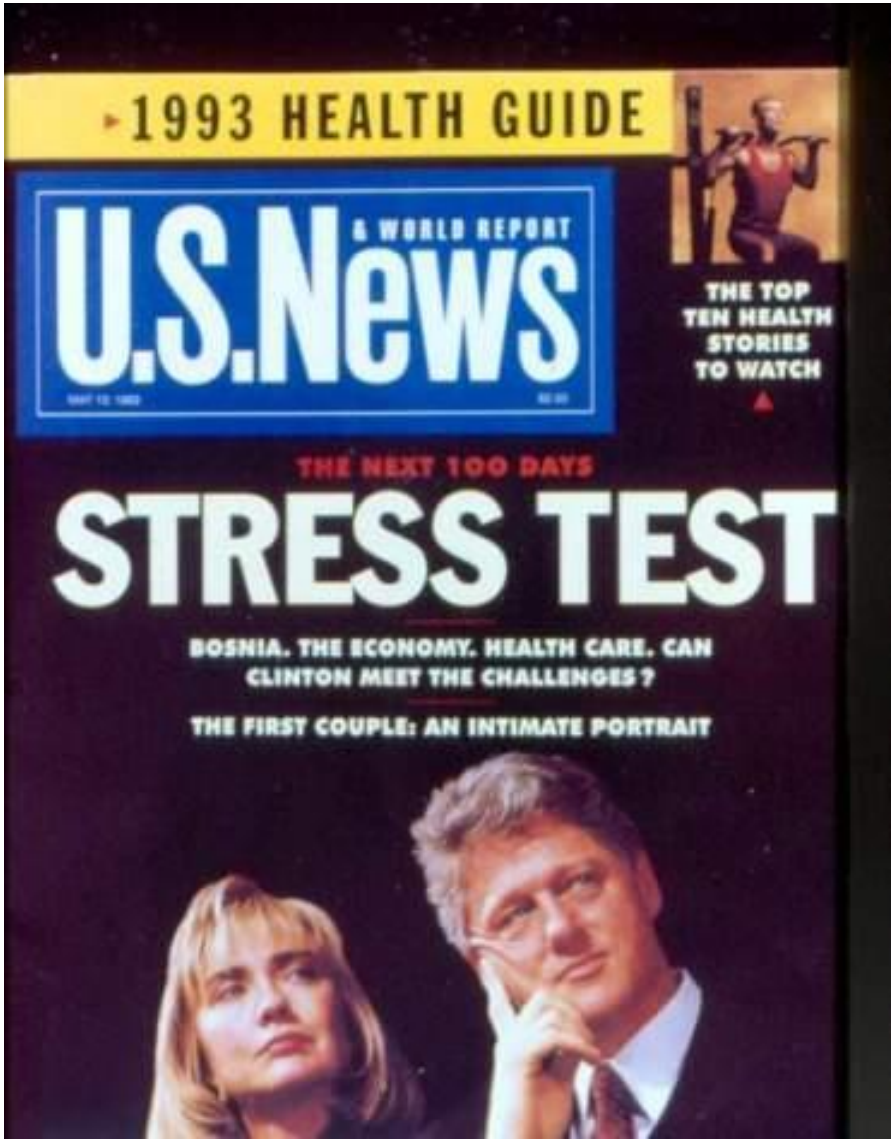
Maron, et al. JACC. 2005, 45,8:1318-21

Component of Preparticipation Examination

- Required
  - History
  - Physical exam
  - Determination of eligibility
- Recommended
  - CBC
  - UA
  - Sickle cell screening
  - Echocardiogram
  - Education

Component of Preparticipation Examination

- May be helpful but not routinely recommended
  - EKG
  - Stress test with echocardiogram or nuclear
  - HIV testing, chemistry profile
  - Strength measurements
  - Maximum O_2 consumption and anaerobic threshold
- Not helpful
  - More detailed blood tests
  - Stress test without echocardiogram


1993 HEALTH GUIDE
U.S.News
& WORLD REPORT
THE TOP TEN HEALTH STORIES TO WATCH
THE NEXT 100 DAYS
STRESS TEST
BOSNIA. THE ECONOMY. HEALTH CARE. CAN CLINTON MEET THE CHALLENGES?
THE FIRST COUPLE: AN INTIMATE PORTRAIT

IMA Sport Screening Echocardiograms 1988-2008

- 5,500 High School Freshmen
- 1,500 College Athletes
- 800 other athletes
- Time: 2.5 minutes per Athlete
- 3 Technicians
- 2 Additional Support Personnel
- 1 Cardiologist

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IMA Sport Screening Echocardiograms 1988-2008

- 7,800 athletes ages 12-25
- 11% have some abnormality
- ~3% need some form of cardiac follow up
- 0.5% disqualified for cardiovascular abnormalities

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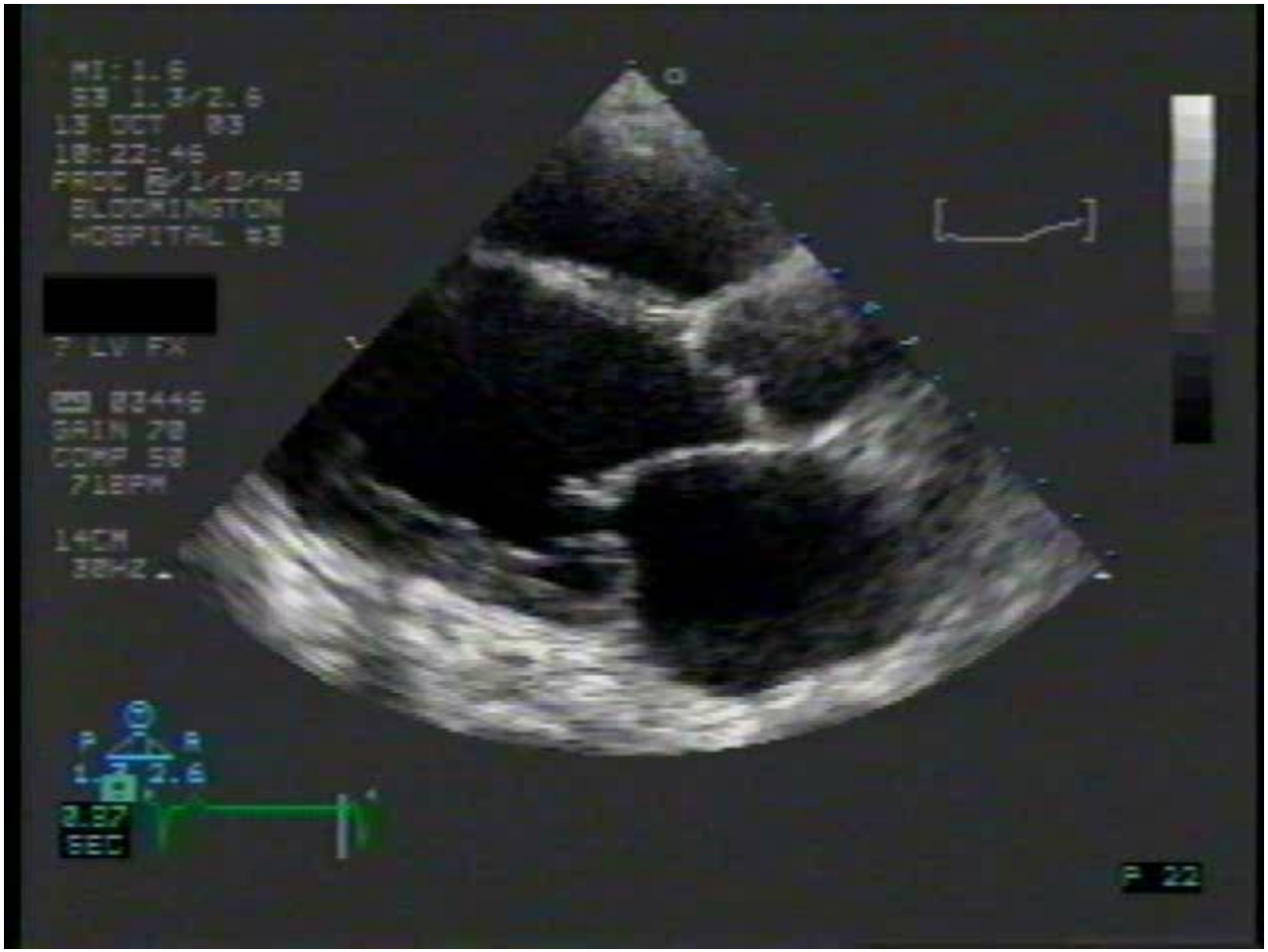
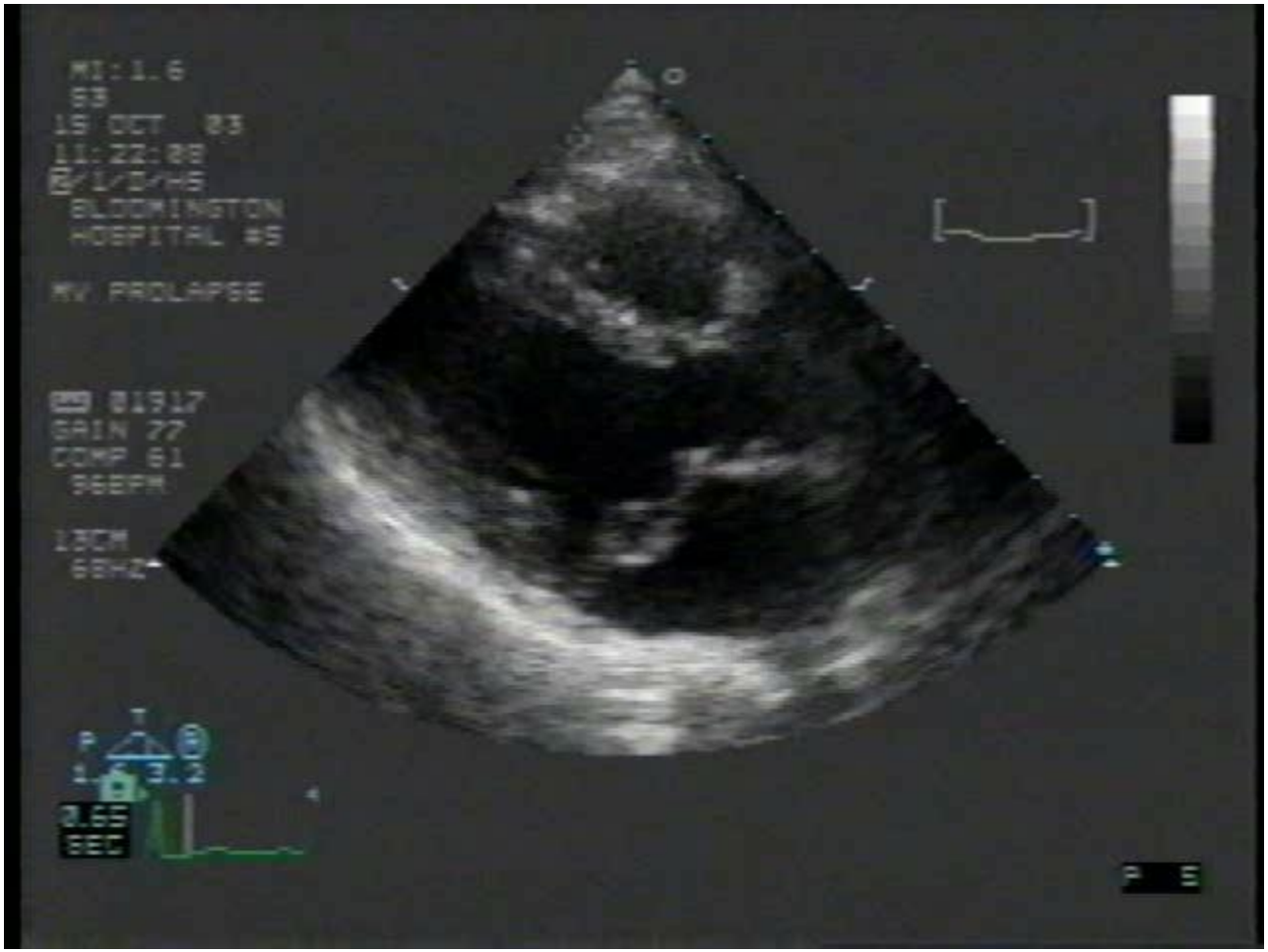
1992 Olympic Track Team, Barcelona, Spain

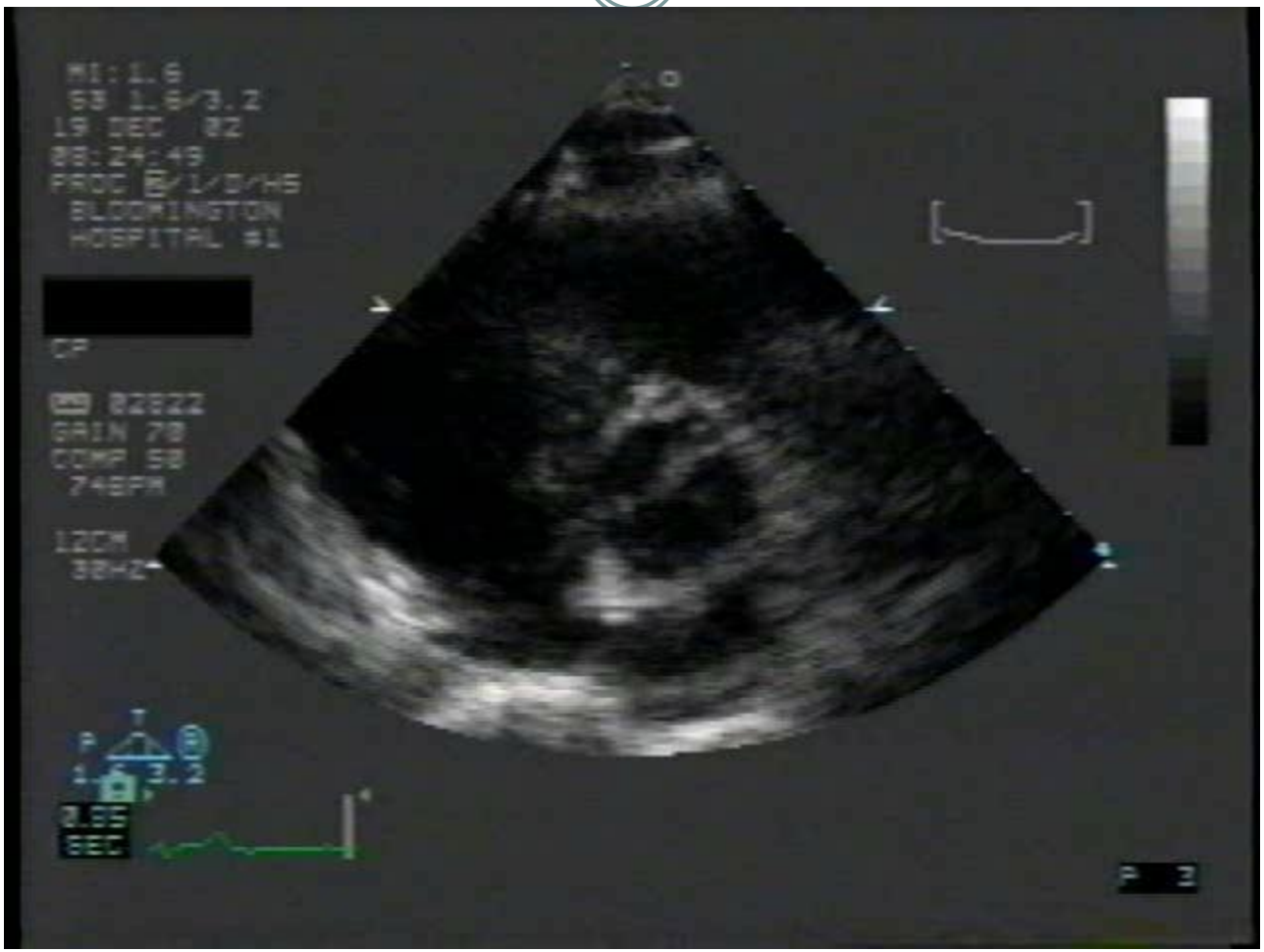
Congestive Cardiomyopathy



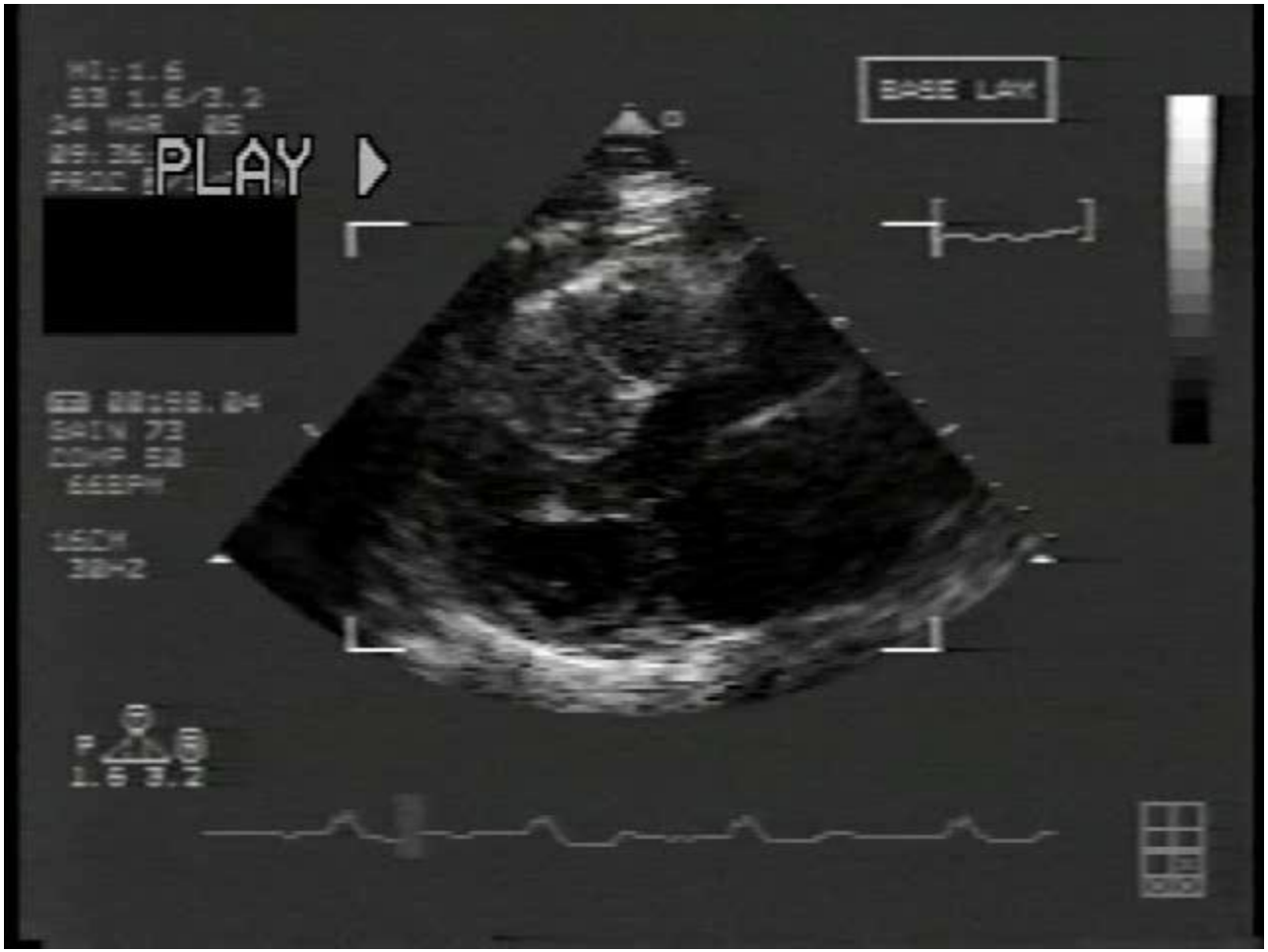
Mitral Valve Prolapse



Congenital Aortic Stenosis



HCM



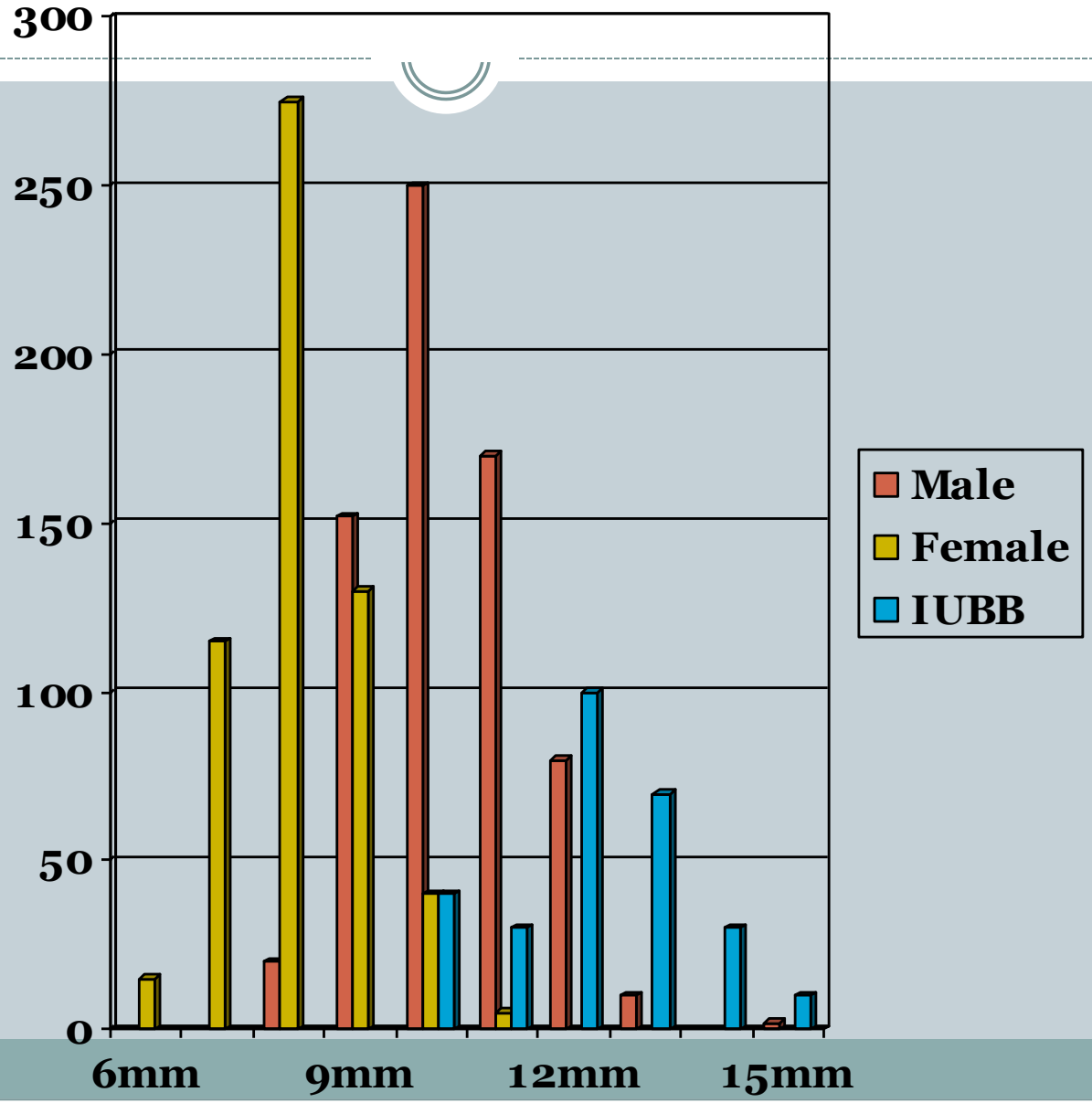
HCM

- Most common familial heart disease (1 in 500)
- Substantial heterogeneity
  - Presentation
  - Phenotypic expression negligible to extreme hypertrophy
  - Clinical course
  - Genetic substrate
- LVOT obstruction and arrythmias are cause for signs and symptoms
- Drug therapy does not reliably mitigate intraventricular pressure gradient at rest (Disopyramide)
- Beta Blockers do with exercise
- Most common cause of sudden death

Echo in HCM

- Unusual geometry and hypertrophy
- Exaggerated anterior displacement of the mitral valve apparatus and papillary muscles
- Primary enlargement and elongation of mitral valve leaflet
- Many doppler detected flow abnormalities
- Systolic anterior motion of the MV is the source of the obstruction 95% of the time
- PW/IVS >13mm
- +/- LVOT obstruction

LV Thickness in Young Athletes



Br J Sports Med
2009;43:649-656

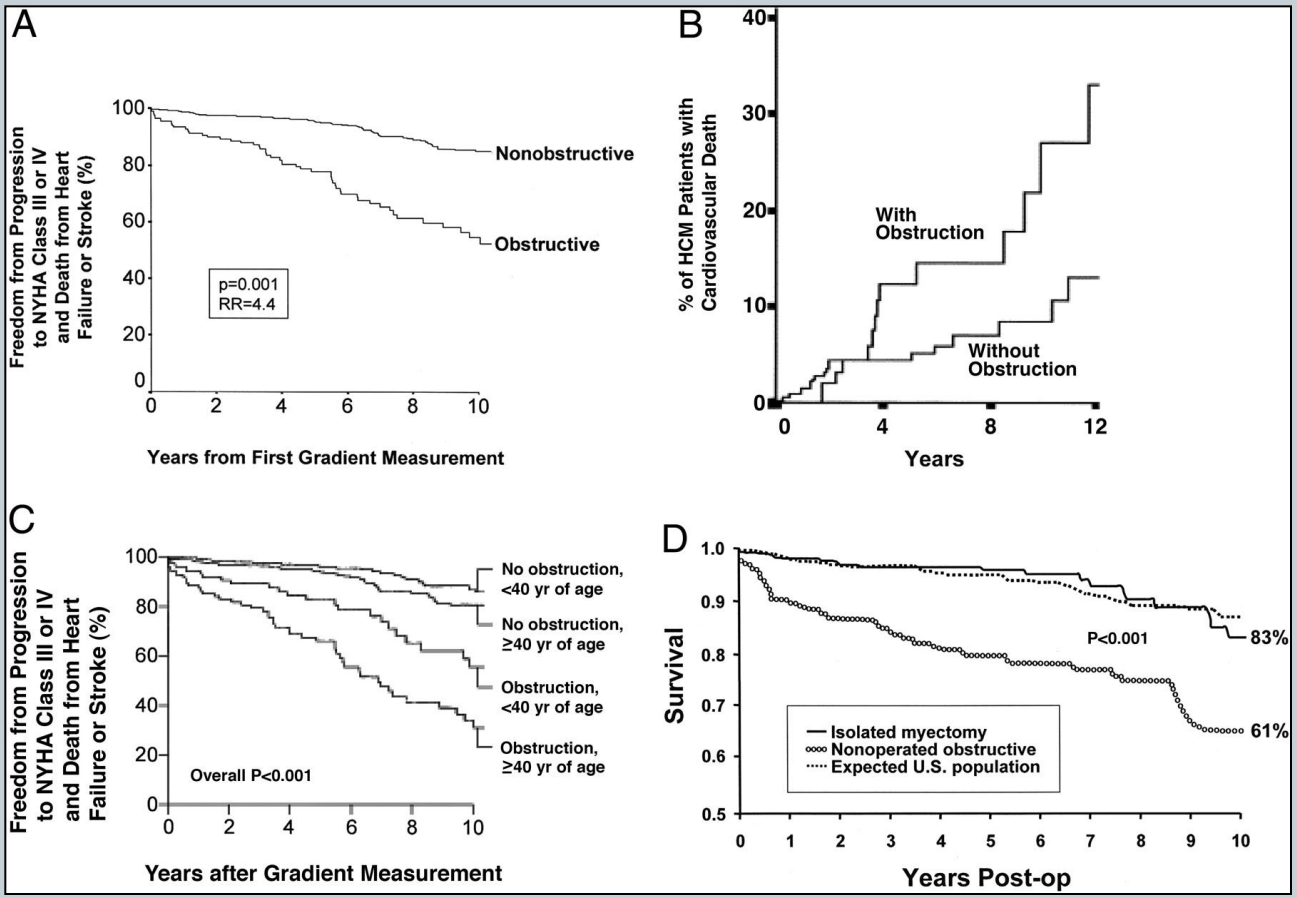
Rink 2009

HCM vs. Athlete's Heart Criteria

HCM		Athlete's Heart
+	asymmetric LVH	-
+	LVEDD<45mm	-
-	LVEDD>55mm	+
+	enlarged left atrium	-
+	diastolic dysfunction	-
+	Bizarre ECG changes	-
+	female patient	-
+	family history of HCM	-
+	regression of changes with deconditioning	-
-	peak O2 consumption	+
+	typical histology	-
+	positive genetic testing	-
-	electromechanical asynchronicity	+

Clin Res Card 2009, 98;2:80-88

Data From HCM Cohort Studies Supporting the Clinical Significance of LV Outflow Gradient

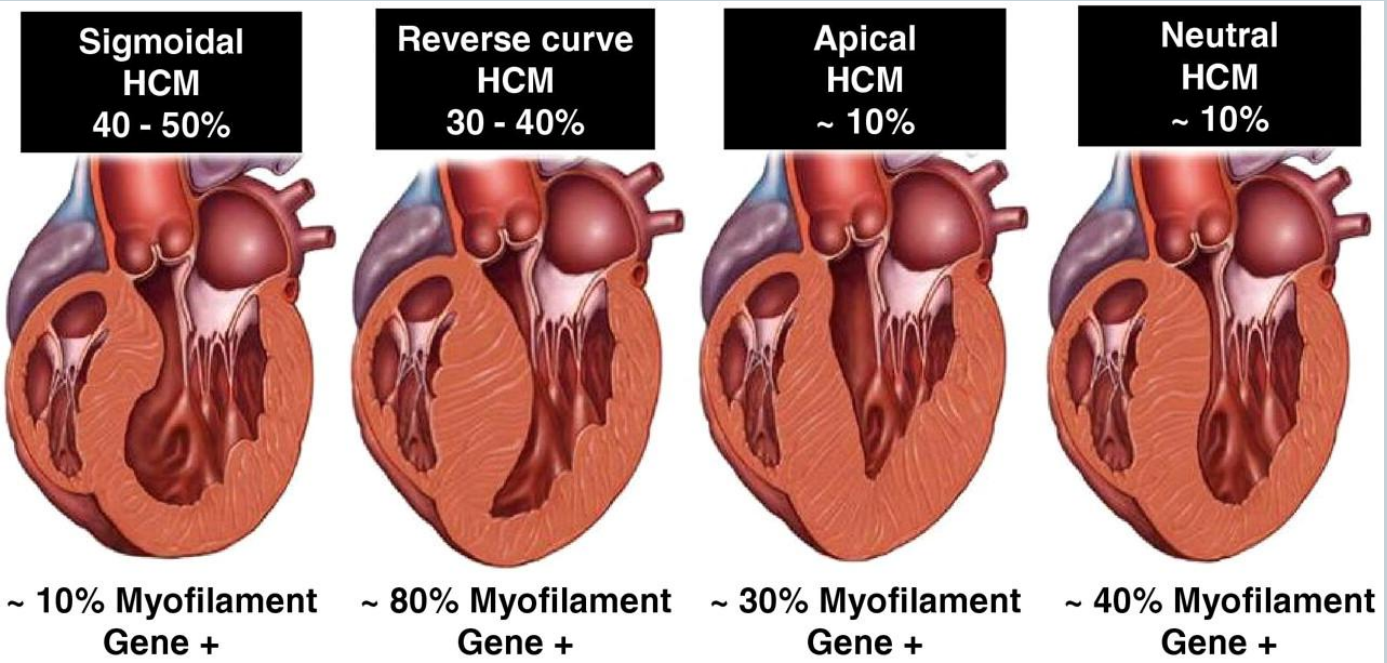


Maron, B. J. et al. J Am Coll Cardiol 2009;54:191-200

Genetic Testing of HCM

- Four companies offer testing for the eight most common myofilament associated genes (MYBPC3 and MYH7 most common)
- Myofilament encoding genes account for 35-65%
- Primary role is for diagnosis, not prognosis and not therapeutic
- Insurance may not pay

Septal Morphologies in HCM



Bos, J. M. et al. J Am Coll Cardiol 2009;54:201-211

Office Evaluation to Differentiate Athlete's Heart from HCM

- Initial Evaluation
  - Complete history
  - Physical examination
  - Echo evaluation in detail
  - EKG
- If primary or secondary relative has HCM and all above WNL recommend:
  - Genetic testing with index case and then the patient.
  - Follow above annually until age 25

What to do to differentiate Athlete's Heart from HCM

- Follow-up Evaluation
  - Stress echocardiogram (LVOT flow before and after exercise)
  - Cardiac MRI with late gadolinium enhancement (ischemic scar and non-ischemic fibrosis)
  - Stop all competitive sports and weight lifting
  - Echoes on primary relatives
  - If diagnosis of HCM in primary or secondary relatives do genetic testing on relative then on athlete

What to do if diagnosis of HCM

- If syncope, VT or SCA – AICD and register with ICD Sports Registry
- Consider medications if LOT obstruction or symptoms
- Psycho-social evaluation
- Education
- Arrange appropriate follow-up

Risk Stratification of Sudden Cardiac in Athletes with HCM

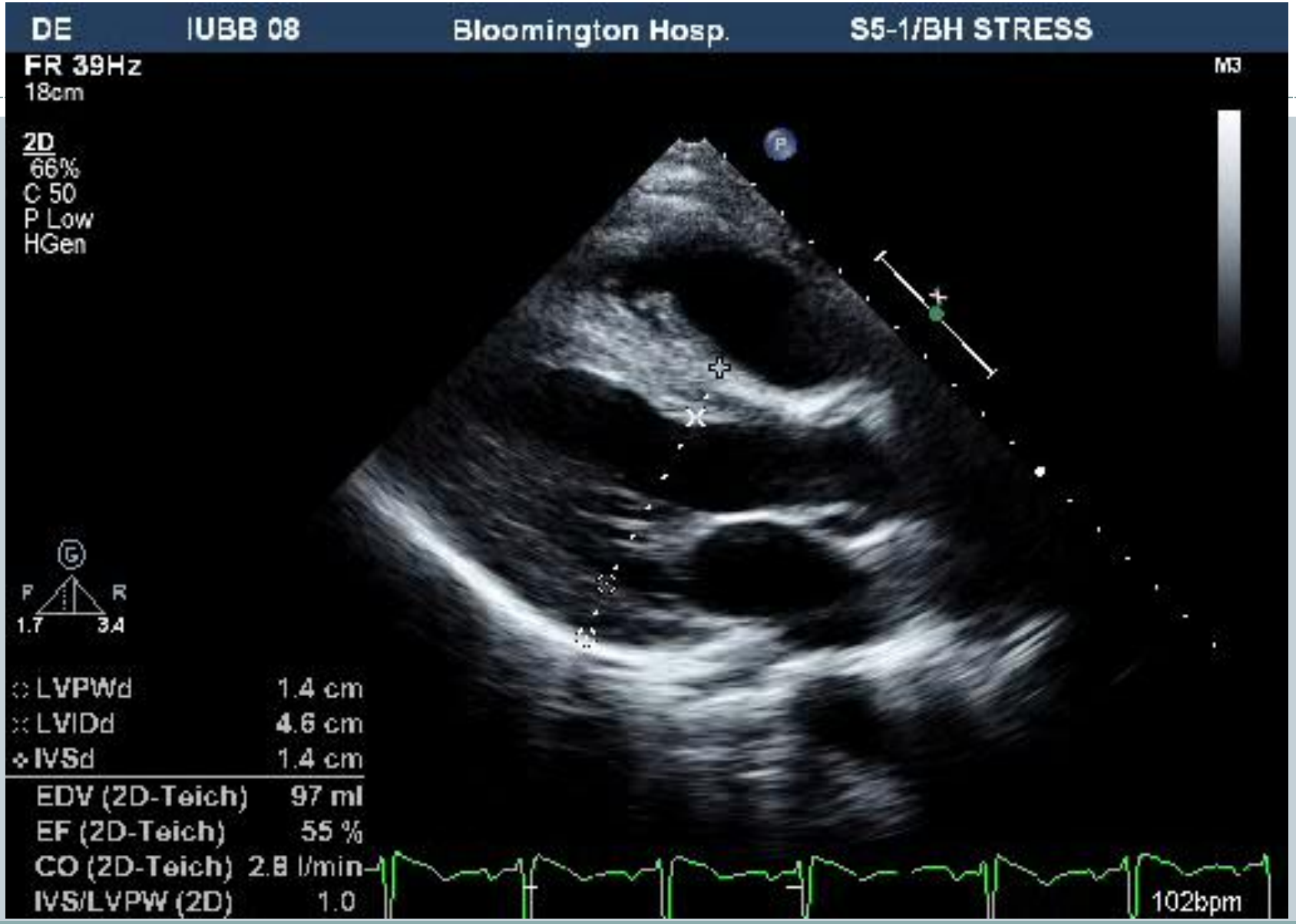
- Death associated with HCM 1-6% per person. Only 10-20% of those with HCM are at risk for SCA.
- Higher risk if:
  - Age 12-35
  - Previous aborted SCA
  - Unexplained syncope
  - Extreme LV hypertrophy >30mm
  - Blood pressure drop with exercise
  - Family history of SCA
  - Non-sustained ventricular tachycardia documented
  - Competitive higher intensity, higher static sports

Arrhythmogenic Right Ventricular Cardiomyopathy

- Structural and functional abnormalities of RV. Replacement of myocardium by fatty and fibrous tissue. May involve LV.
- Ventricular tachyarrhythmia occurring mostly with exercise
- Echo RV dilatation (also in athletes) and decreased RV function
- MRI – use late gadolinium enhancement (fibrosis) fatty changes (T1 weighted block blood series)

Controversies

- Incidence of Sudden Death in Athletes
  - US 1:160,000 to 1:300,000 per athlete per year
  - Italy 1:28,000 young athlete 12-25
  - Canada/US prospective 1:27,000
- What is diagnosis of HCM by echo
- What diseases require disqualifications?
- Is ECG required routinely in athletes?
- Can an athlete participate with an AICD in place?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IUBB 08
Bloomington Hosp.
S5-1/BH STRESS
FR 39Hz
18cm
M3
2D
66%
C 50
P Low
HGen
P
F
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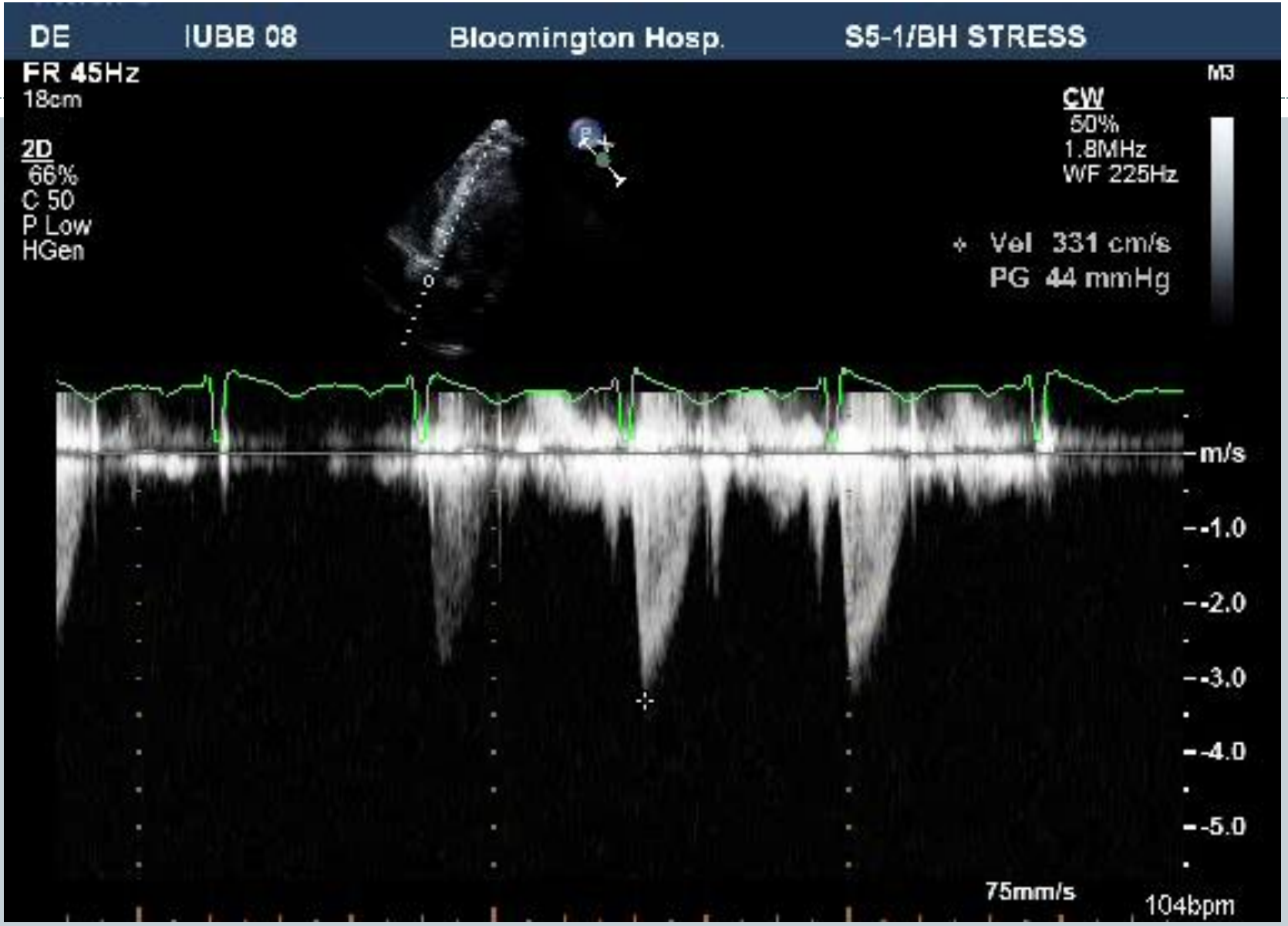
1.7
3.4
LVPWd 1.4 cm
LVIDd 4.6 cm
IVSd 1.4 cm
EDV (2D-Teich) 97 ml
EF (2D-Teich) 55 %
CO (2D-Teich) 2.8 l/min
IVS/LVPW (2D) 1.0
102bpm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S5-1/BH STRESS
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P Low
HGen
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50%
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WF 225Hz
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PG 44 mmHg
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-1.0
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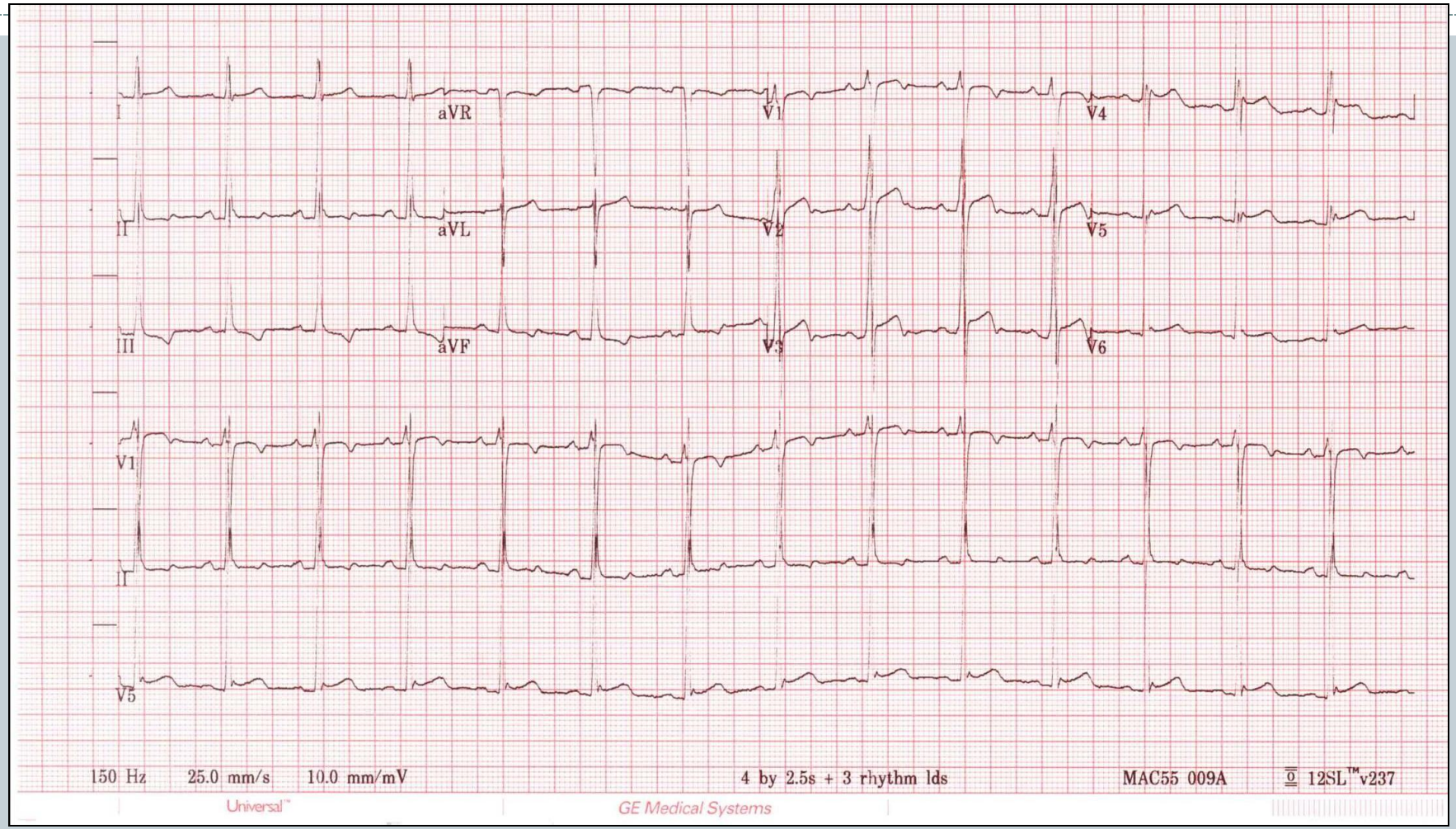
-3.0
-4.0
-5.0
75mm/s
104bpm


I
aVR
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V4
II
aVL
V2
V5
III
aVF
V3
V6
V1
II
V5
150 Hz
25.0 mm/s
10.0 mm/mV
4 by 2.5s + 3 rhythm lds
MAC55 009A
12SL™v237
Universal™
GE Medical Systems

Nick Knapp Case

- Cardiac arrest - 17 year old basketball player
- AICD placed
- Signs scholarship offer and enrolls in Northwestern University
- Declared ineligible by Northwestern
- Medical work up is negative
- Athlete sues Northwestern in effort to play
- After three court appearances, federal appellate court recognized the appropriateness of reliance on published guidelines of the 26th Bethesda Conference
- Transfers to an Ohio university. Plays basketball. Receives a shock for sinus tachycardia. Continues to play after one month.
- Has defibrillator removed. Continues to play basketball 15 years later.

BJSM, Sep 2009; 649-655

Brandon

- Age 17 was diagnosed with hypertrophic cardiomyopathy during a routine sports physical on his 15th birthday just weeks before starting his freshman year of high school. Two months after being diagnosed, Brandon had an ICD implanted. He has always been a serious and competitive athlete. Brandon went on to play on the JV basketball team his freshman year and ran varsity track. His ICD saved his life when his heart went into a dangerous rhythm while jogging during PE class. Brandon, now a junior at Dallas High School, is the starting varsity point guard and continues with competitive level competition and conditioning.

Sarah

- Sarah was diagnosed with hypertrophic cardiomyopathy, who now runs triathlons, completing two in the past year. She writes, “Having an ICD for the past fourteen years has been more than life-saving. It has given me the freedom to do all the activities I love. Despite a heart condition, I am healthy and strong. I have the confidence to swim, bike, run and play tennis thanks to the safety-net an ICD provides.”

Can We Prevent Sudden Death in Athletes?

No !

Yes !

Sometimes

What Should be Included in the Preparticipation Examination to Prevent Sudden Death?

- History questionnaire with follow up for positives
- Physical examination
- EKG
- Stress Echocardiogram
- Laboratory tests
  - Sickle cell trait, CBC, CMP, UA
- PFT before and after exercise
- Field test / Stress test
- Education
- React to warning signs (1/2 have previous symptoms)

So what can we as cardiologists do?

- Educate
- Screening echocardiograms
- Supply defibrillators
- Educate in use of above
- Support AHA and ACC
- Support ICD Sports Registry

  http://icdsports.org/

Characteristics of 1101 Patients with Hypertrophic Cardiomyopathy According to the Presence or Absence of Left Ventricular Outflow Tract Obstruction at Base Line

Table 1. Characteristics of 1101 Patients with Hypertrophic Cardiomyopathy According to the Presence or Absence of Left Ventricular Outflow Tract Obstruction at Base Line.*

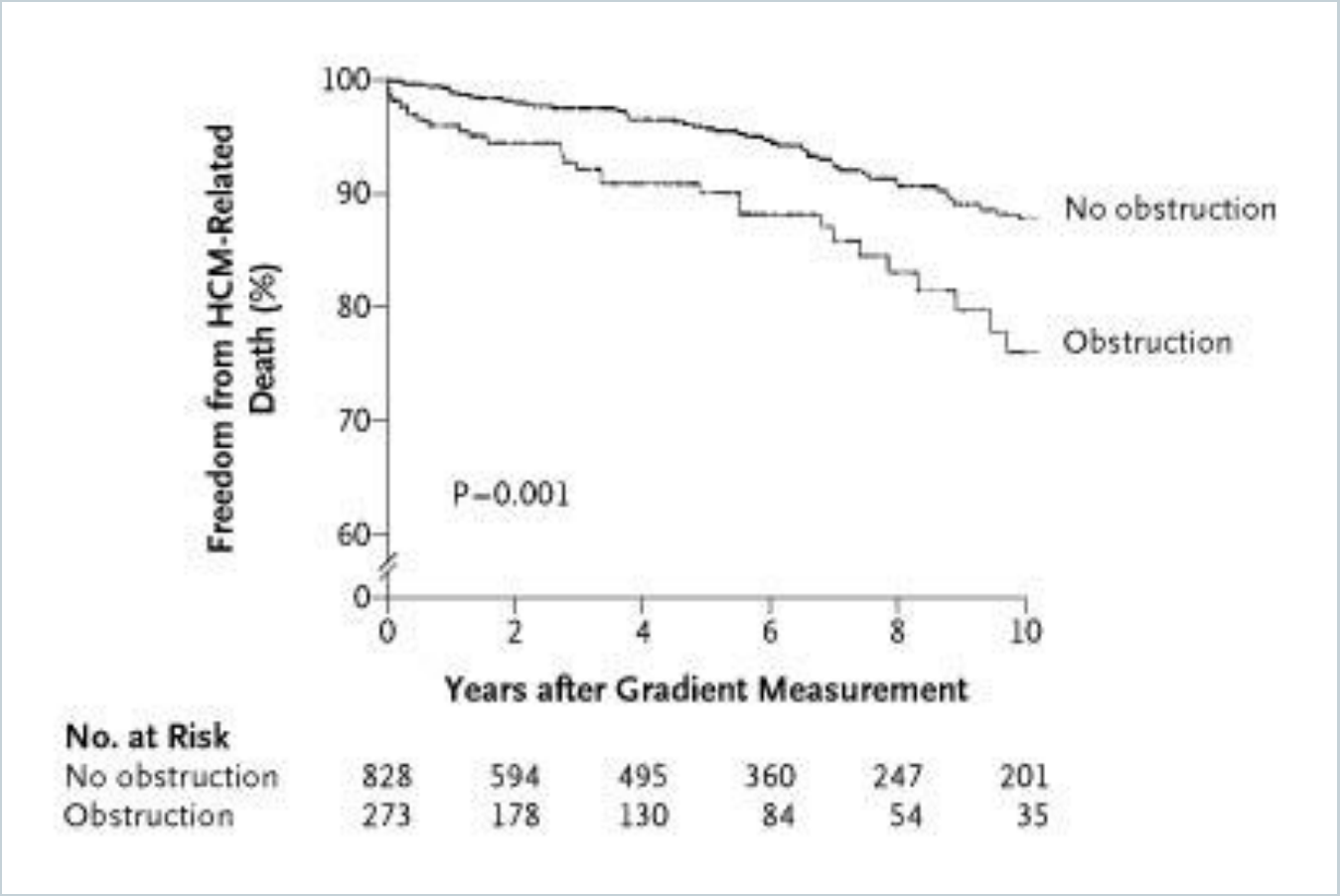
Characteristic	All Patients (N=1101)	Patients without Obstruction (N=828)	Patients with Obstruction (N=273)	P Value
Base-line measurements				
Male sex — no. (%)	655 (59)	529 (64)	126 (46)	<0.001
Age — yr	45±20	44±19	50±21	<0.001
NYHA class				
Mean	1.6±0.7	1.5±0.6	1.8±0.8	<0.001
I — no. (%)	596 (54)	490 (59)	106 (39)	NA
II — no. (%)	398 (36)	280 (34)	118 (44)	NA
III or IV — no. (%)	107 (10)	58 (7)	49 (18)	NA
Atrial fibrillation — no. (%)	220 (20)	147 (18)	73 (27)	<0.01
Syncope — no. (%)	152 (14)	96 (12)	56 (21)	<0.001
Drugs — no. (%)				
Beta-blockers	429 (39)	278 (34)	151 (55)	0.20
Verapamil	381 (35)	260 (31)	121 (44)	0.30
Disopyramide	35 (3)	11 (1)	24 (9)	0.50
Amiodarone	153 (14)	113 (14)	40 (15)	0.70
Echocardiographic measurements				
Left ventricular outflow tract gradient — mm Hg	21±33	5±7	70±32	NA
Maximal left ventricular thickness — mm	22±33	22±6	24±6	<0.001
Left atrial dimension — mm	42±8	41±8	45±8	<0.001
Left ventricular end-diastolic dimension — mm	43±7	44±7	41±7	<0.001
Follow-up measurements				
Duration of follow-up — yr	6.3±6.2	6.7±6.4	5.1±5.3	<0.001
NYHA class†				
Mean	1.7±0.8	1.6±0.7	2.2±0.9	<0.001
I — no. (%)	506 (48)	435 (55)	71 (28)	NA
II — no. (%)	323 (31)	249 (31)	74 (29)	NA
III or IV — no. (%)	216 (21)	107 (14)	109 (43)	NA
Major interventions for obstruction or symptoms — no. (%)				
Septal myectomy	40 (4)	—	40 (15)	NA
Mitral-valve replacement	7 (1)	2 (0.2)	5 (2)	NA
Alcohol septal ablation	19 (2)	—	19 (7)	NA
Heart transplantation	8 (1)	8 (1)	—	NA
Dual-chamber pacemaker	8 (1)	—	8 (3)	NA
Age at death — yr	54±19	52±18	59±19	0.14
Death related to hypertrophic cardiomyopathy — no. (%)	127 (12)	88 (11)	39 (14)	NA
Sudden death	71 (6)	51 (6)	20 (7)	NA
Heart failure	41 (4)	29 (4)	12 (4)	NA
Stroke	15 (1)	8 (1)	7 (3)	NA

* Plus–minus values are means ±SD. NA denotes not applicable.
† The 56 patients who died of heart failure or stroke were excluded from the analysis.

Maron, M. et al. N Engl J Med 2003;348:295-303

The NEW ENGLAND JOURNAL of MEDICINE

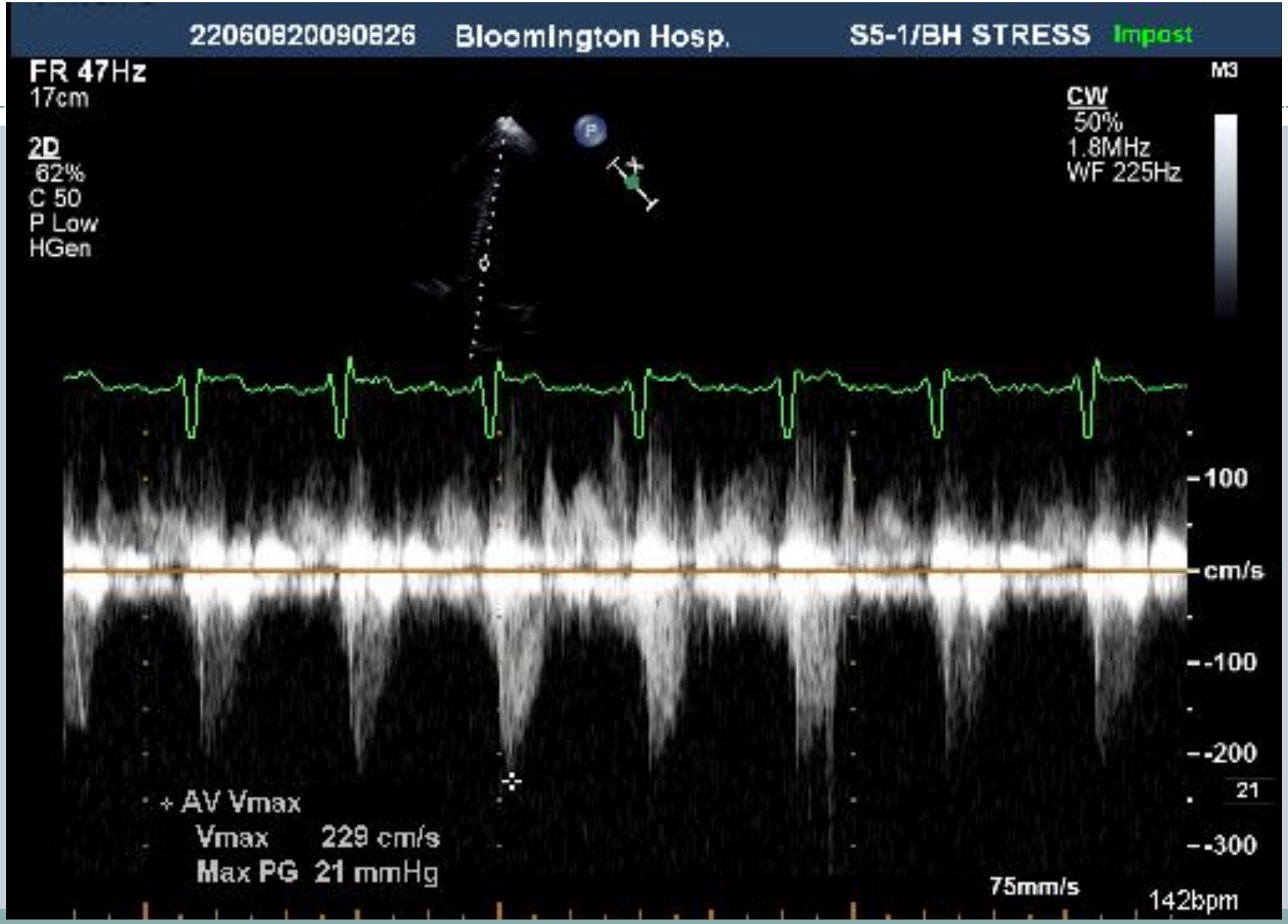
Probability of Hypertrophic Cardiomyopathy (HCM)-Related Death among 273 Patients with a Left Ventricular Outflow Gradient of at Least 30 mm Hg under Basal Conditions and 828 Patients without Obstruction at Entry



No. at Risk						
No obstruction	828	594	495	360	247	201
Obstruction	273	178	130	84	54	35

Maron, M. et al. N Engl J Med 2003;348:295-303

The NEW ENGLAND JOURNAL of MEDICINE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S5-1/BH STRESS Impost
FR 47Hz
17cm
2D
62%
C 50
P Low
HGen
CW
50%
1.8MHz
WF 225Hz
M3
100
cm/s
-100
-200
21
-300
+ AV Vmax
Vmax 229 cm/s
Max PG 21 mmHg
75mm/s
142bpm

Incidence of Sudden Death

- 1-3/100,000 person years
- Males 9x higher than females
- Most common cause is inherited/congenital cardiac disease

1,866 Deaths in the US 1980-2006

- Deaths 1994-2006 2x greater than in 1980-1993
- 90% were male
- 82% occurred during exercise
- 56% due to cardiovascular disease
- Majority of these HCM
- More Afro-Americans than anticipated

Circ 2009, 119:1085-1092

Catastrophic Events

	High School 1982-1992	High School 1992-2002	College 1982-1992	College 1992-2002
Direct catastrophes				
Deaths	67	91	7	9
Nonfatal	148	141	24	49
Serious	147	167	57	72
Total	**362**	**399**	**88**	**130**
Indirect Deaths				
Heart	114	201	25	47
Other	20	21	12	15
Total	**134**	**222**	**37**	**62**

Cardiac Problems and Sudden Death in Athletes, L.Rink 2005

Causes of 200 Cases of Sudden Death in Young Athletes

Diagnosis	Number	%
Hypertrophic cardiomyopathy	44	22.1
Coronary artery anomaly	30	15.1
Myocarditis	21	10.5
Arteriosclerotic coronary artery disease	22	11.1
Left ventricular hypertrophy	12	6.0
Right ventricular dysplasia	7	3.5
Conduction pathology	5	2.5
Mitral valve prolapse	9	4.5
Ruptured aorta	4	2.0
No cause determined	23	11.6
Other	22	11.1

Cardiac Problems and Sudden Death in Athletes, L.Rink 1995

Hypertrophic Cardiomyopathy

- Most common cause of sudden death in young athletes <30 yr old
- 0.2% of population 1:500
- 12 mutant genes and over 400 specific mutations
- Wall thickness of LV>15mm
- No single clinical morphologic or electrophysiologic factor has emerged as a reliable predictor of risk

LV Mass Dependant On

- Body size
- Sport
- Gender
- Genotype
- Any associated structural abnormalities such as AS

How to Differentiate Athletes Heart from Hypertrophic Cardiomyopathy

- History of family
- Echo
  - Walls >1.5mm
  - SAM MV
  - L atrial dilatation
  - LVOT pressure gradient
  - Reduction in LV mass with rest – 6 months
- MRI

FIMS Position Statement on Evaluation of Athlete to Prevent Sudden Death

- Rare 1:100,000
- Underlying cardiac disease is responsible for most events
- Most effective strategy not known
- Procedures unlikely to be cost effective
- If disease found in athlete or family, see 36th Bethesda Conference Recommendations

FIMS: International Federation of Sports Medicine

Genetic Testing

- Index case results are the gold standard
- Screen all primary relatives by ECG and echo
- Screen all secondary relatives if athletes
- Genetic test may not be covered by insurance

What are Appropriate Screenings?

- Yearly echo on 12-25 year olds
- Echo every 3-5 years thereafter
- Genetic test may not be covered by insurance