

Case. 35 yrs Male. Recurrent Syncope
HCM With LVOT Obstruction
(PG 37mmHg) & 30mm-Septal
thickness
- Myectomy first

삼성서울병원 순환기내과
박 경 민

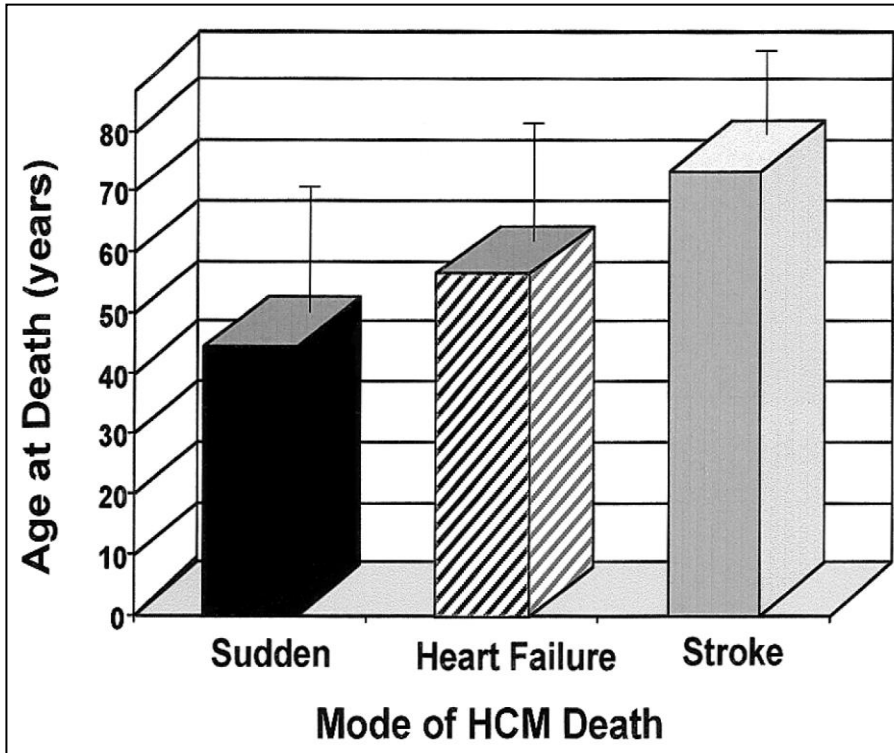


Natural History

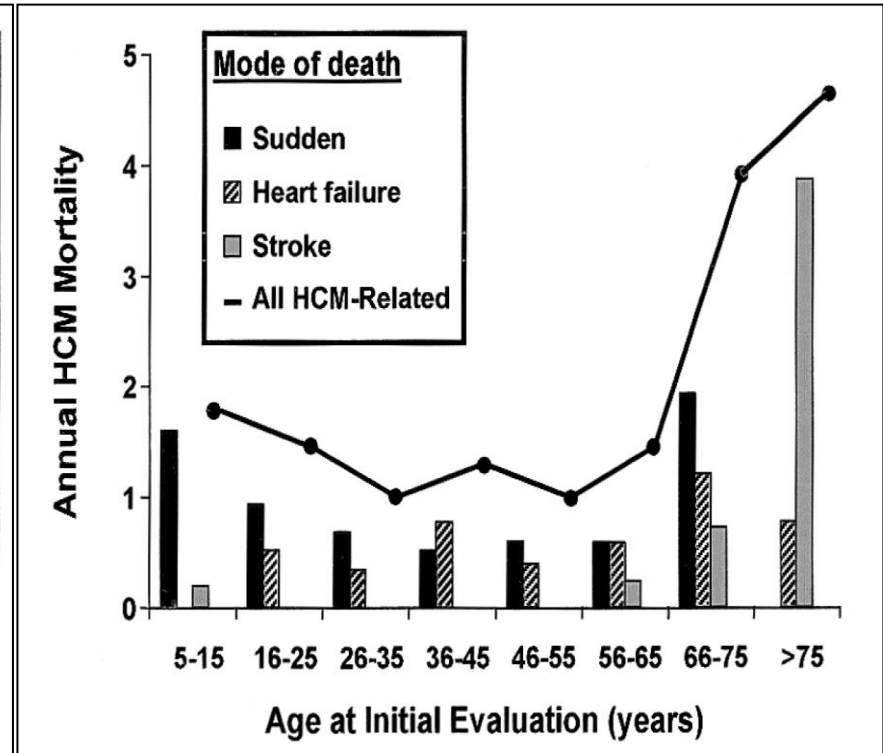
- ▶ **Incidence** 0.1% ~ 0.2%
- ▶ **Normal life expectancy**
- ▶ **Mortality**
 - ▶ Adults – 1% / yr
 - ▶ Children - 2% / yr
- ▶ **Most common cause of SCD in the young**
- ▶ **Subgroups at higher risk for important disease complications**
 - ▶ sudden and unexpected death
 - ▶ progressive heart failure
 - ▶ atrial fibrillation (AF) and consequent embolic phenomenon

Epidemiology of HCM-Related Death

: Revisited in a Large Non-Referral-Based Patient Population



Ages at which 3 modes of HCM-related death occurred in 86 patients.



Annual HCM-related mortality depicted prospectively from age at study entry

High Risk

Secondary prevention

1. Prior cardiac arrest
2. Sustained ventricular tachycardia

Primary prevention

one or more of the following

1. Family history of one or more premature HCM-related deaths, particularly if sudden and multiple
2. Unexplained syncope, especially if recent and in the young
3. Hypotensive or attenuated blood pressure response to exercise
4. Multiple, repetitive (or prolonged) NSVT on Holter in young
5. Massive LVH (wall thickness, ≥ 30 mm)

2011 ACCF/AHA guidelines

Selection of Patients for ICDs—Recommendations

Class I

1. The decision to place an ICD in patients with HCM should **include application of individual clinical judgment, as well as a thorough discussion of the strength of evidence, benefits, and risks** to allow the informed patient's active participation in decision making.
2. ICD placement is recommended for patients with HCM with **prior documented** cardiac arrest, v. fib, or hemodynamically significant VT.

Class IIa. Benefit >> Risk, Additional studies with focused objectives needed

1. It is reasonable to recommend an ICD for patients with HCM with:

- b. A maximum LV wall thickness greater than or equal to 30mm
- c. One or more , unexplained syncope episodes.

2. An ICD can be useful in select patients with NSVT (particularly those <30 years of age) in the presence of other SCD risk factors or modifiers.

3. An ICD can be useful in select patients with HCM with an abnormal blood pressure response with exercise in the presence of other SCD risk factors or modifiers.

4. It is reasonable to recommend an ICD for high-risk children with HCM, based on unexplained syncope, massive LV hypertrophy, or family history of SCD, after taking into account the relatively high complication rate of long-term ICD implantation.

ICD guidelines in Korea

- 비후성 심근병증 환자로서 아래의 ① ~ ⑤ 중 두 가지 이상에 해당되는 경우
 - (1) 실신의 증상
 - (2) 급사의 가족력
 - (3) 좌심실중격의 과도한 비후(>30mm)
 - (4) 24시간 활동 중 심전도에서 나타난 비지속성 심실빈맥
 - (5) 운동부하검사 상 이상 혈압증가 반응이 없는 경우
(충분한 운동부하에도 혈압상승이 < 20mmHg 인 경우)



Considerations

- Age
- Strength of the risk factors
- Risk-benefit

Consider Age 35

- Relatively young
- Long term complications of ICD
 - ☞ Inappropriate shock
 - ☞ Leads complication
 - ☞ Device infection

Device complications and inappropriate implantable cardioverter defibrillator shocks in patients with hypertrophic cardiomyopathy

G Lin,¹ R A Nishimura,¹ B J Gersh,³ D Phil,¹ S R Ommen,¹ M J Ackerman,² P A Brady¹

• ICD complications

Complication	No of patients (%)
Pneumothorax (at ICD implant)	1 (<1)
Pericardial effusion (at ICD implant)	3 (2)
Pocket haematoma	4 (2)
Early (\leq 1 month)	3
Late (with generator change)	1
Upper extremity deep venous thrombosis	1 (<1)
Lead revision	24 (13)
Acute (\leq 24 h)	6
Chronic ($>$ 24 h)	18
ICD infection	8 (5)
Early (\leq 1 month)	1
Late ($>$ 1 month)	7
ICD revision for high defibrillation threshold testing	6 (3)
Subcutaneous array	4
Lead revision	1
Generator change	1
Inappropriate shocks	42 (23)
Atrial fibrillation	20
Sinus tachycardia	16
Device malfunction	6

- 181 patients
(44 years; 62% male)
- mean follow-up: 59 months
- 65 patients (36%) had a total of 88 device complications, including 42 (23%) patients with inappropriate shocks

Device complications and inappropriate implantable cardioverter defibrillator shocks in patients with hypertrophic cardiomyopathy

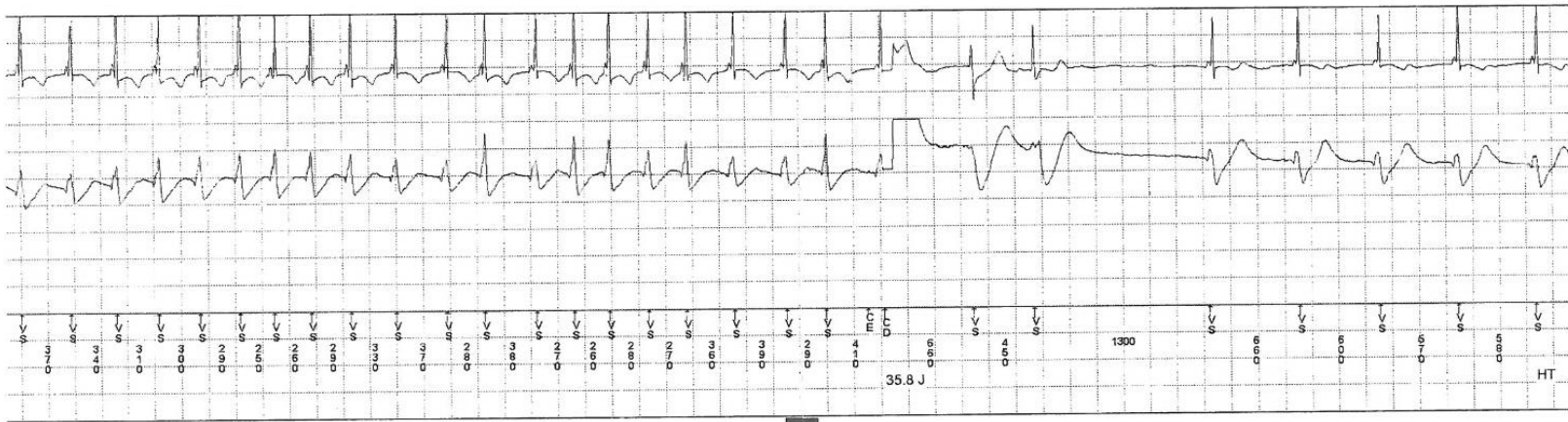
	Hazard ratio	χ^2	95% CI	p Value
Univariate risk				
Age <35 years	2.71	10.03	1.47 to 5.00	<0.01
Gender (male)	1.15	0.18	0.44 to 1.64	0.68
Left ventricular outflow tract gradient >36 mm Hg	1.96	3.16	0.93 to 3.87	0.08
Septal thickness \geq 30 mm	0.67	0.64	0.20 to 1.68	0.42
Ejection fraction <50%	1.07	0.009	0.17 to 3.52	0.92
Atrial fibrillation	1.88	4.16	1.02 to 3.52	0.04
Use of beta blocker	1.42	1.09	0.79 to 2.88	0.30
Dual-chamber implantable cardioverter defibrillator	0.94	0.03	0.51 to 1.76	0.85
Myectomy	0.82	0.35	0.42 to 1.54	0.55
Septal ablation	0.64	0.23	0.04 to 2.95	0.63
No of risk factors for cardiac arrest	1.07	0.09	0.66 to 1.72	0.78
\geq 2 risk factors	1.17	0.22	0.59 to 2.35	0.64
Multivariate risk				
Age <35 years	3.37	13.82	1.80 to 6.32	<0.01
Atrial fibrillation	2.48	7.95	1.32 to 4.74	<0.01

Predictors and Clinical Impact of Inappropriate Implantable Cardioverter-Defibrillator Shocks in Korean Patients

Jeong Hoon Yang, Kyeongmin Byeon, Hye Ran Yim, Jung Wae Park, Seung-Jung Park, June Huh, June Soo Kim, and Young Keun On

Etiology	No inappropriate shock (n=114)	Inappropriate shock (n=34)
Ischemic CMP (n, %)	36 (31.6)	9 (26.5)
Dilated CMP (n, %)	18 (15.8)	5 (14.7)
Hypertrophic CMP (n, %)	15 (13.2)	9 (26.5)
Idiopathic (n, %)	35 (30.7)	10 (29.4)
Others (n, %)	10 (8.8)	1 (2.9)

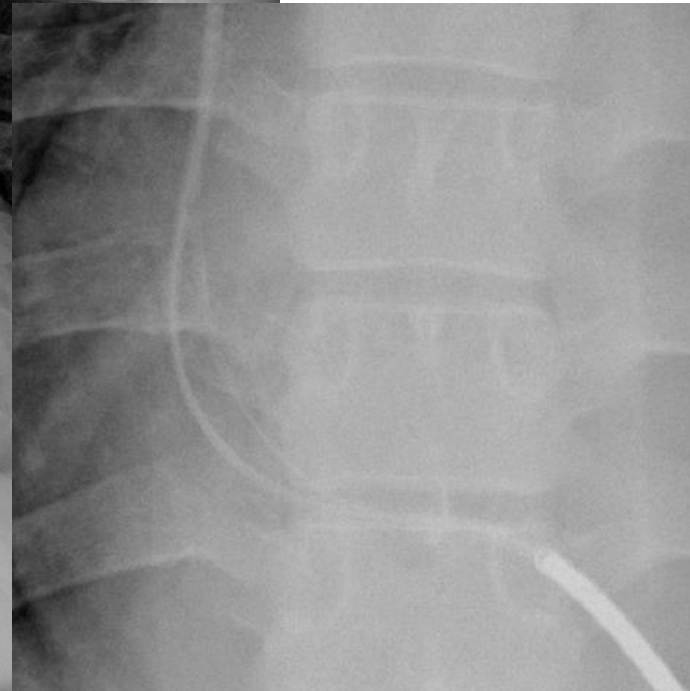
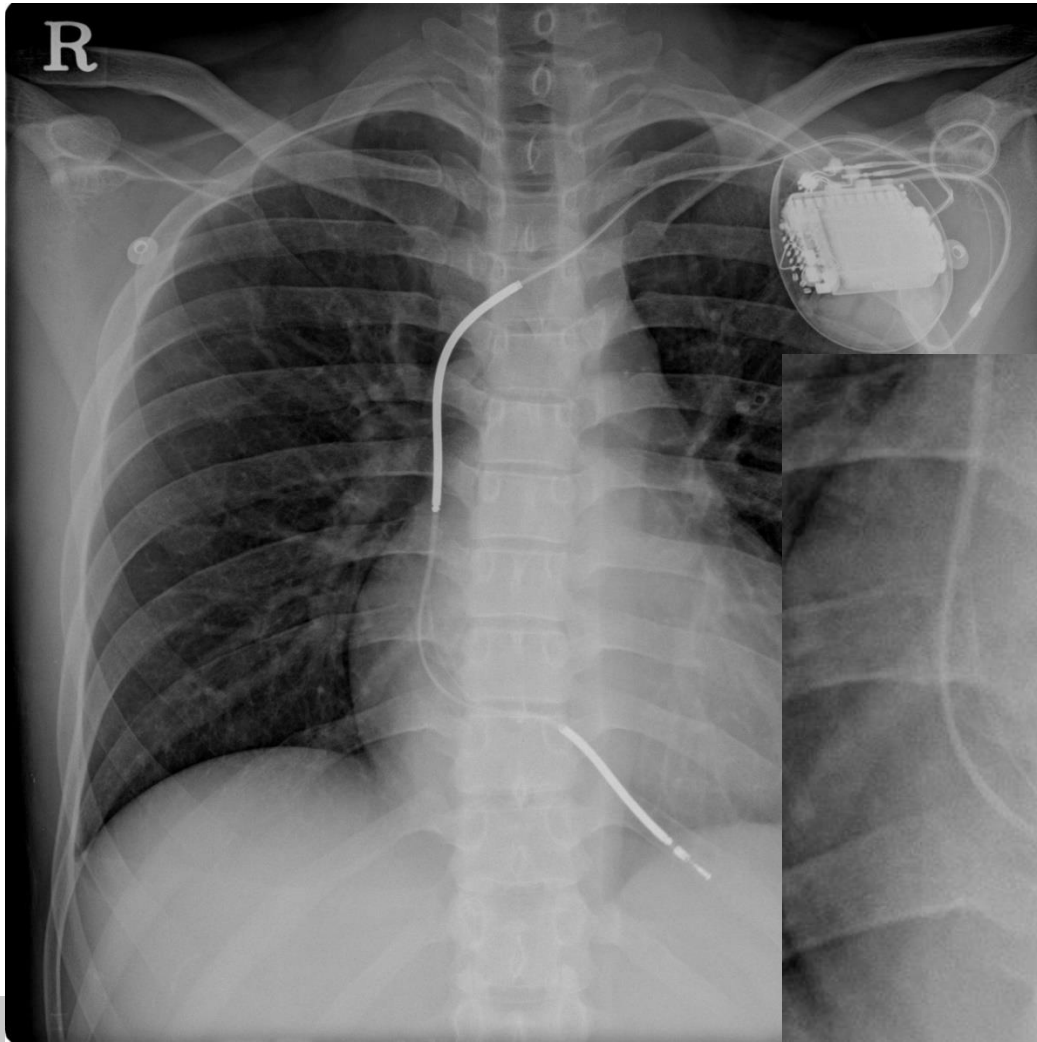
Case 1. 2012.3 ICD 삽입. 2013. 4 inappropriate shock



Case 2. 23세 남자. 2012년 12월 ICD insertion
2014년 3월 pocket site infection



Case 3. 26세 남자. 2007년 ICD 삽입
2011년 7월 recurrent shock therapy – lead problem
- 이후 현재까지 우울증으로 정신과 치료받고 있음



Tachy Episode Directory (continued)

Total Episodes: 80

Alerts (60 Episodes)	Date/Time	Episode Type	CL (ms)	Therapy	Duration	EGM	Alerts (60 Episodes)	Date/Time	Episode Type	CL (ms)	Therapy	Duration	EGM
≥3 VT/VF episodes in 24 hours	12 Jul 2011 8:01 pm	VF	275	20J	0:10		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 8:04 am	VF	165	Aborted	0:08	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 8:00 pm	VF	140	Aborted	0:10		≥3 VT/VF episodes in 24 hours	12 Jul 2011 8:52 am	VF	130	20J	0:12	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 5:58 pm	VF	270	Aborted	0:14		≥3 VT/VF episodes in 24 hours	12 Jul 2011 8:30 am	VF	150	20J	0:14	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 5:32 pm	VF	205	Aborted	0:08		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 8:30 am	VF	145	Aborted	0:08	
≥3 VT/VF episodes in 24 hours	12 Jul 2011 5:08 pm	VF	230	20J	0:12		≥3 VT/VF episodes in 24 hours	12 Jul 2011 7:36 am	VF	220	20J	0:12	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 4:58 pm	VF	240	Aborted	0:08		≥3 VT/VF episodes in 24 hours	12 Jul 2011 6:49 am	VF	250	20J	0:12	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 4:54 pm	VF	205	Aborted	0:10		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 6:49 am	VF	275	Aborted	0:08	
≥3 VT/VF episodes in 24 hours	12 Jul 2011 4:32 pm	VF	290	20J	0:16		≥3 VT/VF episodes in 24 hours	12 Jul 2011 6:26 am	VF	215	20J	0:12	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 3:45 pm	VF	230	Aborted	0:12		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 6:19 am	VF	265	Aborted	0:10	
≥3 VT/VF episodes in 24 hours	12 Jul 2011 3:32 pm	VF	130	20J	0:12		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 5:09 am	VF	140	Aborted	0:08	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 3:08 pm	VF	225	Aborted	0:08		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 5:04 am	VF	295	Aborted	0:08	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 3:06 pm	VF	165	Aborted	0:08		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 5:01 am	VF	205	Aborted	0:14	
≥3 VT/VF episodes in 24 hours	12 Jul 2011 2:39 pm	VF	300	20J	0:18		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 5:01 am	VF	125	Aborted	0:10	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 1:44 pm	VF	170	Aborted	0:12		≥3 VT/VF episodes in 24 hours	12 Jul 2011 5:00 am	VF	170	20J	0:12	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 1:44 pm	VF	205	Aborted	0:08		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 4:39 am	VF	285	Aborted	0:08	
≥3 VT/VF episodes in 24 hours	12 Jul 2011 12:49 pm	VF	125	20J	0:12		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 4:35 am	VF	260	Aborted	0:08	
≥3 VT/VF episodes in 24 hours	12 Jul 2011 12:27 pm	VF	210	20J	0:14		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 4:34 am	VF	190	Aborted	0:08	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 12:26 pm	VF	270	Aborted	0:08		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 4:24 am	VF	215	Aborted	0:08	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 12:12 pm	VF	265	Aborted	0:16		≥3 VT/VF episodes in 24 hours	12 Jul 2011 2:45 am	VF	170	20J	0:10	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 11:26 am	VF	140	Aborted	0:08		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 2:33 am	VF	150	Aborted	0:06	
≥3 VT/VF episodes in 24 hours	12 Jul 2011 11:08 am	VF	300	20J	0:14		≥3 VT/VF episodes in 24 hours	12 Jul 2011 1:27 am	VF	150	20J	0:14	
At least one shock unsuccessful, ≥3 VT/VF episodes in 24 hours	12 Jul 2011 11:07 am	VF	285	20J, Aborted	0:18		≥3 VT/VF episodes in 24 hours	12 Jul 2011 12:52 am	VF	255	20J	0:20	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 10:59 am	VF	200	Aborted	0:06		≥3 VT/VF episodes in 24 hours	12 Jul 2011 12:49 am	VF	265	20J	0:16	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 10:07 am	VF	140	Aborted	0:08		≥3 VT/VF episodes in 24 hours	12 Jul 2011 12:10 am	VF	160	20J	0:06	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 10:04 am	VF	250	Aborted	0:06		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 12:10 am	VF	190	Aborted	0:06	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 10:04 am	VF	150	Aborted	0:06		≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 12:10 am	VF	125	Aborted	0:08	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 10:01 am	VF	130	Aborted	0:08		≥3 VT/VF episodes in 24 hours, Non-sustained episode	11 Jul 2011 11:55 pm	VF	190	Aborted	0:10	
≥3 VT/VF episodes in 24 hours, Non-sustained episode	12 Jul 2011 10:00 am	VF	190	Aborted	0:06		≥3 VT/VF episodes in 24 hours, Non-sustained episode	11 Jul 2011 11:31 pm	VF	130	Aborted	0:08	

The rate of inappropriate ICD shocks and frequency of device complications in HCM patients are **not insignificant** and are most common **in younger patients** and those with atrial fibrillation.

Inappropriate ICD shocks are the most common device complication and **should be accounted for when counselling high-risk HCM patients for ICD implantation**

Consider syncope

- One of the most difficult clinical presentations in pts. with hCMP
 - ≡ 15% neurally mediated syncope or unexplained syncope
- Recent (< 6 months) unexplained syncope
 - associated with an increased risk for sudden death in all age groups
- Remotes (> 6 months) episodes of syncope
 - not associated with increased risk in older patients

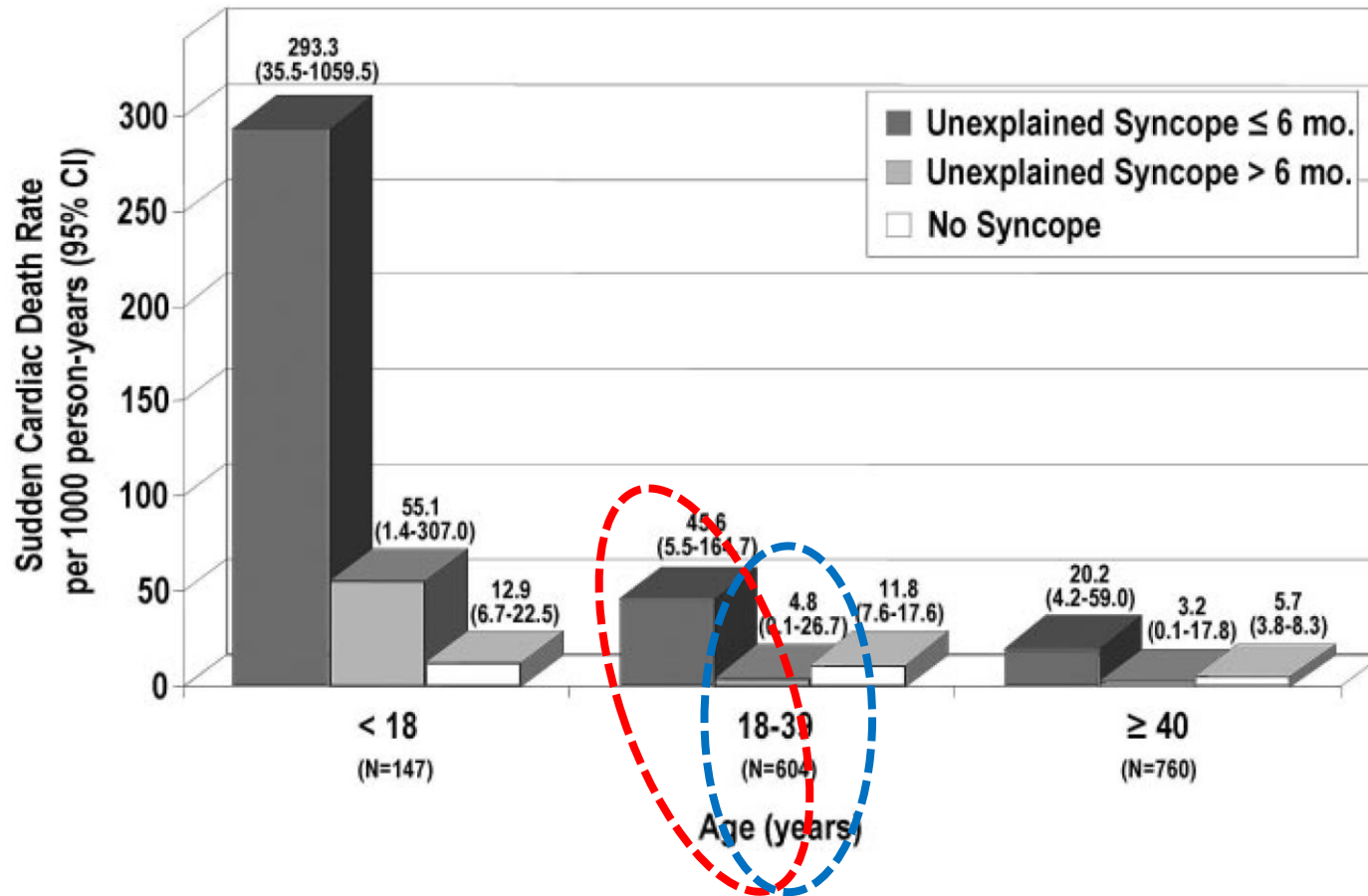
Syncope and Risk of Sudden Death in Hypertrophic Cardiomyopathy

Paolo Spirito, Camillo Autore, Claudio Rapezzi, Paola Bernabò, Roberto Badagliacca, Martin S. Maron, Sergio Bongioanni, Fabio Coccolo, N.A. Mark Estes, Caterina S. Barilla, Elena Biagini, Giovanni Quarta, Maria Rosa Conte, Paolo Bruzzi and Barry J. Maron

Circulation. 2009;119:1703-1710; originally published online March 23, 2009;

Variable	Death Due to Any Cause				HCM-Related Death (Sudden Death, HF, or Stroke)				Sudden Death			
	No. of Deaths (%)	Incidence per 1000 Person-Years (95% CI)	Relative Risk (95% CI)	P	No. of Deaths (%)	Incidence per 1000 Person-Years (95% CI)	Relative Risk (95% CI)	P	No. of Sudden Deaths (%)	Incidence per 1000 Person-Years (95% CI)	Relative Risk (95% CI)	P
Without syncope (n=1306)	182 (14)	24.6 (21.2–28.5)	1 (Reference)		131 (10)	17.7 (14.8–21.0)	1 (Reference)		61 (5)	8.2 (6.3–10.6)	1 (Reference)	
Unexplained syncope (n=153)	22 (14)	29.4 (18.4–44.5)	1.19 (0.74–1.90)	0.44	15 (10)	20.0 (11.2–33.0)	1.13 (0.64–1.97)	0.65	11 (7)	14.7 (7.3–26.2)	1.78 (0.88–3.51)	0.08
Unexplained and recurrent syncope* (n=63)	10 (16)	34.6 (16.6–63.6)	1.41 (0.69–2.67)	0.30	6 (10)	20.8 (7.6–45.2)	1.18 (0.47–2.76)	0.70	3 (5)	10.4 (2.1–3.4)	1.26 (0.32–4.15)	0.52†
Neurally mediated syncope (n=52)	10 (19)	37.8 (18.1–69.4)	1.53 (0.76–3.02)	0.19	8 (15)	30.2 (13.1–59.5)	1.71 (0.77–3.60)	0.14	2 (4)	7.6 (0.9–27.2)	0.91 (0.00–3.83)	1.0†

Syncope



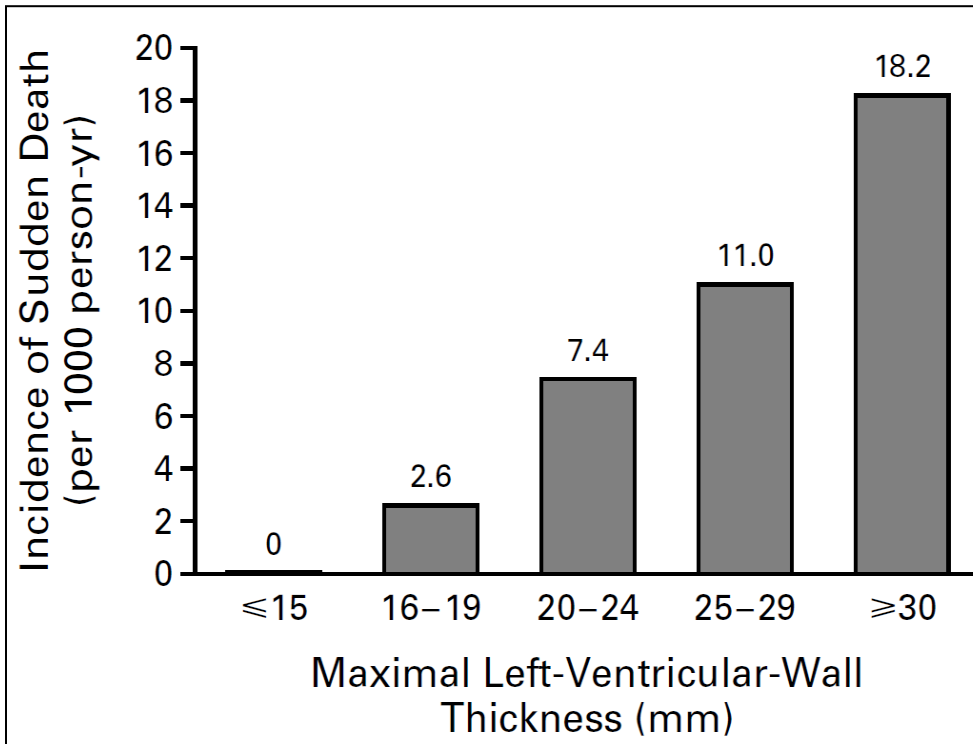
Consider septal thickness $\geq 30\text{mm}$

$\geq 30\text{ mm}$ is independently
associated with SCD

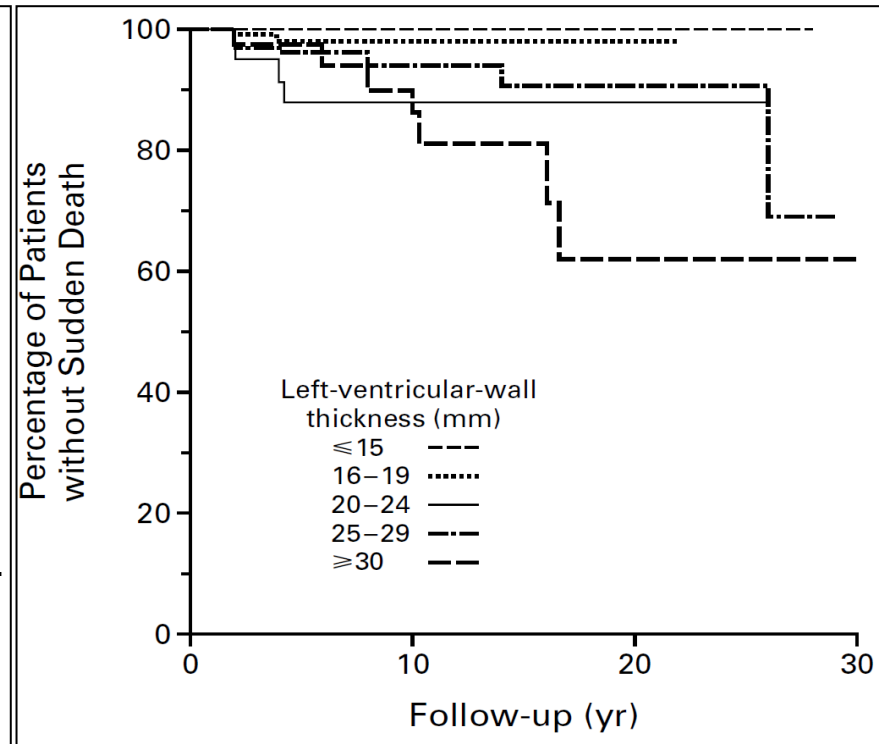
Elliott PM, et al. J Am Coll Cardiol. 2000;26:2212-8
Spirito P, et al. Circulation 2009;119:170-3-10
Spirito P, et al. N Engl J med 2000;342:1778-85

MAGNITUDE OF LEFT VENTRICULAR HYPERTROPHY AND RISK OF SUDDEN DEATH IN HYPERTROPHIC CARDIOMYOPATHY

PAOLO SPIRITO, M.D., PIETRO BELLONE, M.D., KEVIN M. HARRIS, M.D., PAOLA BERNABÒ, M.D.,
PAOLO BRUZZI, M.D., PH.D., AND BARRY J. MARON, M.D.



Relation between maximal LV wall thickness and the risk of SCD



Kaplan-Meier Estimates of the proportions of SCD

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VARIABLE	No. of PATIENTS	SUDDEN DEATH		DEATH DUE TO HEART FAILURE		DEATH FROM ANY CAUSE		DURATION OF FOLLOW-UP person-yr
		NO. OF PATIENTS	INCIDENCE PER 1000 PERSON-YR (95% CI)	NO. OF PATIENTS	INCIDENCE PER 1000 PERSON-YR (95% CI)	NO. OF PATIENTS	INCIDENCE PER 1000 PERSON-YR (95% CI)	
Age								
<20 yr	64	8	14.2 (6.1–28.0)	0	0 (0–6.5)	8	14.2 (6.1–28.0)	563
20–39 yr	120	3	4.0 (0.8–11.6)	3	4.0 (0.8–11.6)	7	9.3 (3.7–19.2)	754
40–59 yr	154	6	5.5 (2.0–11.9)	3	2.7 (0.6–8.0)	18	16.4 (9.7–26.0)	1094
≥60 yr	142	6	8.6 (3.1–18.7)	9	12.9 (5.9–24.4)	32	45.8 (31.3–64.6)	699
P value			0.39		0.003		<0.001	

The highest rate of SCD was observed in the **youngest patients (< 20 yrs)** with a wall thickness of 30mm or more

Consider LVOT obstruction

- Resting LVOT pressure gradient ≥ 30 mmHg
 - SCD is positively correlated with severity of LVOT obstruction
 - independent determinant of symptoms of progressive HF and death
- Relief of OT obstruction through surgical myectomy is associated with very low rates of SCD

Effect of Left Obstruction in Hypertro

Martin S. Maron, M.D., I
Susan A. Casey, R.N.,
Franco Cecch

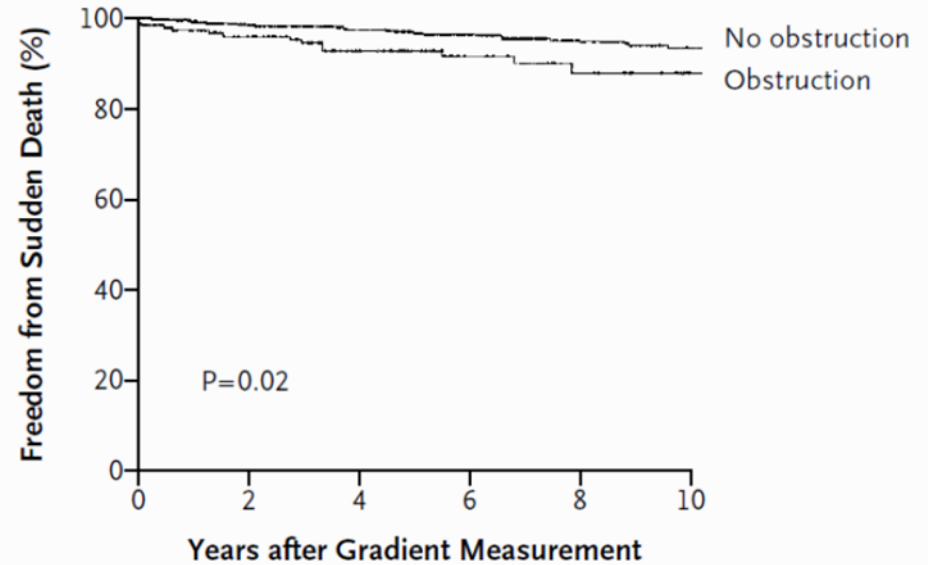


Table 2. Results of Age-Adjusted Multivariate Cox Pro and Outcome.*

No. at Risk	0	2	4	6	8	10
No obstruction	770	557	464	334	231	188
Obstruction	224	144	103	66	39	25

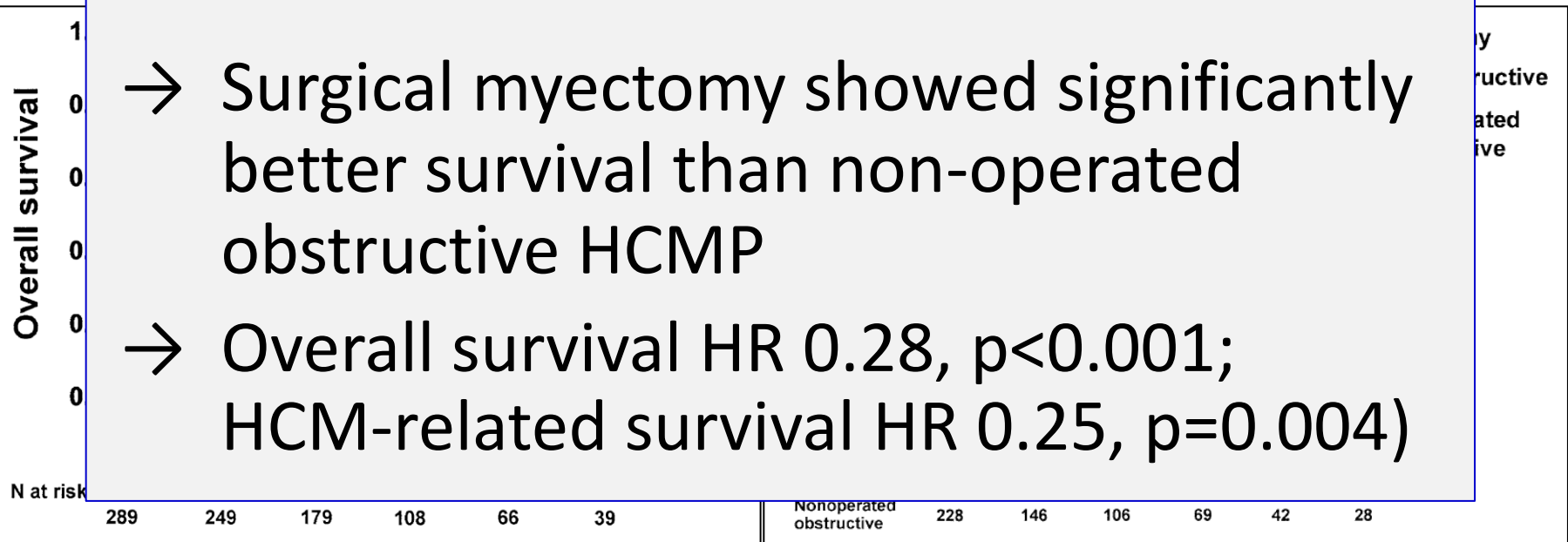
Variable	Death from Any Cause		HCM-Related Death		Progression to NYHA Class III or IV or Death from Heart Failure or Stroke†		Sudden Death from HCM‡	
	Relative Risk (95% CI)	P Value	Relative Risk (95% CI)	P Value	Relative Risk (95% CI)	P Value	Relative Risk (95% CI)	P Value
Left ventricular outflow obstruction (≥30 mm Hg)	1.6 (1.1–2.2)	0.02	1.6 (1.1–2.4)	0.02	2.7 (2.0–3.5)	<0.001	1.9 (1.1–3.5)	0.01†
NYHA class II, III, or IV at entry	1.5 (1.1–2.1)	0.02	1.9 (1.2–2.9)	0.002	3.4 (2.4–4.8)	<0.001	—	0.12
Paroxysmal or chronic atrial fibrillation	1.4 (1.0–1.9)	0.04	1.6 (1.1–2.4)	0.01	1.3 (1.1–1.6)	0.046	—	0.72
Maximal left ventricular thickness ≥30 mm	1.6 (1.1–2.4)	0.01	1.8 (1.1–2.8)	0.01	—	0.09	—	0.82
Female sex	—	0.22	—	0.29	1.4 (1.1–1.8)	0.02	—	0.75

Hypertrophic and Dilated Cardiomyopathy

Long-Term Effects of Surgical Septal Myectomy on Survival in Patients With Obstructive Hypertrophic Cardiomyopathy

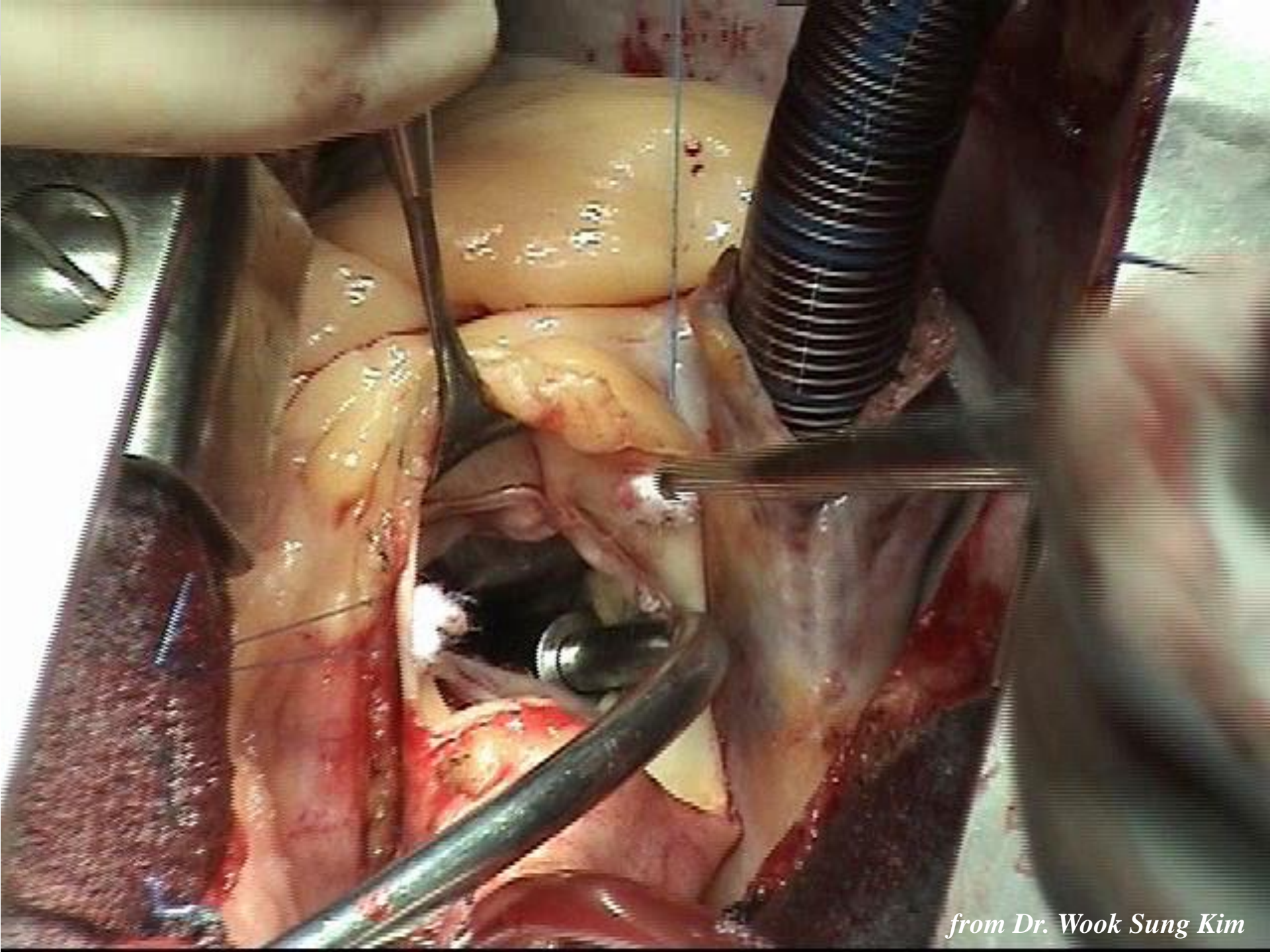
Steve R. Ommen, MD, FACC* Barry J. Maron, MD, FACC,*‡ Iacopo Olivotto, MD,§
 Martin S. Maron, MD|| Franco Cecchi, MD§ Sandro Betocchi, MD, FACC¶
 Berna Robert Gordon Roches

- **Subgroup analysis with ≤ 45 yrs**



- Survival free from all-cause mortality
 Surgical myectomy for obstructive hcmp & general population

- Survival free from sudden cardiac death



from Dr. Wook Sung Kim

Septal Myectomy

Studies of early and long-term results of myectomy from experienced centers

Authors/year	Time period	Patients (N)	Early mortality (all cases) (%)	Early mortality (isolated myectomy) (%)	Mean follow-up time (y)	Survival at 5 y (%)	Survival at 10 y (%)
Cohn et al 1992 [46]	1972–1991	31	0	0	6.5	100	86
Schulte et al 1993 [44]	1963–1991	364	4.9 ^a	2.9 ^b	8.2	—	88
Ten Berg et al 1994 [47]	1977–1992	38	0	0	6.8	100	—
Heric et al 1995 [48]	1975–1993	178	6.2	4	3.7	86	70
Schoendube et al 1995 [49]	1979–1992	58	1.7	—	7.0	—	86
Robbins and Stinson 1996 [50]	1972–1994	158	3.2	2.3	6.1	85	72
Schoenbeck et al 1998 [51]	1965–1995	110	3.6	—	11.7	93	80
Woo et al 2005 [52]	1978–2002	338	1.5	0.8	7.7	95	83
Ommen et al 2005 [53]	1983–2001	289 ^c	—	0.8	6.2	96	83

^a 3.0% between years 1981 to 1990.

^b 1.3% between years 1981 to 1990.

^c Surgical cohort only included patients undergoing isolated myectomy. *Heart Failure Clin* 2007;3:275-288

Clinical and Echocardiographic predictors of overall mortality

Variable*	Overall Survival		HCM-Related Survival	
	HR (95% CI)	p Value	HR (95% CI)	p Value
Age >50 yrs	2.6 (1.6–4.3)	<0.0001	2.0 (1.1–3.4)	0.01
Female	2.0 (1.3–3.2)	0.002	2.1 (1.2–3.6)	0.006
Septal thickness ≥20 mm	1.7 (1.02–2.7)	0.04	2.0 (1.1–3.6)	0.02
Resting LVOT gradient ≥64 mm Hg	1.6 (0.98–2.5)	0.06	1.7 (1.0–3.0)	0.0496
Invasive treatment†	0.6 (0.4–0.97)	0.04	—	0.30 (NS)

Toronto, Ontario, Canada

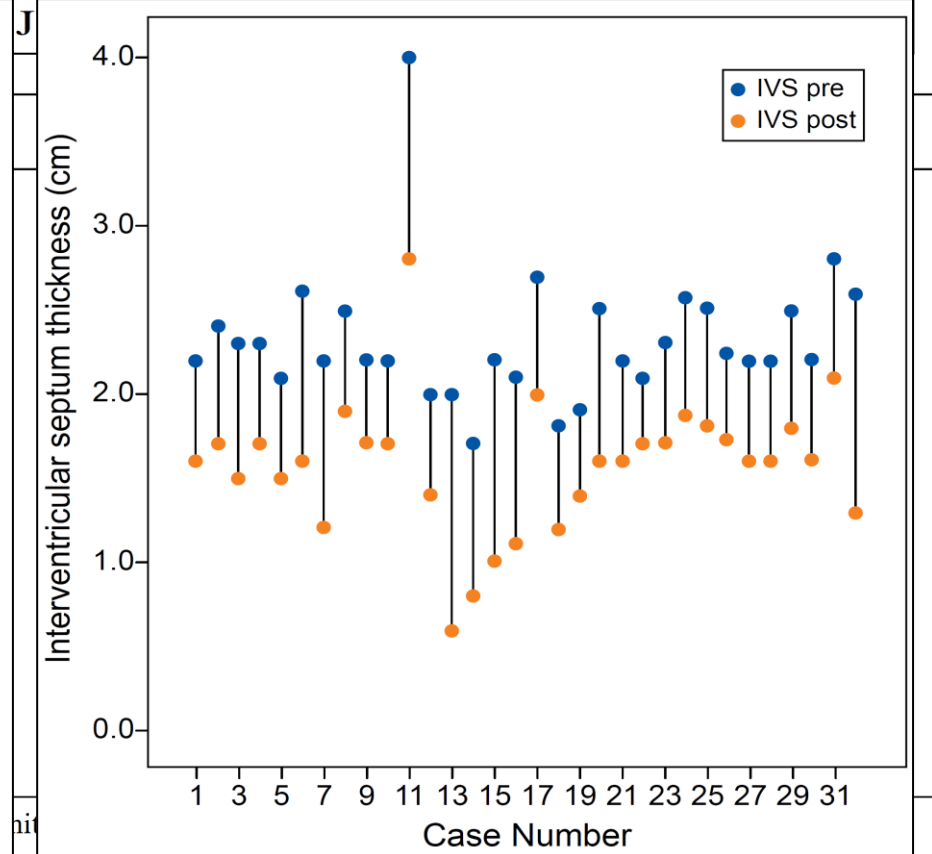
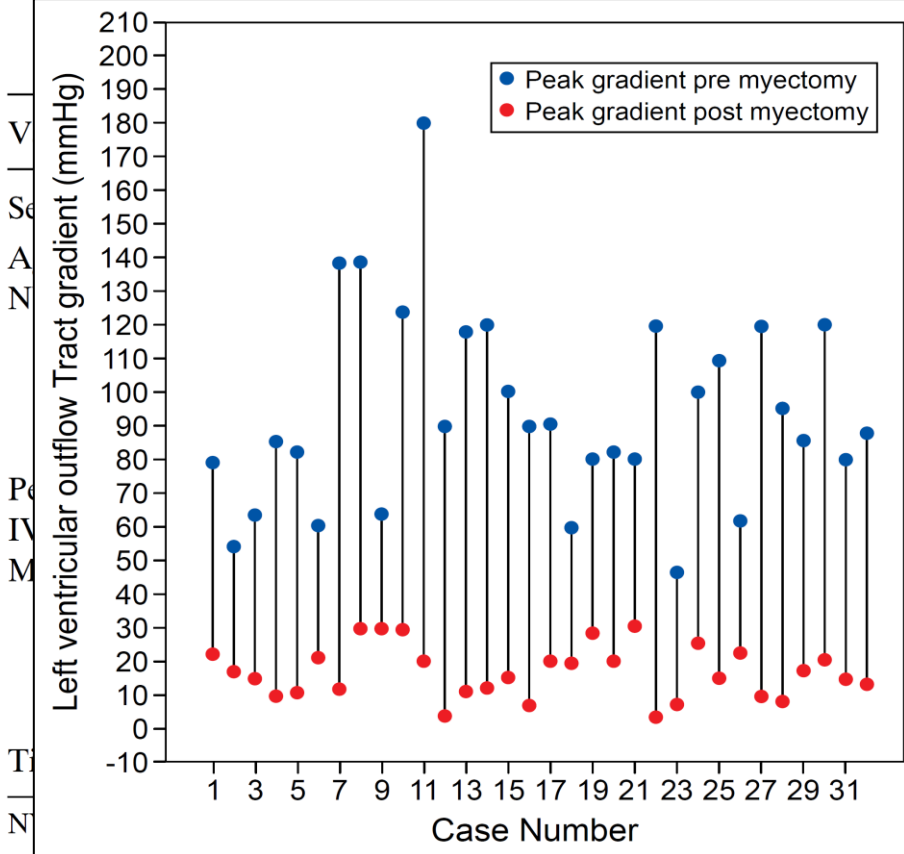
Overall mortality and equivalents of mortality

	Conservative Group (n = 246)	Invasive Group (n = 403)	Total (n = 649)
Mortality			
HCM-related mortality	19 (7.7)	28 (6.9)	47 (7.2)
Sudden cardiac death	8 (3.3)	7 (1.7)	15 (2.3)
Non-HCM-related death	16 (6.5)	8 (2.0)	24 (3.7)
Overall mortality	35 (14.2)	36 (8.9)	71 (10.9)
Equivalents of mortality			
Resuscitated cardiac arrest	2 (0.8)	4 (1.0)	6 (0.9)
Appropriate ICD discharge*	1 (0.4)	3 (0.7)	4 (0.6)
Total mortality and equivalents of mortality	38 (15.4)	43 (10.7)	81 (12.5)

- Conservative group (n=246)
Invasive group (n=403)
- Invasive procedure;
 - Septal myectomy (n=287)
 - SEA (n=85)
 - DDD pacing (n=31)
- septal thickness ≥ 13 mm
- Mean f/u; 7.2 ± 5.5 yrs

Surgical Septal Myectomy for Hypertrophic Cardiomyopathy in Greece: A Single-Center Initial Experience

GEORGIOS K. EFTHIMIADIS^{1*}, ANTONIS PITSIS^{2*}, EFSTATHIOS D. PAGOURELIAS¹,
VASILEIOS KAMPERIDIS¹, TIMOTHEOS KELPIS², SOULTANA MEDITSKOU³, STAVROS HADJIMILTIADES¹,



Does Myectomy Convey Survival Benefit in hCMP ?

Clinical and Echocardiographic Determinants of Long-Term Survival After Surgical Myectomy in Obstructive Hypertrophic Cardiomyopathy

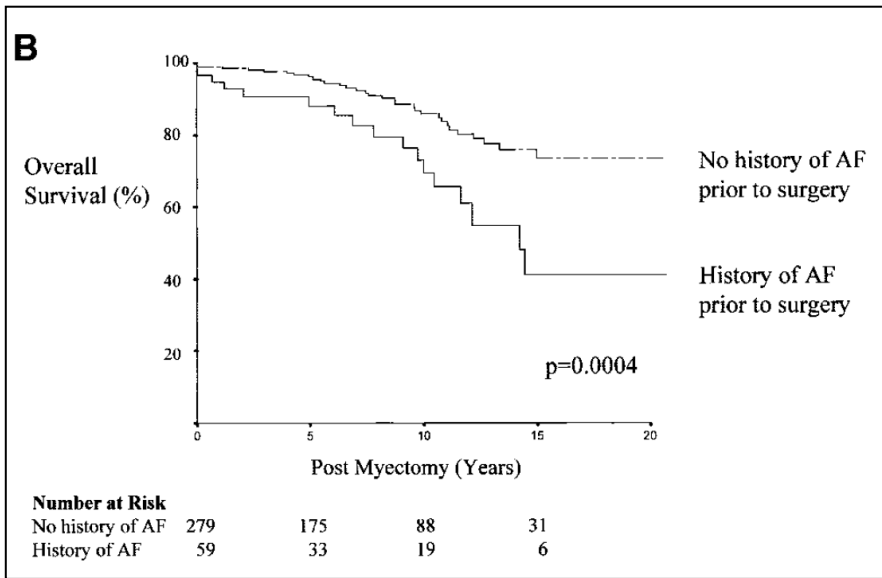
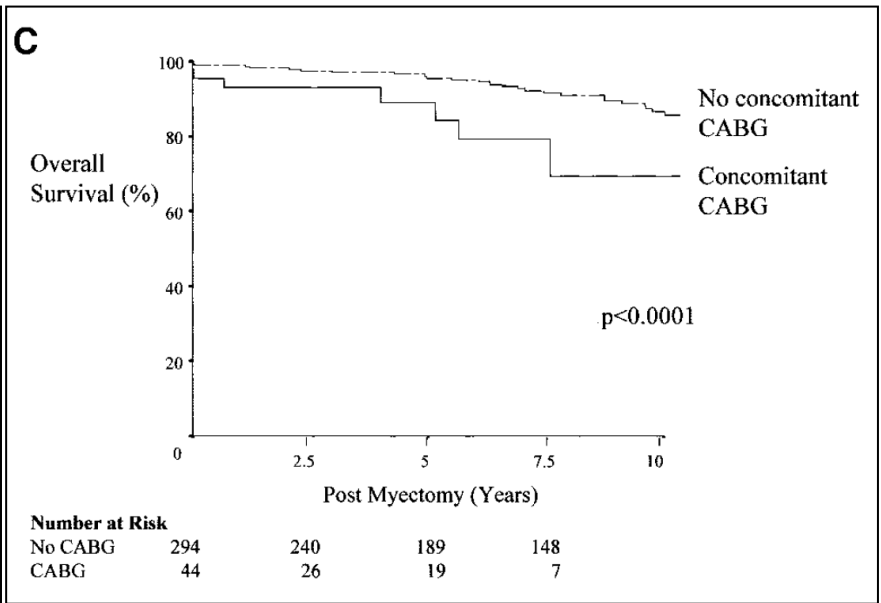
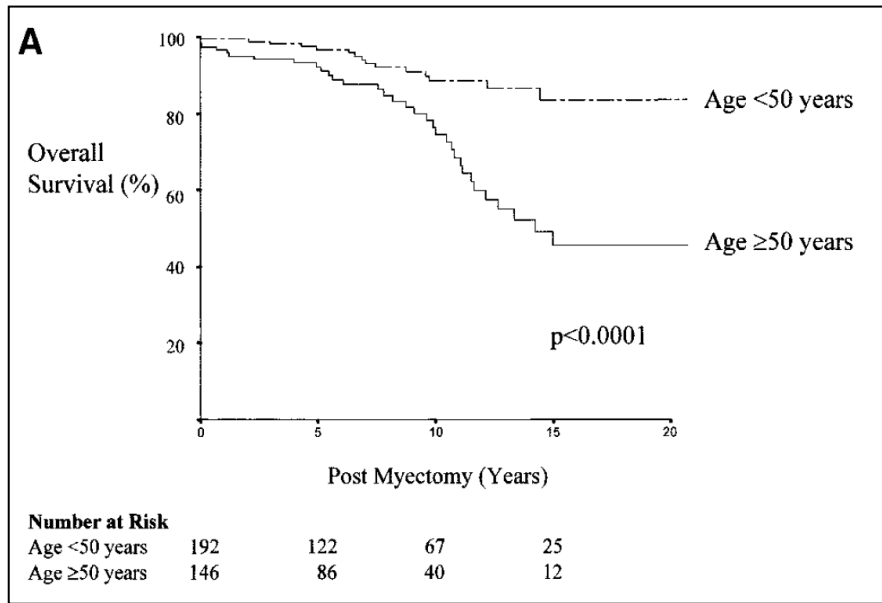
Anna Woo, William G. Williams, Richard Choi, E. Douglas Wigle, Evelyn Rozenblyum, Katie Fedwick, Samuel Siu, Anthony Ralph-Edwards and Harry Rakowski

Circulation. 2005;111:2033-2041; originally published online April 11, 2005;

TABLE 3. Clinical and Echocardiographic Predictors of Overall Mortality Among 338 Patients Who Underwent Myectomy

Variable	Individual Analysis HR		Multivariable Analysis HR	
	(95% CI)	P	(95% CI)	P
Age \geq 50 y*	3.3 (1.9–5.8)	<0.0001	2.8 (1.5–5.1)	0.0008
Female gender*	1.7 (1.0–2.8)	0.06	2.5 (1.5–4.3)	0.0009
History of preoperative AF*	2.7 (1.6–4.8)	0.0004	2.2 (1.2–4.0)	0.008
LA diameter \geq 46 mm*	2.9 (1.6–5.3)	0.0005	2.9 (1.6–5.4)	0.0008
Septal/posterior thickness ratio \geq 1.8*	0.5 (0.3–0.8)	0.009	0.8 (0.4–1.5)	0.5
Concomitant CABG*	4.8 (2.3–10.2)	<0.0001	3.7 (1.7–8.2)	0.001

Kaplan-Meier survival curve for total mortality in 338 patients who underwent myectomy (solid line)



Kaplan-Meier plots of overall postmyectomy survival stratified by different variables:

(A) age 50 years or age 50 years at time of surgery ($P < 0.0001$),

(B) history of preoperative AF ($P = 0.0004$),

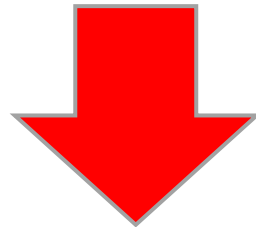
(C) need for concomitant CABG surgery ($P < 0.0001$).

What is the most appropriate next step in management ?

- A. Ambulatory (Holter) ECG
- B. EP study with aggressive programmed stimulation protocol to induce ventricular arrhythmias
- C. ICD implant for primary prevention
- D. Refer for myectomy operation

In this case,

1. 35 yo (relatively young) with unexplained syncope
2. LVOT PG = 37mmHg/ septal thickness = 30 mm
3. Normal life expectancy
4. Symptoms (?) : dyspnea, limited exercise capacity
5. Progressive disease complications: HF, atrial fibrillation



Surgical Myectomy

In conclusions

- In patient with HCMP,
 - a thorough discussion of the strength of evidence, benefits, and risks to allow the informed patient's active participation in decision making of the procedures
 - please consider patient's age, clinical symptoms & signs
 - myectomy is a safe and effective procedure for symptom control and survival benefit

HCMP registry in SMC (n=1457)

septal thickness \geq 30mm 환자 review (1.1%)

n	type	age	sex	Risk factors	medication	clinical
1	septal/apical	51	M	-	verapamil, carvedilol	
2	septal/ant	62	F	family hx	bisoprolol, diltiazem	
3	septal	14	M	recent onset syncope	bisoprolol	ICD, inappropriate shock (+)
4	septal	16	M	ventricular tachycardia	amlodipine	ICD + Myectomy
5	septal	46	M	-	bisoprolol	
6	septal	36	M	family hx	bisoprolol	환자 및 보호자와 상의 후 ICD 넣지 않음
7	septal	42	F	-	bisoprolol	
8	septal	50	M	-	bisoprolol, diltiazem	
9	septal	17	M	non-sustained VT	-	Myectomy, 환자 및 보호자와 상의 후 ICD 넣지 않음
10	septal	19	M	-	atenolol	
11	septal	56	M	non-sustained VT	carvedilol	환자 및 보호자와 상의 후 ICD 넣지 않음
12	septal	31	M	non-sustained VT	bisoprolol	환자 및 보호자와 상의 후 ICD 넣지 않음
13	septal	36	M	syncope, inadequate BP rise	verapamil, bisoprolol	myectomy or ICD 고려 중이나 환자가 결심을 못함
14	septal	44	M	-	verapamil, carvedilol	verapamil, carvedilol 약에 반응 없으면 수술 고려
15	septal/apical	74	M	-	amiodarone	
16	septal	35	M	VT, family hx		환자가 ICD 삽입 또는 myectomy 수술을 결정하지 못함
17	septal	71	M	-	carvedilol	

경청해 주셔서 감사합니다

