

RISK STRATIFICATION FOR SUDDEN CARDIAC DEATH

Dr. Antonio Curnis
EP Lab – Department of Cardiology
Spedali Civili di Brescia - Italy

PREVALENCE OF HCM AS CAUSE OF SCD IN YOUNG PATIENTS

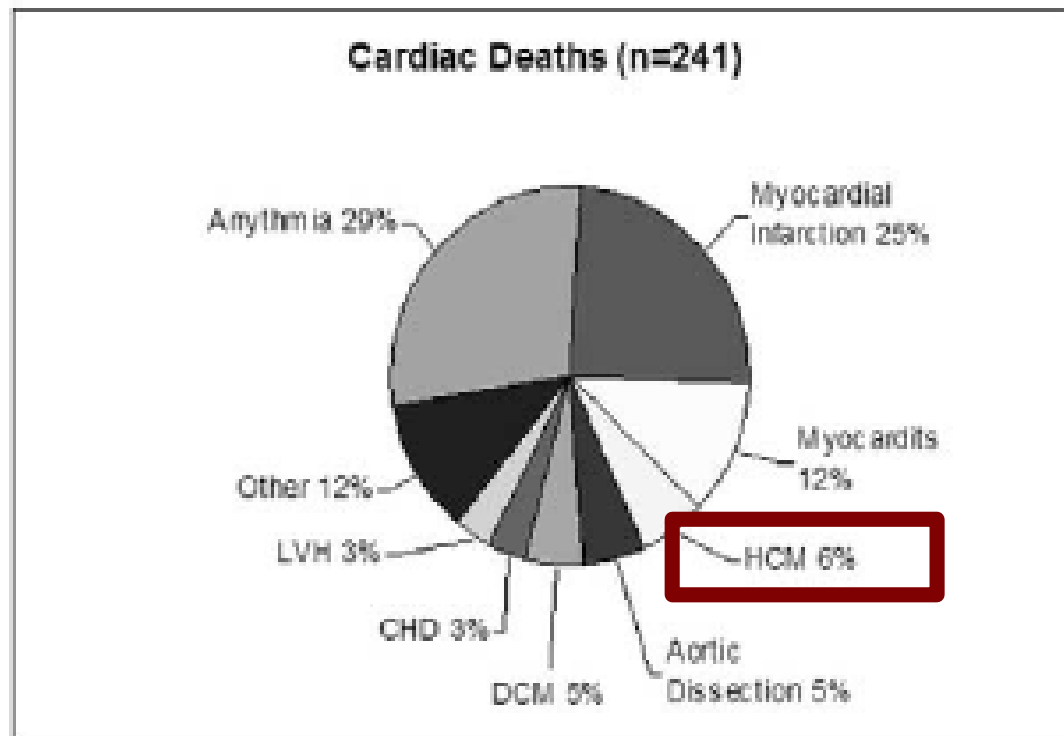
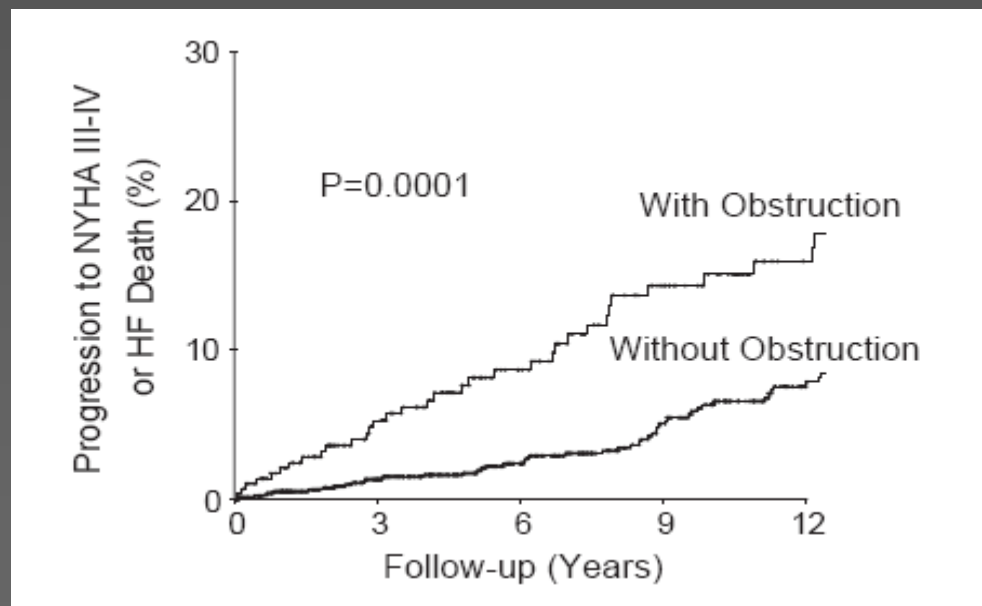
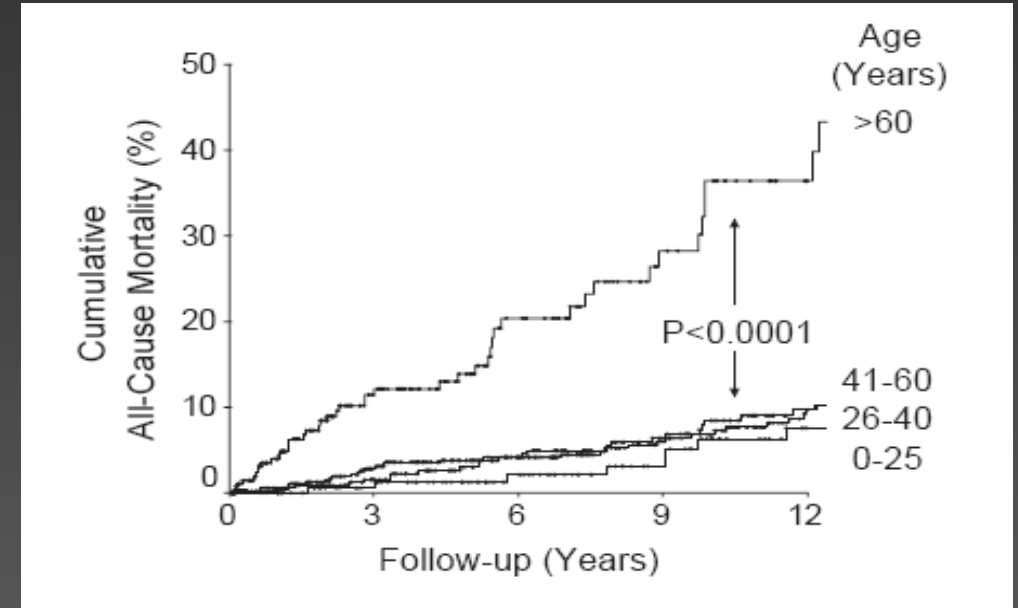
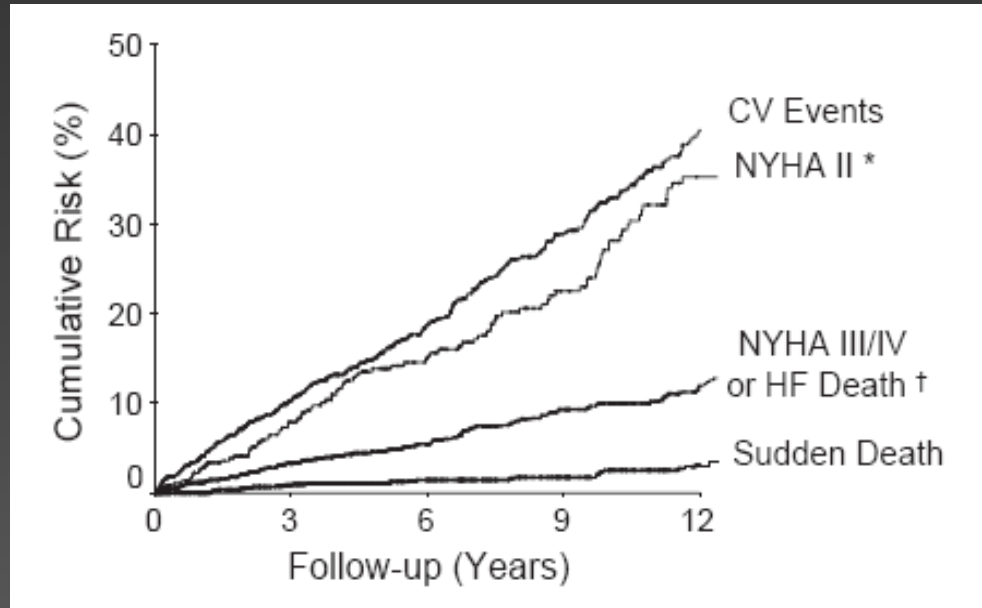


Figure 1 Total number of deaths attributed to cardiac causes. CHD = congenital heart disease; DCM = dilated cardiomyopathy; HCM = hypertrophic cardiomyopathy; LVH = left ventricular hypertrophy.

NATURAL HISTORY OF HCM



Data derived from the Italian Registry for Hypertrophic Cardiomyopathy

SUDDEN DEATH

- x Higher prevalence of malignant ventricular arrhythmia in two subsets of patients
 - x Aged 11 – 20 yrs
 - x Aged > 55 yrs
- x Lower incidence since 21 to 55 yrs

MECHANISMS OF SUDDEN CARDIAC DEATH - 1

- ✓ Paroxysmal AF
- ✓ Sinus tachycardia
 - ✓ Abnormal vascular responses
 - ✓ Myocardial ischemia
- ✓ Sustained monomorphic ventricular tachycardia
- ✓ Rapid AV conduction via an accessory pathway
- ✓ AV block

MECHANISMS OF SUDDEN CARDIAC DEATH - 2

- x Recent data reported that appropriated ICD discharges are related to:
 - x Monomorphic VT
 - x Fast VT degenerated in VF
 - x “Primary” VF

REMEMBER: UNDERSTANDING THE FINAL ARRHYTHMIA DOES NOT MEAN TO UNDERSTAND THE UNDERLYING MECHANISM THAT LED TO THE ARRHYTHMIA

ROLE OF MYOCARDIAL ISCHEMIA ?

POSSIBLE MECHANISMS OF MYOCARDIAL ISCHEMIA IN HCM

Increased myocardial
myocardial
oxygen demand

Reduced
perfusion

Myocardial hypertrophy
disease
Diastolic dysfunction
vascular response
Myocyte disarray
coronary vascular
Left ventricular outflow obstruction
Arrhythmia
bridges

Small vessels
Abnormal
Increased
resistance
Myocardial

ARE THERE RECOGNISED MARKER OF RISK OF SCD IN HCM ?

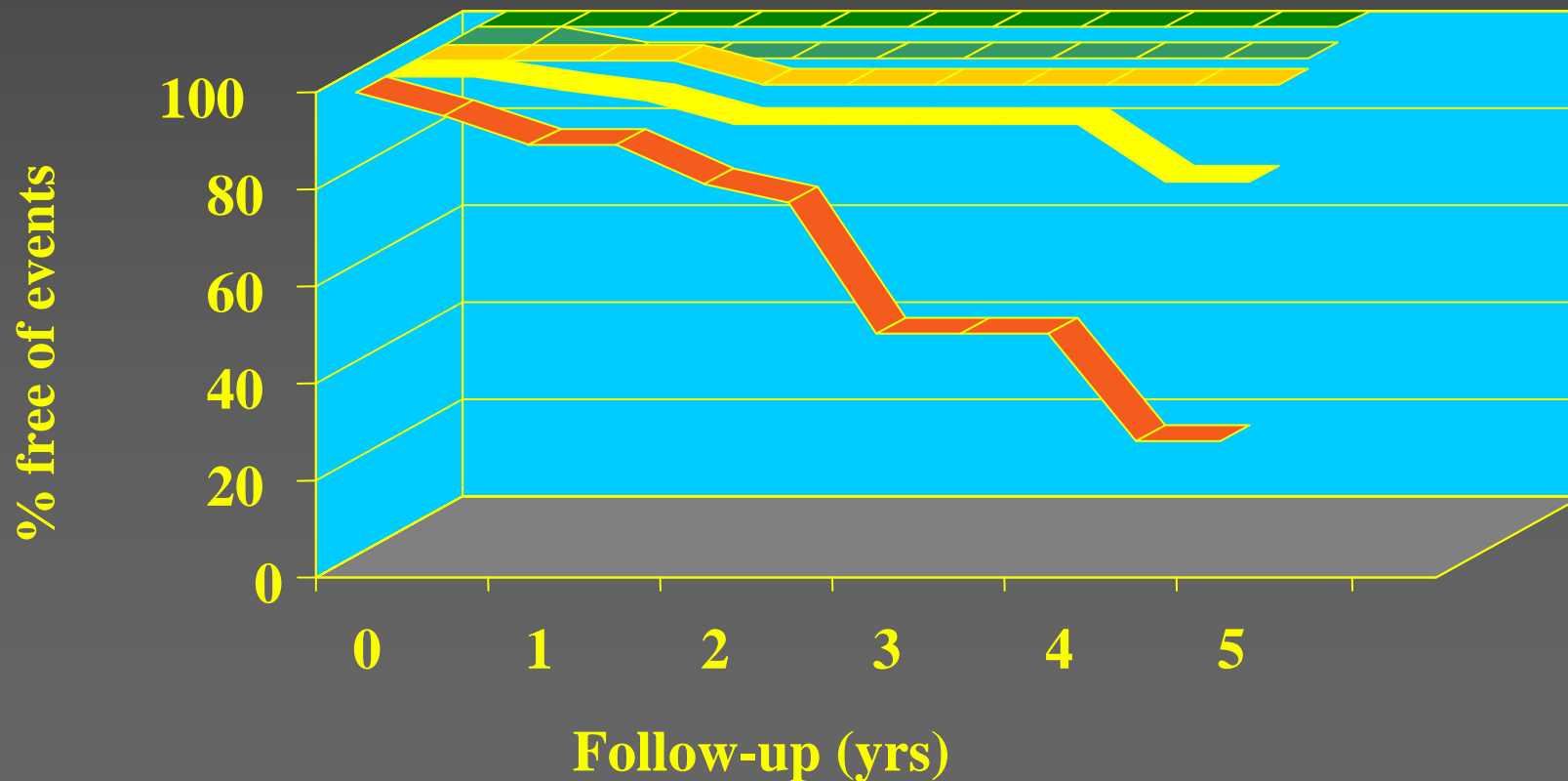
- Previous cardiac arrest
- NSVT at Holter or exercise
- Abnormal exertional blood pressure response
- Unexplained syncope
- Familial history of premature sudden death
- Severe left ventricular hypertrophy (≥ 3 cm)

RISK STRATIFICATION: WHICH POINTS MUST BE CONSIDERED ?

- x History
 - x Syncope and symptoms
 - x Prior cardiac arrest
 - x Family history
- x Echocardiogram
- x Holter monitoring
- x Blood pressure response to exercise testing

HISTORY AND CLINICAL PRESENTATION

- Cardiac arrest
- Pre-syncope
- Asymptomatic VT+
- Syncope
- Asymptomatic VT-



CLINICAL HISTORY

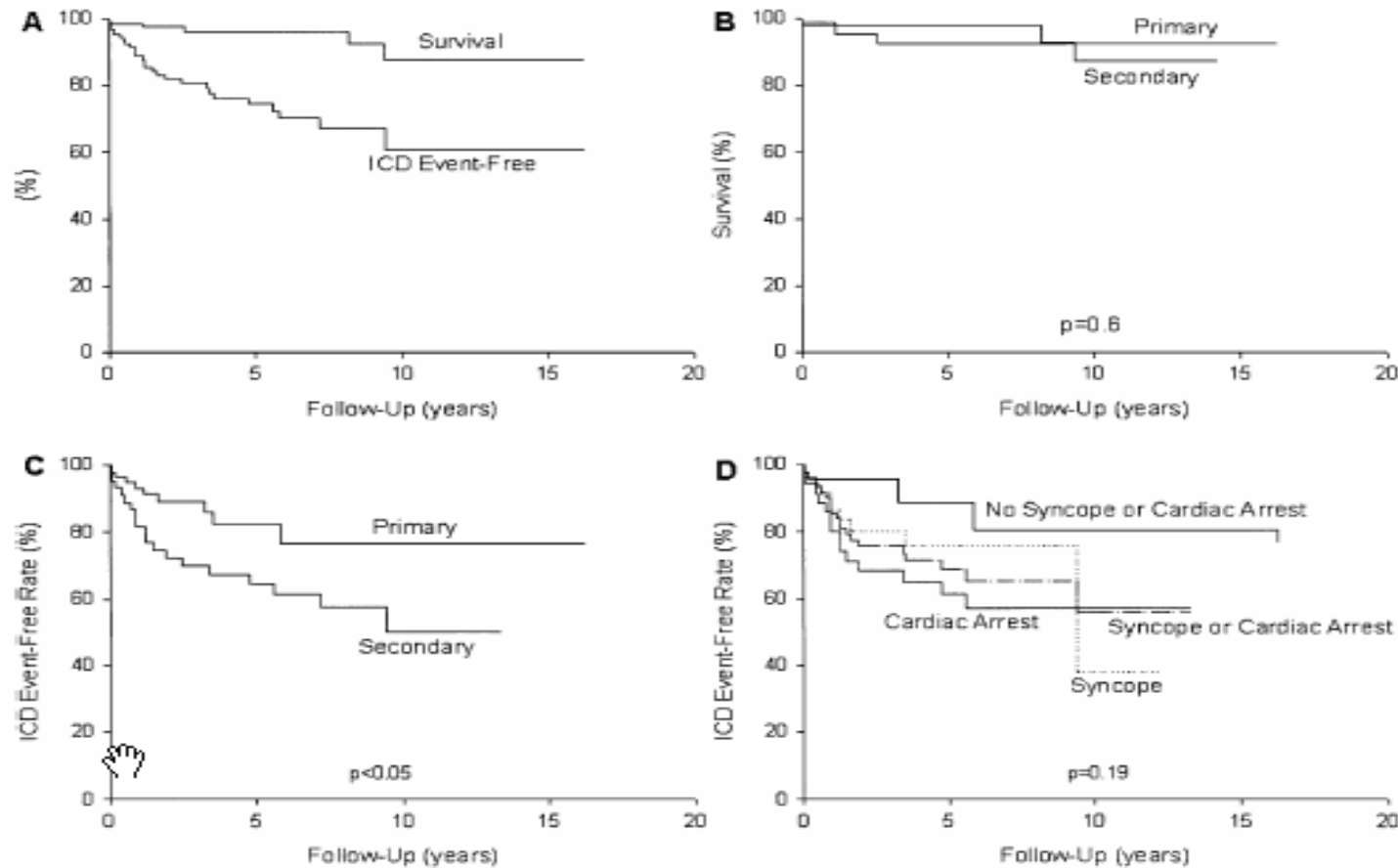


Figure 1. Panel A. Survival rates and therapeutic ICD intervention-free rates for all of the patients; Panel B. Survival rates in patients in whom ICDs were implanted for primary and secondary prevention of sudden death; Panel C. Comparison of appropriate ICD intervention-free rates in patients in whom ICDs were implanted for primary and secondary prevention of sudden death; and Panel D. Comparison of ICD intervention-free rates in patients who presented with cardiac arrest, syncope, cardiac arrest and syncope, and patients who had neither presentations.

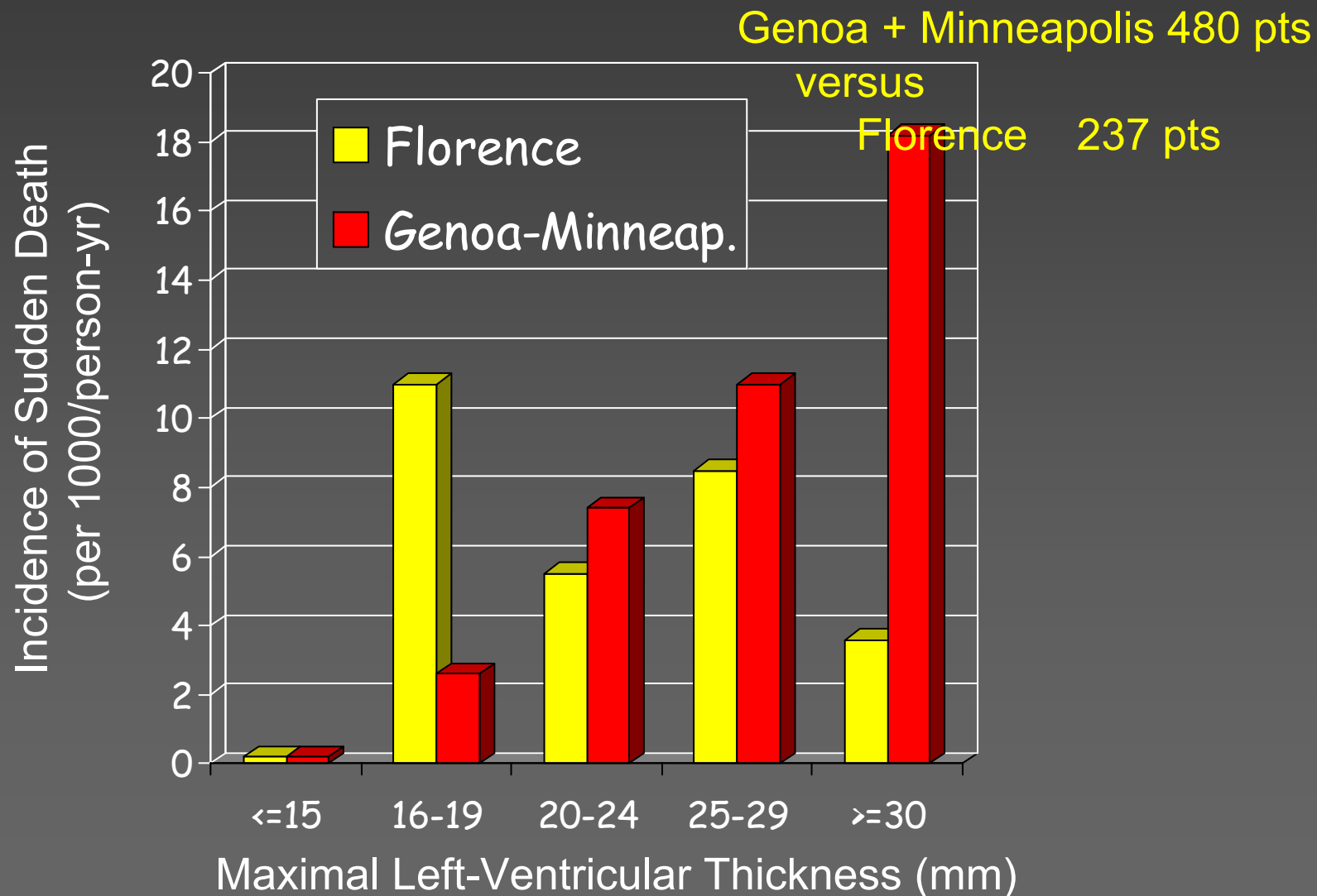
IMPORTANCE OF LV WALL THICKNESS

	n.subgr.	SUDDEN DEATH		CONGESTIVE HEART FAILURE	
		OR	p	OR	p
NYHA FC	2	-		9.48	0.001
LVOT OBSTRUCTION (basal gradient > 30 mmHg)	2	-		5.52	0.005
LV WALL THICKNESS 0.04	5	1.76	0.003	1.92	

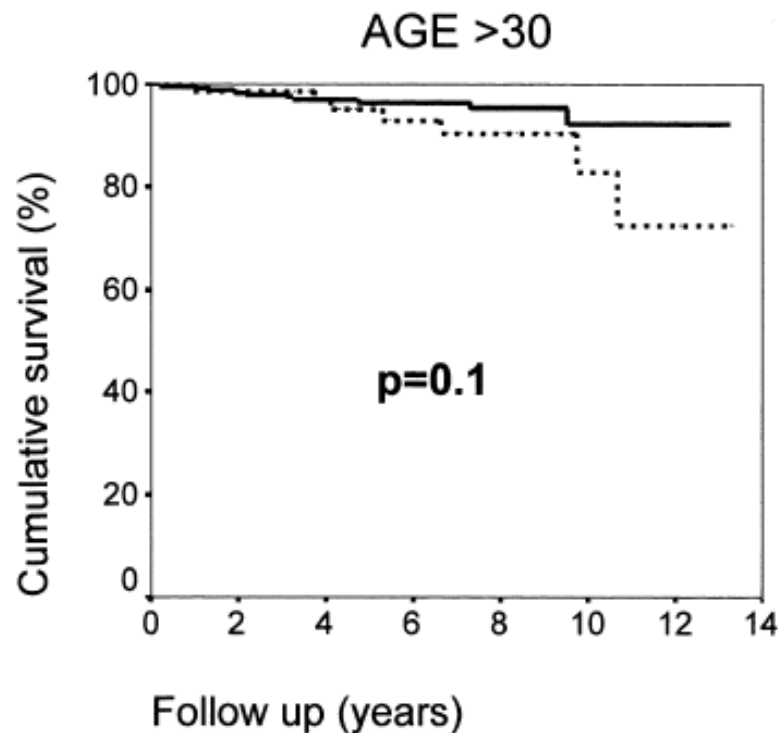
Spirito P et al. NEJ M 2000; 342: 1778
 Multivariate analysis, adjusted for age
 Genoa + Minneapolis 480 pts

Identification and treatment of pts at risk of SD

Maximal LV Wall Thickness and Risk of Sudden Death



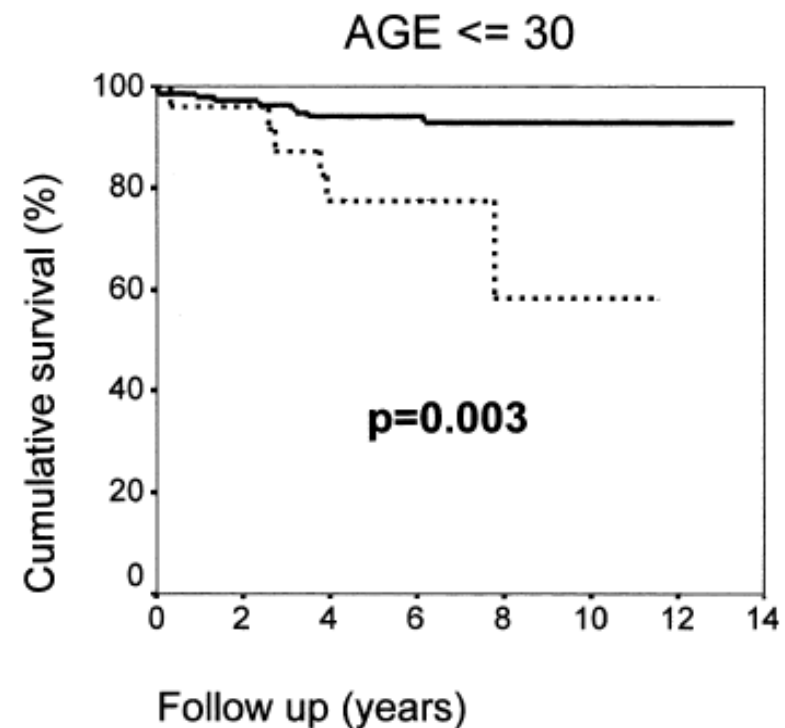
VALUE OF NSVT AT HOLTER: SCD RISK



Number of patients at risk

Without NSVT	279	235	184	119	66	23	7
With NSVT	78	68	57	38	28	9	6

A

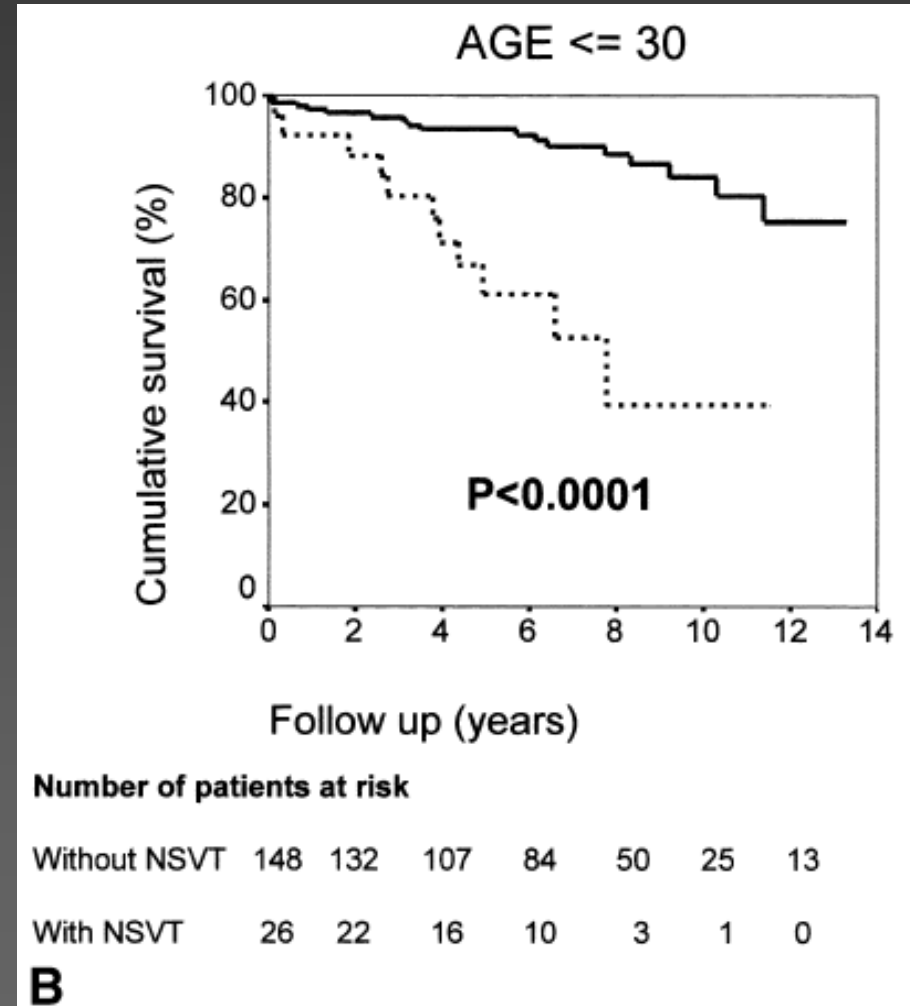
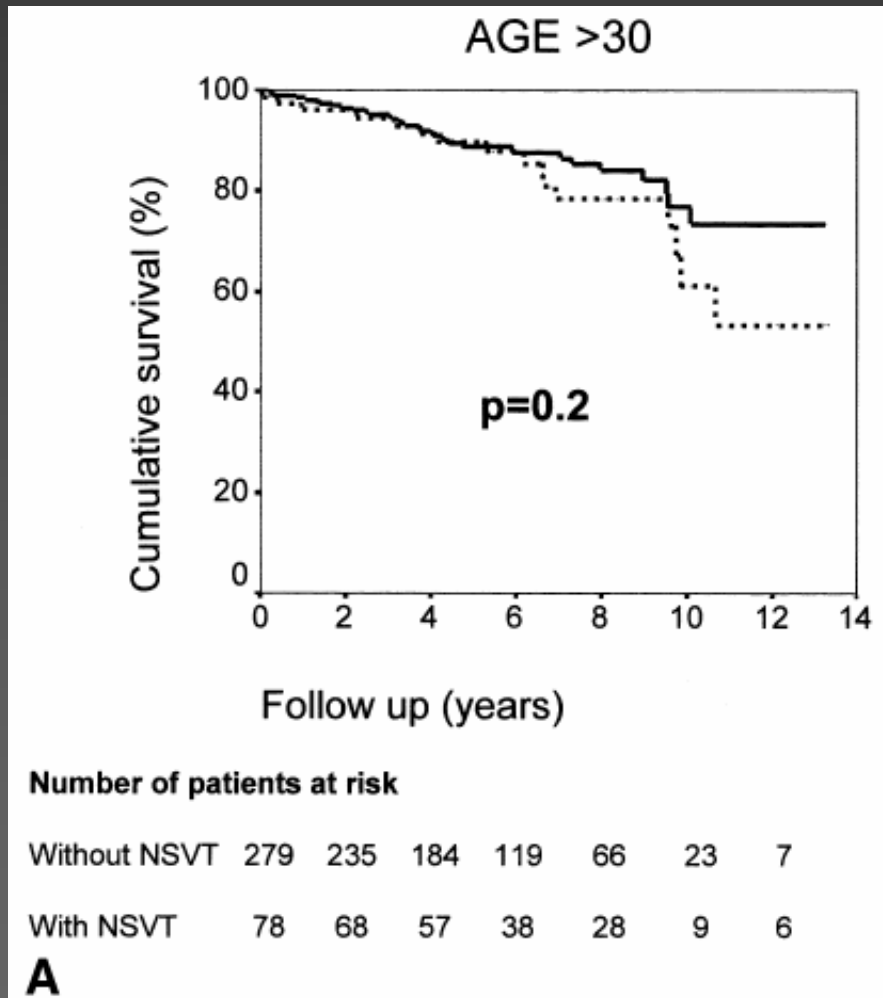


Number of patients at risk

Without NSVT	148	132	107	84	50	25	13
With NSVT	26	22	16	10	3	1	0

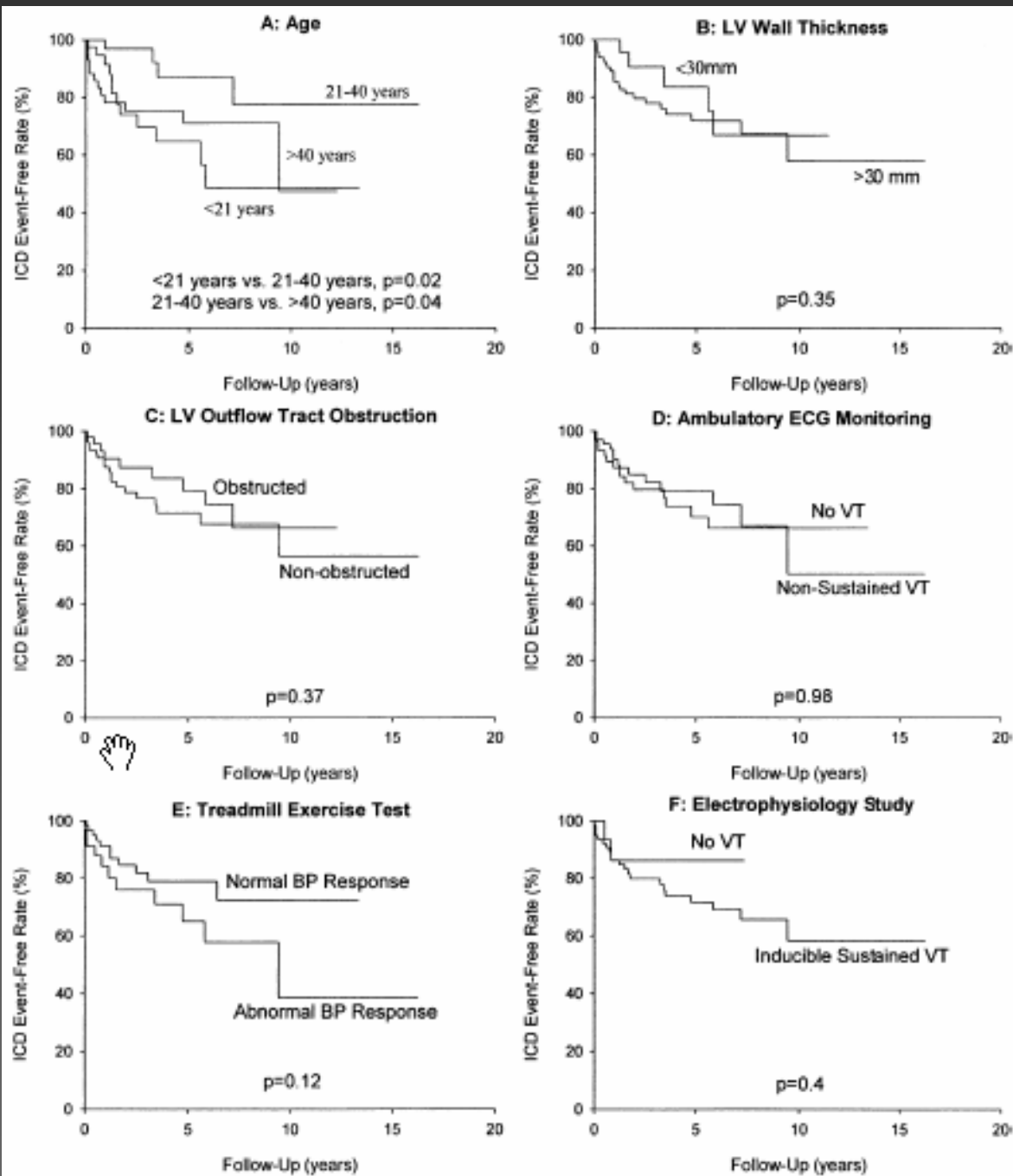
B

VALUE OF NSVT AT HOLTER: TOTAL MORTALITY RISK



BLOOD PRESSURE RESPONSE TO EXERCISE

- In healthy subjects, BP should increase during exercise
- In up to 25% of HCM, BP could remain “flat” during exercise or even decrease
- This fact is due to activation of baroreceptors of the LV - for an excess of wall stress during exercise – that lead to vagal discharge and vasodilation
- As other risk factors is more sensitive in young (< 40 yrs) patients



SUMMARY OF DIFFERENT RISK FACTORS

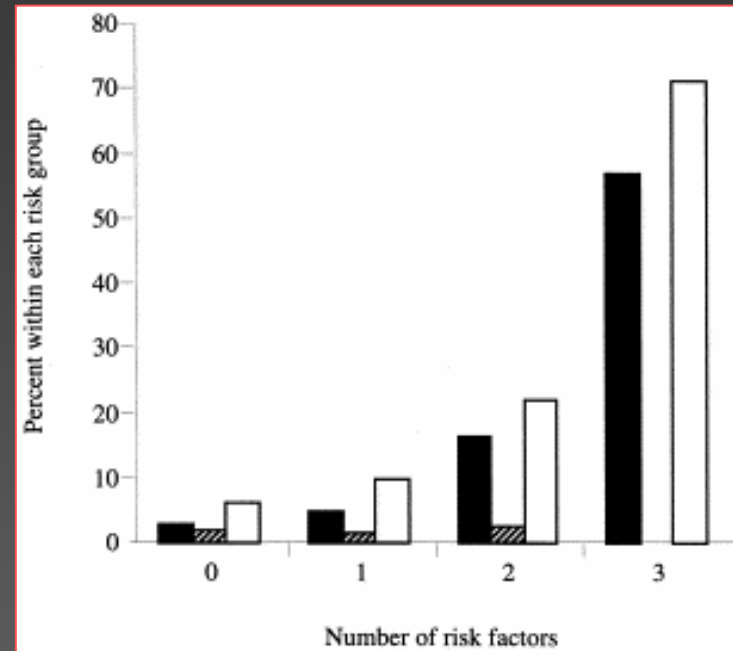
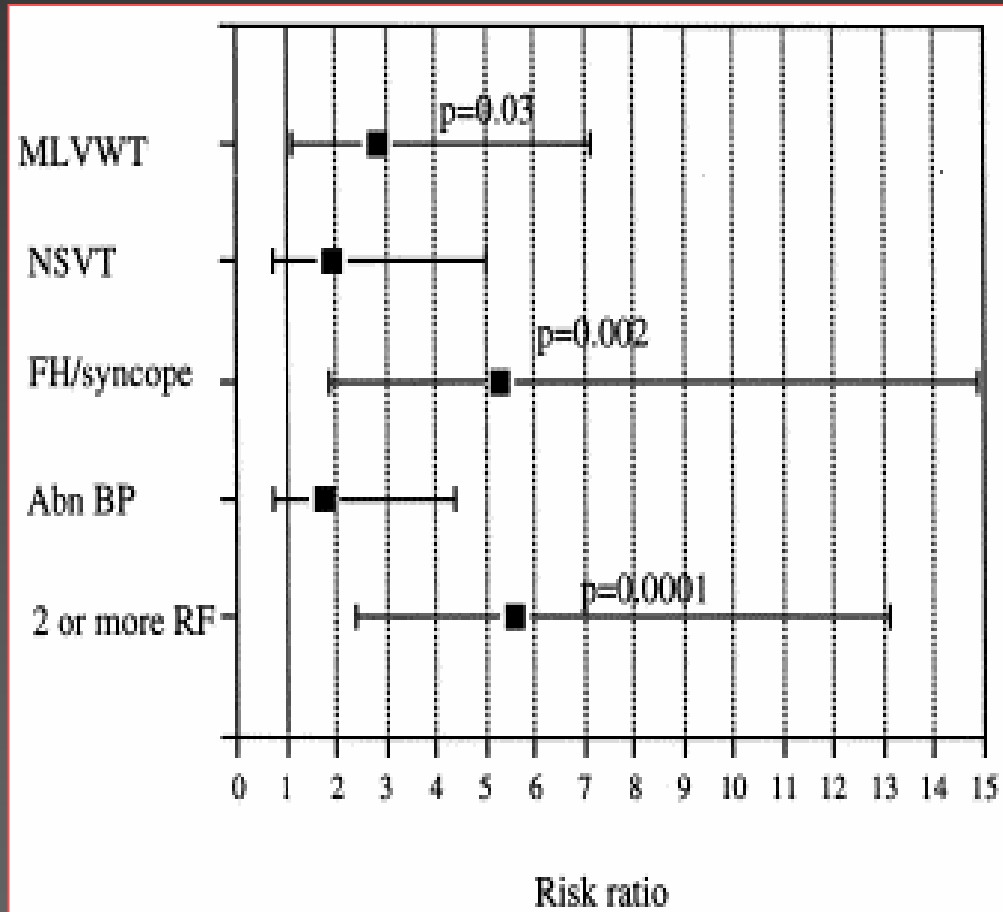
Begley, PACE 2001

Figure 2. The relation of therapeutic ICD intervention-free rates to 'risk factors' of sudden death: Panel A. Age; Panel B. Severity of LV wall thickness; Panel C. LV outflow obstruction; Panel D. Non-sustained VT during ambulatory electrocardiographic (ECG) monitoring; Panel E. Abnormal blood pressure (BP) response during treadmill exercise test (BP did not predict outcome also in the subset of subjects aged < 40 years); and Panel F. Induced sustained ventricular arrhythmia.

SENSITIVITY, SPECIFICITY, PPV, NPV OF DIFFERENT RISK FACTORS

Risk factor	Sensitivity (%)	Specificity (%)	PPA (%)	NPA (%)
Abnormal blood pressure response: <40 years old ¹⁹	75	66	15	97
NSVT: adult <45 years old ¹⁸	69	80	22	97
NSVT: ≤ 21 years old ²³	<10	89	<10	85
Inducible VT/VF: High risk population ^{w30}	82	68	17	98
*Syncope: <45 years old ³	35	82	25	86
*Family history: at least one unexplained sudden death ± HCM ³	42	79	28	88
†LVH ≥ 3 cm ¹⁷	26	88	13	95
††Two or more risk factors ²	45	90	23	96

SCD RISK STRATIFICATION: USEFULNESS OF A MULTIFACTORIAL APPROACH



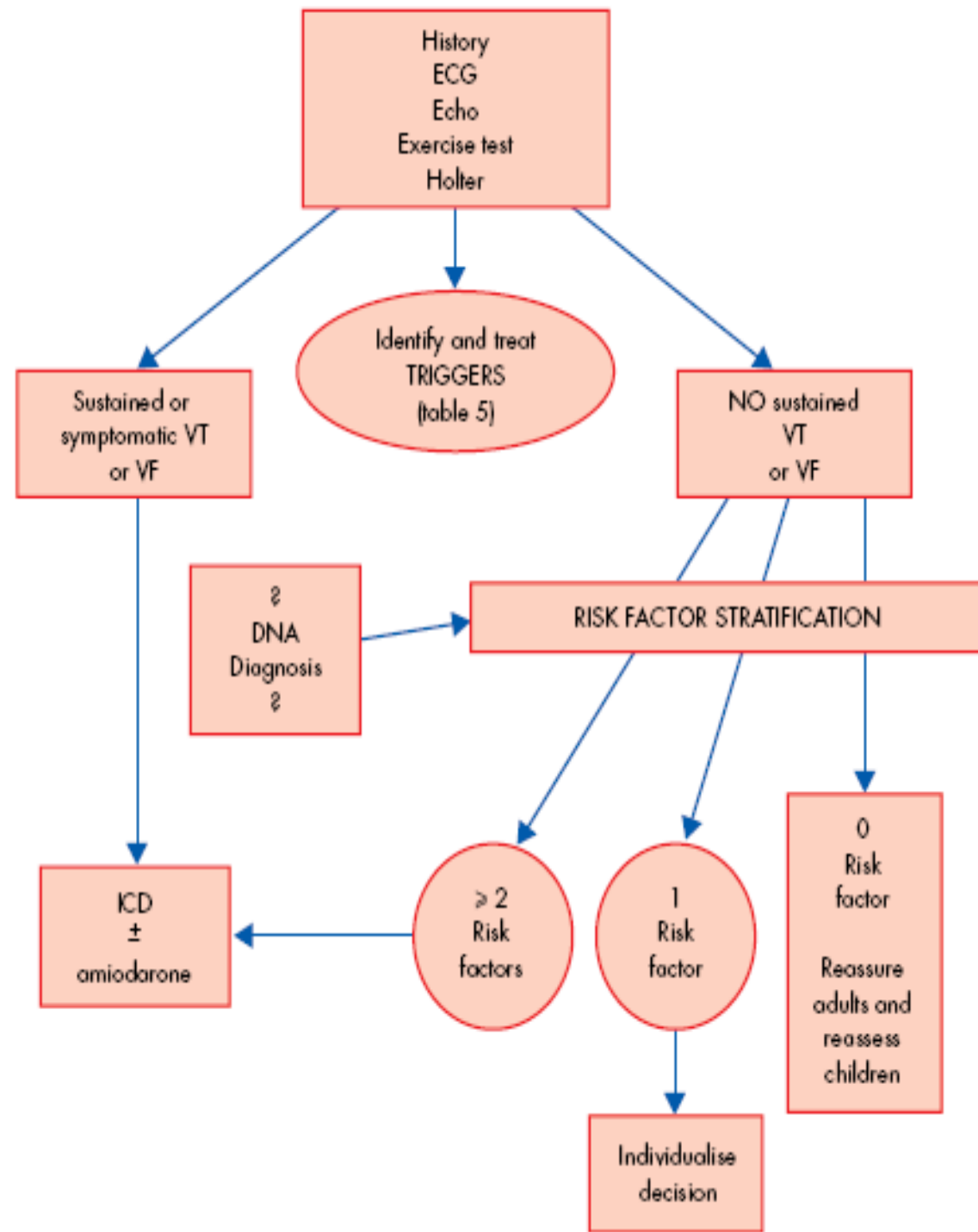
n=368 (14-65 years, 239 M)

No amiodarone

FU 3.6±2.5 years, (2 days-9.6 yrs)

22 Sudden deaths

Annual risk of SD 1,7%



CONCLUSIONS

- Even if there are emerging data derived from more sophisticated instrumental and genetical examinations, simple tests remain of high value in stratifying SCD risk in HCM patients
- Especially in young people (probably because they are at higher risk of SCD) this stratification seems to be extremely useful
- Data derived from large population show that ICD implantation could be extremely useful in reducing total mortality
- This means that SCD remains one of the greater risk in HCM patients