News in HCM / HOCM

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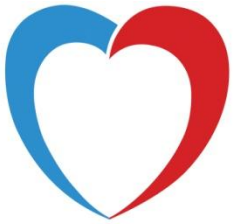
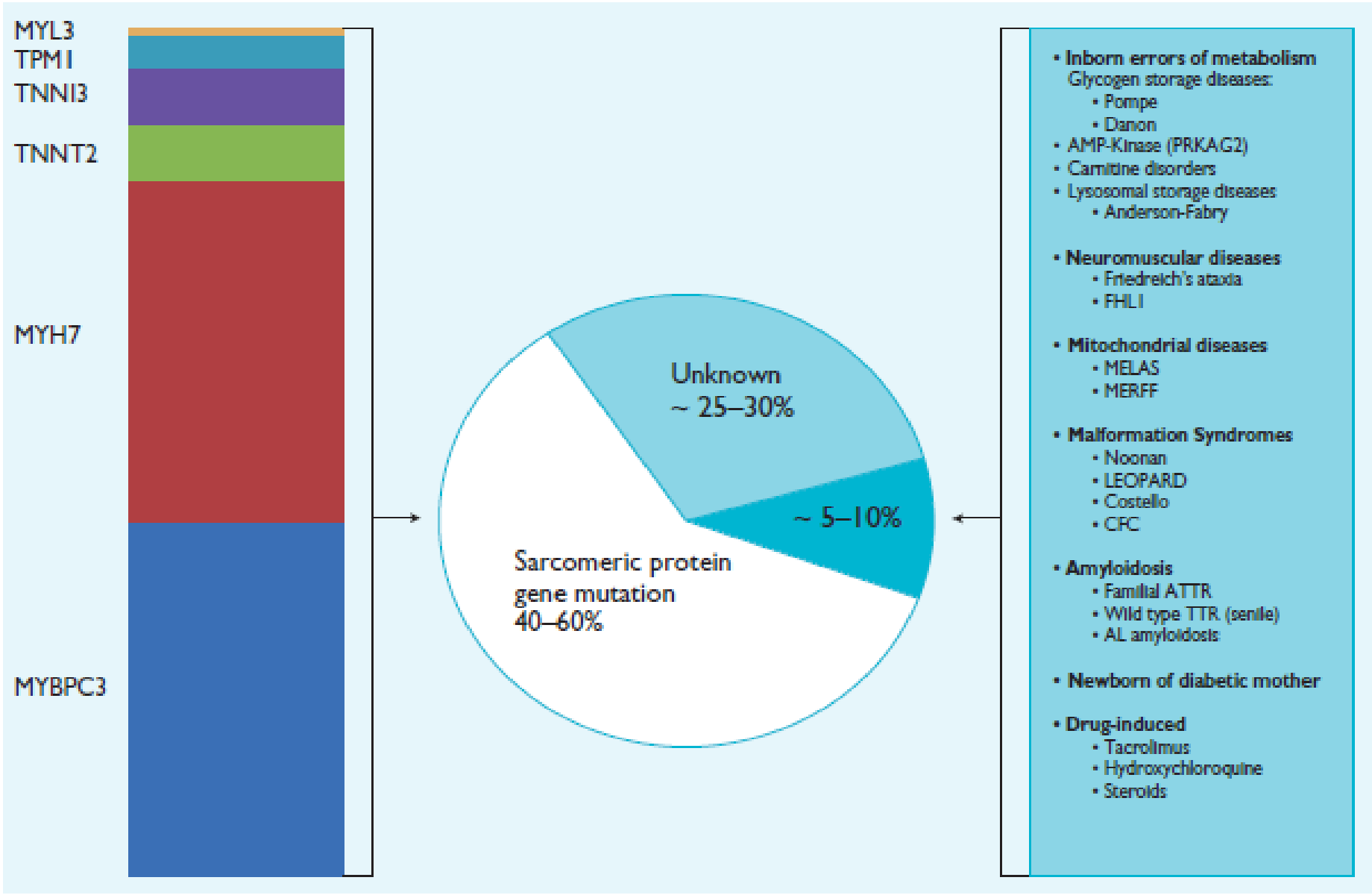


DEPARTMENT OF CARDIOLOGY
MOTOL UNIVERSITY HOSPITAL, 2nd MEDICAL SCHOOL,
CHARLES UNIVERSITY, PRAGUE, CZECH REPUBLIC

Genotype in HCM



Elliott et al. EHJ 2014

Risk of sudden cardiac death

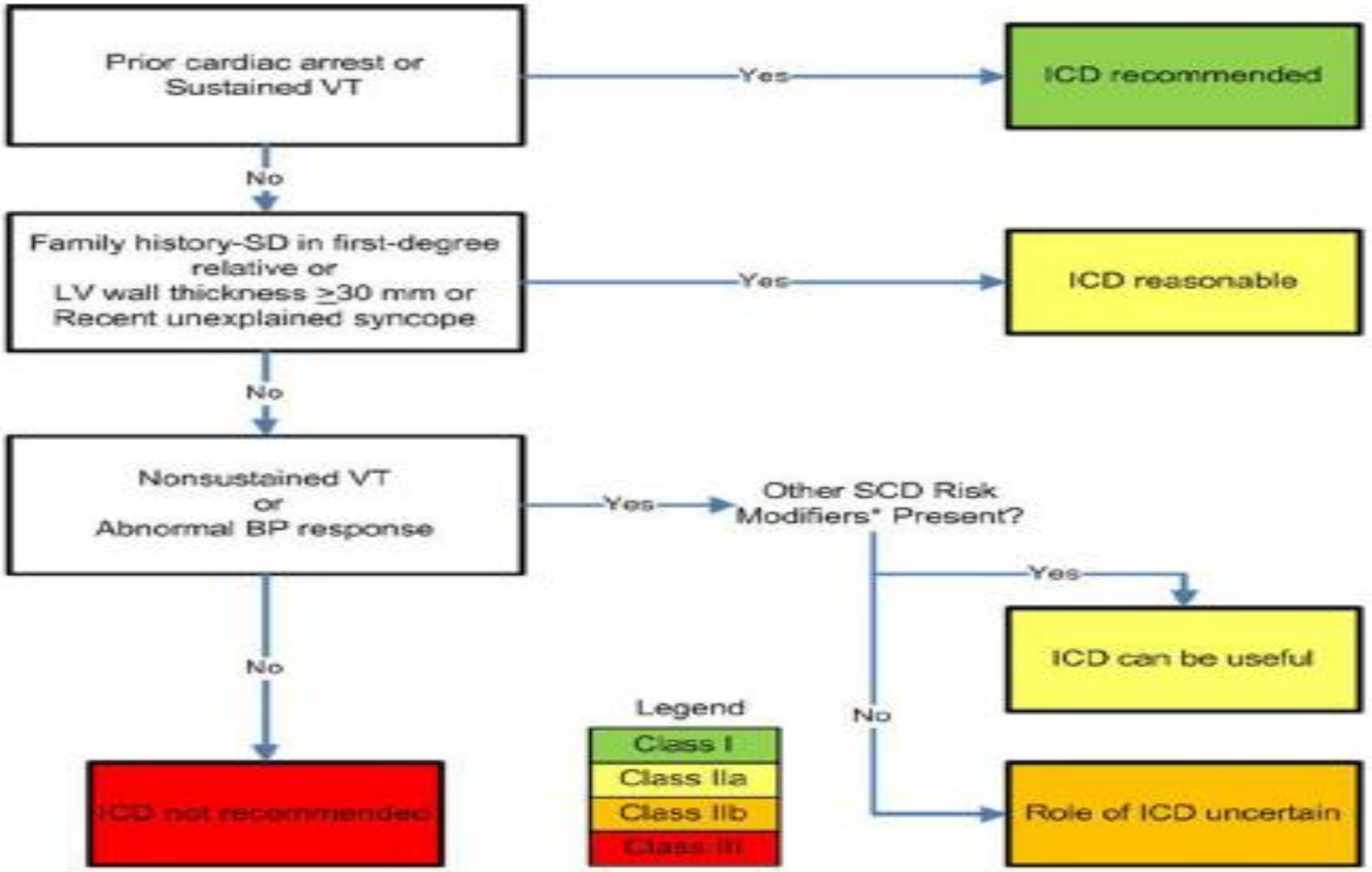
Prognosis

- **SCD**
- Most often cause of SCD in young athletes
- One of the most often causes in childern and young adults
- Obstruction is risk

- **Mortality events**
- 2% per year in childern and young adults
- 1% per year in adults

!Despite therapy, HCM-attributable death occurs in ≈ 1% of HCM pts per year!

Risk stratification - US Guidelines



Gersh et al 2011

Lack of specificity in US model

- Strict compliance to US model could lead to ICD implantation even in 40–60% of HCM patients with a low annual rate of appropriate ICD discharge of ≈2% in large HCM cohorts with implanted ICDs *(Jahnlová IJC 2015;Maron 2015 JACC; O´Mahony Heart 2012).*
- There was a need for a more specific model that would better stratify the individual risk of each patient.

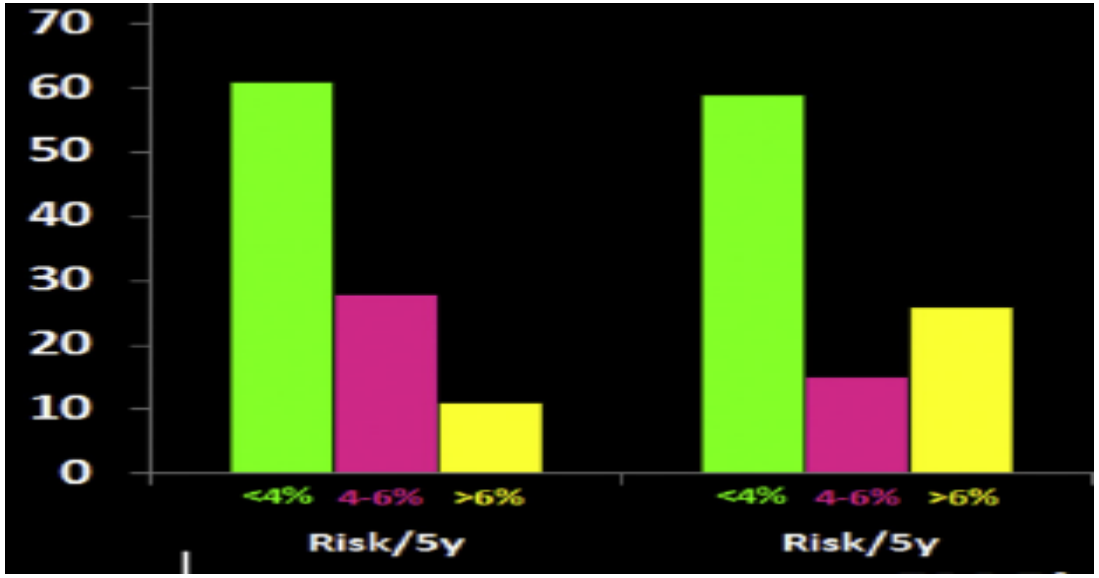
HCM Risk-SCD Calculator

Age	30	**Years**	*Age at evaluation*
Maximum LV wall thickness	22	**mm**	*Transthoracic Echocardiographic measurement*
Left atrial size	54	**mm**	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	56	**mmHg**	*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernouilli equation: Gradient= $4V^2$, where V is the peak aortic outflow velocity*
Family History of SCD	◉ No ○ Yes		*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).*
Non-sustained VT	◉ No ○ Yes		*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.*
Unexplained syncope	◉ No ○ Yes		*History of unexplained syncope at or prior to evaluation.*

Risk of SCD at 5 years (%):	4.75
ESC recommendation:	ICD may be considered

Lack of sensitivity in EU model

- This model was shown to be sufficiently specific and associated with a lower rate of ICD implantations (20–26%) *(Jahnlová IJC 2015; Maron 2015 AJC).*
- Sufficient specificity.
- Insufficient sensitivity.



SCD ICD discharge

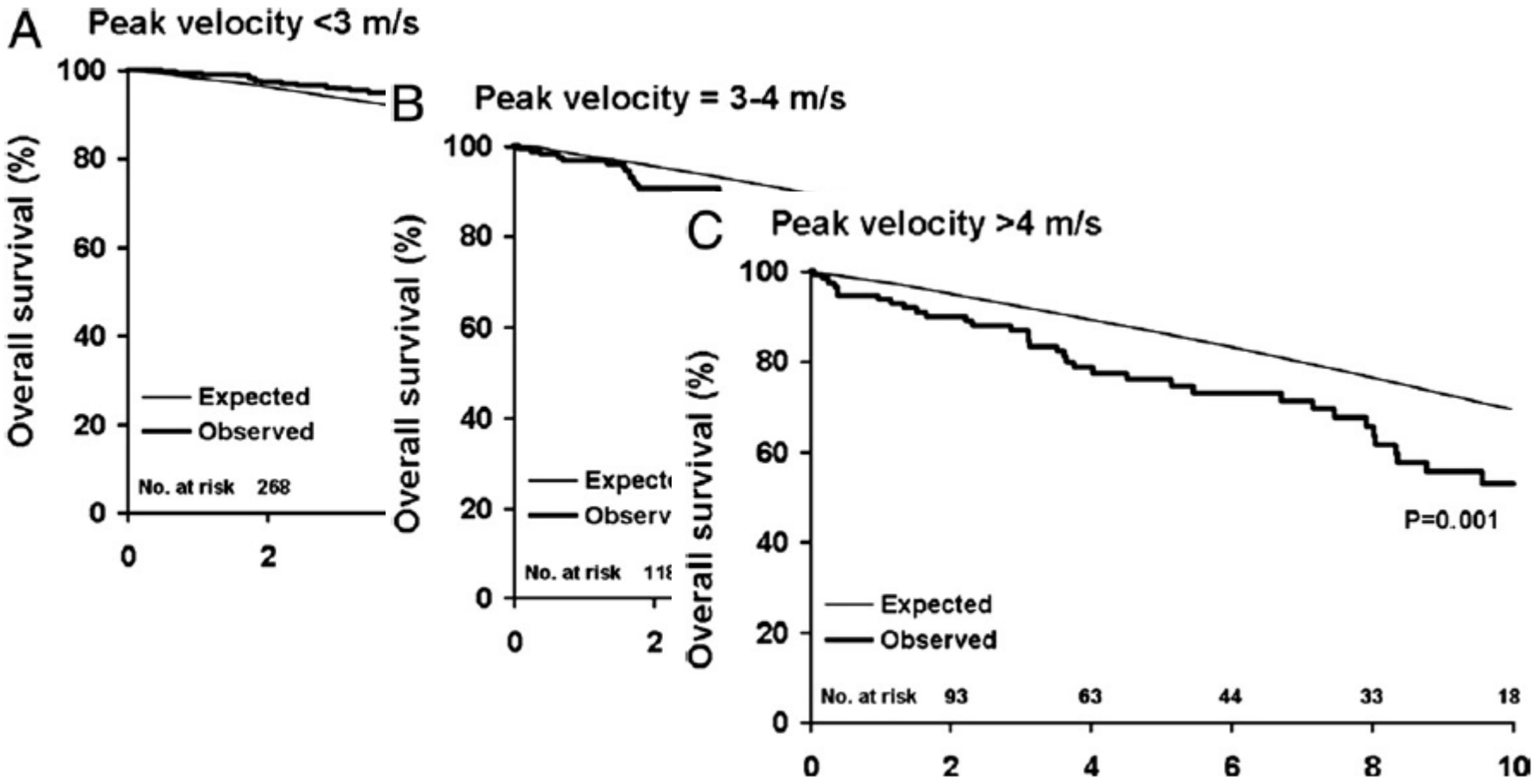
Maron et al. AJC 2015

Take-home messages

- All patients should be informed about both models and their (dis)advantages.
- ICD-related complications including inappropriate discharges, infections, and lead or device dysfunctions should be mentioned since their incidence is higher than the incidence of appropriate discharges!

Survival and LV obstruction

Obstruction and survival
(HOCM vs matched general population)



Sorajja et al. JACC 2009

Obstruction and survival

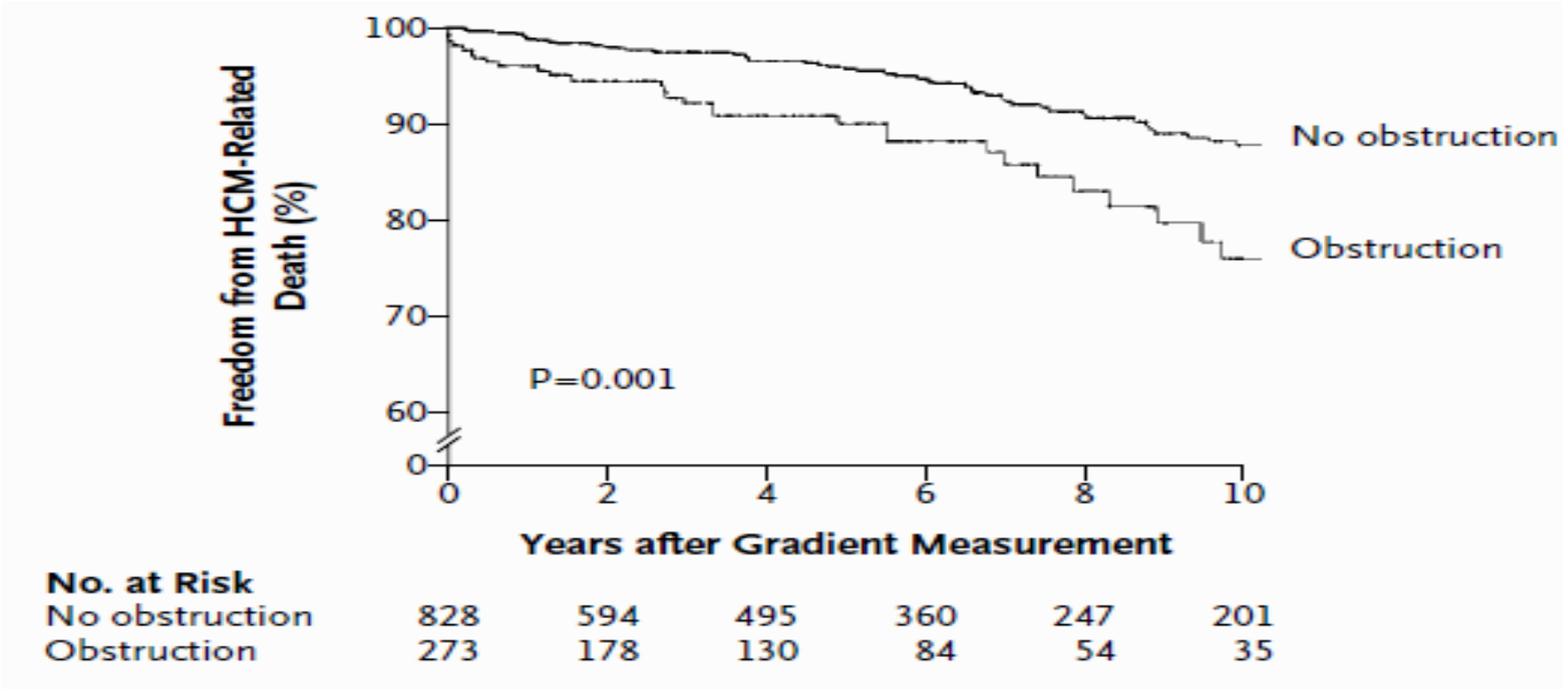
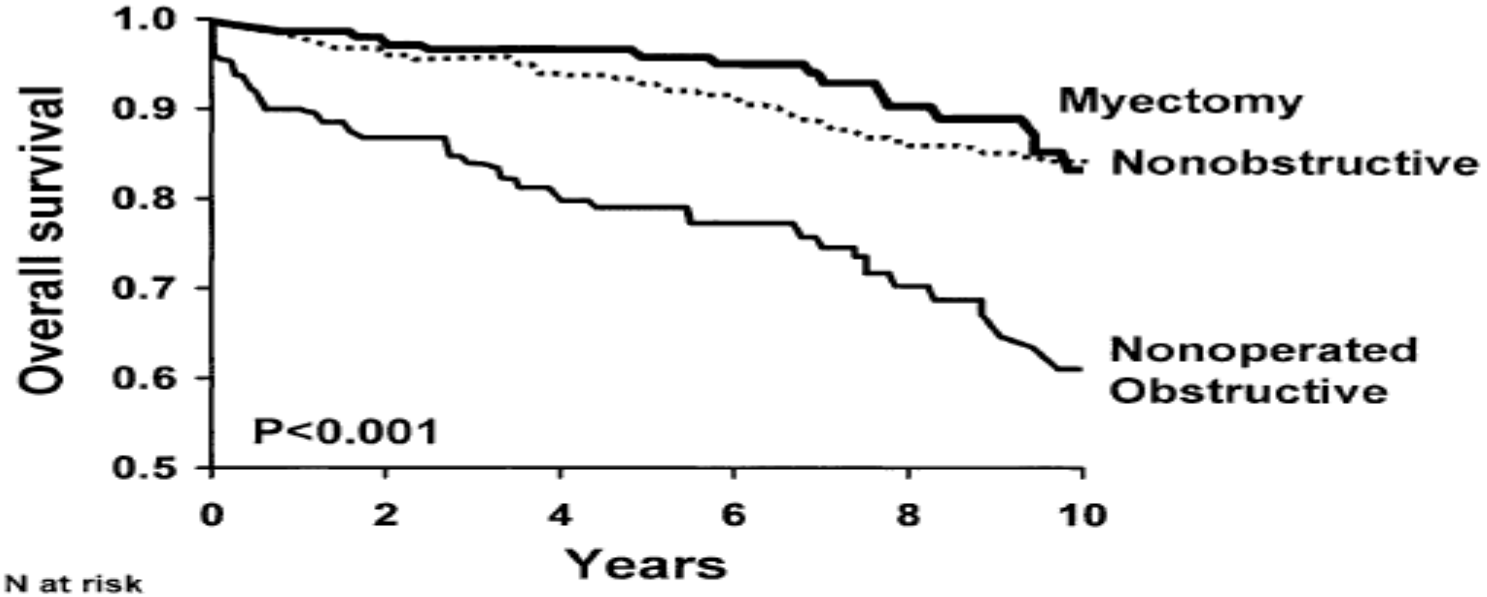


Figure 1. Probability of Hypertrophic Cardiomyopathy (HCM)–Related Death among 273 Patients with a Left Ventricular Outflow Gradient of at Least 30 mm Hg under Basal Conditions and 828 Patients without Obstruction at Entry.

Maron M et al. NEJM 2003

Survival and myectomy



N at risk						
Myectomy	289	249	179	108	66	39
Nonobstructive	820	587	490	355	244	201
Nonoperated obstructive	228	146	106	69	42	28

Figure 2. Survival free from all-cause mortality in three hypertrophic cardiomyopathy patient subgroups: surgical myectomy (n = 289), nonoperated with obstruction (n = 228), and nonobstructive (n = 820). Overall log-rank, $p < 0.001$; myectomy versus nonoperated obstructive hypertrophic cardiomyopathy, $p < 0.001$; myectomy versus nonobstructive hypertrophic cardiomyopathy, $p = 0.8$.

Ommen et al. JACC 2005

Cardiovascular mortality events and post-ASA obstruction

Freedom from cardiovascular mortality events occurring after the first post-ASA check-up in patients with residual **LVOTO ≥ 30 mmHg and < 30 mmHg**
(adjustment for age, sex, baseline LVOTO, and baseline septum thickness).

HR 2.95, 95% CI 1.26-6.91

Veselka et al. ESC congress 2016

Take-home messages

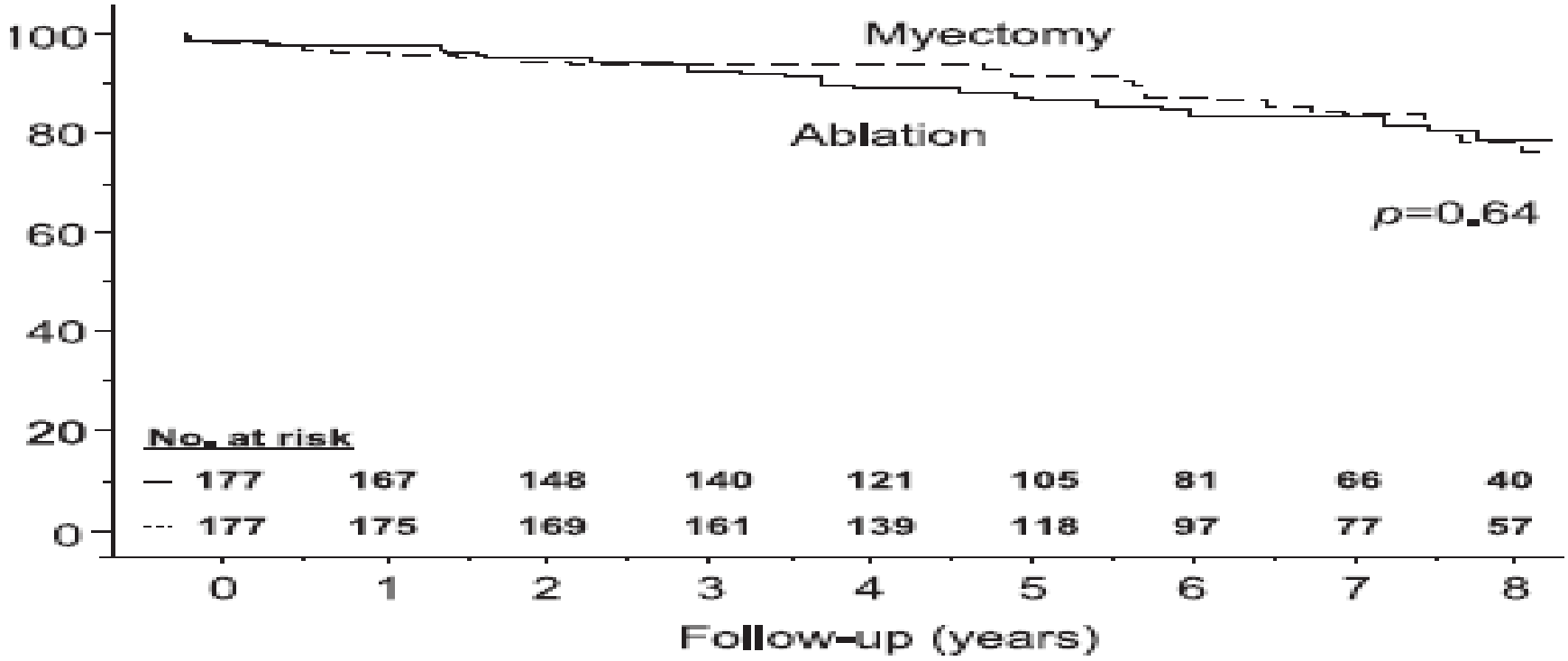
- LVOTO is a risk factor of long-term clinical outcome in HOCM patients treated or untreated with septal reduction therapy.

- Abolishment of LVOTO (it means PG < 30 mmHg) should be pursued in therapy of HOCM patients.

ASA vs Myectomy

Survival

ASA vs Myectomy: Mayo Clinic data



Sorajja P. Circulation 2012

ASA vs MYE

Meta-analyses

- A recent **meta-analysis** of long-term outcomes after septal reduction therapy, including **24 studies** from centers around the world, showed that the **peri-procedural mortality rate of ASA was 1.3%, compared to 2.5% in patients undergoing MYE**.
- When studies from before the year 2000 were excluded, these figures became similarly low (1.3% vs. 1.1%, respectively). The same held true for the long-term mortality rates.

Liebregts M et al.JACC Heart Fail 2015

Results of Ventricular Septal Myectomy and Hypertrophic Cardiomyopathy (from Nationwide Inpatient Sample [1998–2010])

Sidakpal S. Panaich, MD[a], Apurva O. Badheka, MD[a,*], Ankit Chothani, MD[b], Kathan Mehta, MD[c], Nileshkumar J. Patel, MD[d], Abhishek Deshmukh, MD[e], Vikas Singh, MD[f], Ghanshyambhai T. Savani, MD[f], Shilpkumar Arora, MD[a], Nilay Patel, MD[a], Vipulkumar Bhalara, MD[a], Peeyush Grover, MD[f], Neeraj Shah, MD[d], Mahir Elder, MD[a], Tamam Mohamad, MD[a], Amir Kaki, MD[a], Ashok Kondur, MD[a], Michael Brown, MD[a], Cindy Grines, MD[a], and Theodore Schreiber, MD[a]

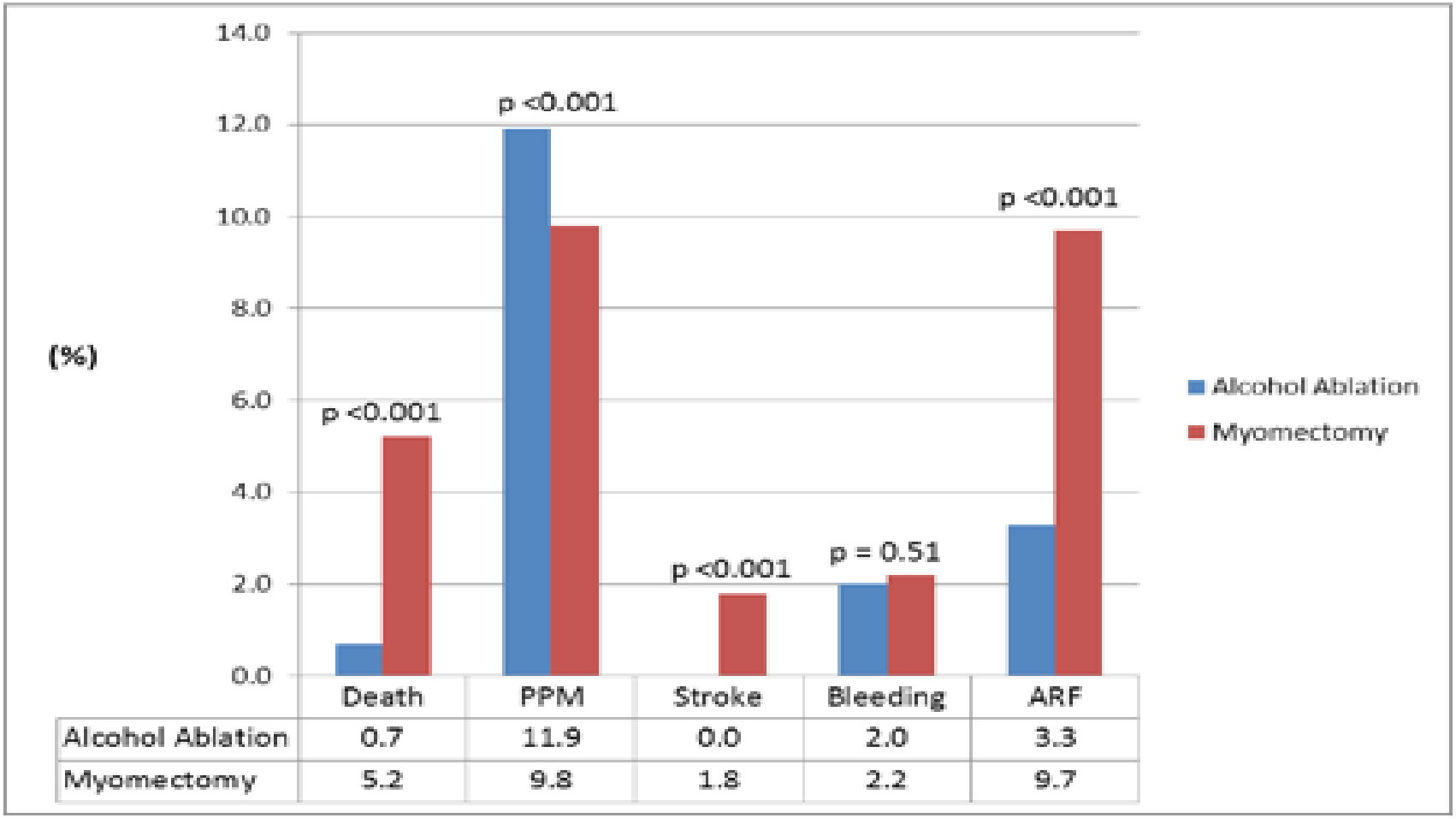
Table 1 shows baseline characteristics of the study population. A total of 665 VSM procedures were available for analysis from 1998 to 2010. The mean age of the study cohort was 56.9 ± 0.6 years. Men constituted 40% of the

The overall postprocedural mortality (Table 2) was 5.9%, while the rate of postprocedural complications was 30.2% (Table 2). Cardiac complications were most common (15.9%), including iatrogenic cardiac complications (10.5%) and complete heart block requiring pacemaker insertion (8.7%). Vascular complications, including access-site complications, occurred in 9.6% of patients, of whom 5.4% required transfusion. Respiratory complications occurred in 3.9% of patients, while 1.4% of patients had renal or metabolic complications.

Am J Cardiol 2014

Septal Myectomy is Associated With Worse In-Hospital Outcomes than Alcohol Septal Ablation: Data From the Nationwide Inpatient Sample in the United States, 2003-2011

Figure 1. Adverse In-Hospital Event Rates after Alcohol Septal Ablation and Septal Myectomy.



	Death	PPM	Stroke	Bleeding	ARF
Alcohol Ablation	0.7	11.9	0.0	2.0	3.3
Myomectomy	5.2	9.8	1.8	2.2	9.7

Looser PM et al. Circulation 2015

Low Operative Mortality Achieved With Surgical Septal Myectomy

Role of Dedicated Hypertrophic Cardiomyopathy Centers in the Management of Dynamic Subaortic Obstruction

*Barry J. Maron, MD
Joseph A. Dearani, MD
Steve R. Ommen, MD
Martin S. Maron, MD
Hartzell V. Schaff, MD
Rick A. Nishimura, MD
Anthony Ralph-Edwards, MD
Harry Rakowski, MD
Mark V. Sherrid, MD
Daniel G. Swistel, MD
Sandhya Balaram, MD
Hassan Rastegar, MD
Ethan J. Rowin, MD
Nicholas G. Smedira, MD
Bruce W. Lytle, MD
Milind Y. Desai, MD
Harry M. Lever, MD

Institution	No. of Myectomies	Age (yrs)	Male (%)	Operative Deaths†	
				n	%
Mayo Clinic, Rochester, Minnesota	1,411	51 ± 14	55	4‡	0.3
Cleveland Clinic, Cleveland, Ohio	1,470§	55 ± 14	55	6	0.4
Tufts Medical Center, Boston, Massachusetts‖	348	52 ± 15	56	4	1.1
Toronto General, Ontario, Canada	306	49 ± 13	62	2	0.6
Mount Sinai-St. Luke's and Roosevelt, New York, New York	160	53 ± 14	48	1	0.6
Totals	3,695	54 ± 14	55	17	0.4

Maron BJ et al. JACC 2015

Low procedure-related mortality achieved with alcohol septal ablation in European patients☆

Josef Veselka [a,*], Morten Kvistholm Jensen [b], Max Liebregts [c], Jaroslav Januska [d], Jan Krejci [e], Thomas Bartel [f], Maciej Dabrowski [g], Peter Riis Hansen [h], Henning Bundgaard [b], Robbert Steggerda [i], Lothar Faber [j]

Center	N	Male (%)	Age (years)	30-day cardiovascular mortality rate (%)
Bad Oyenhausen	249	53	59 ± 13	0.4
Nieuwegein/Groningen	193	53	61 ± 13	0.5
Prague	176	45	58 ± 12	0
Copenhagen/Rigshospitalet	104	43	62 ± 13	0
Trinec	84	37	60 ± 13	2.4
Brno	50	62	60 ± 12	0
Warsaw	50	50	56 ± 11	0
Innsbruck	31	52	57 ± 13	0
Copenhagen/Gentofte	25	56	62 ± 13	0

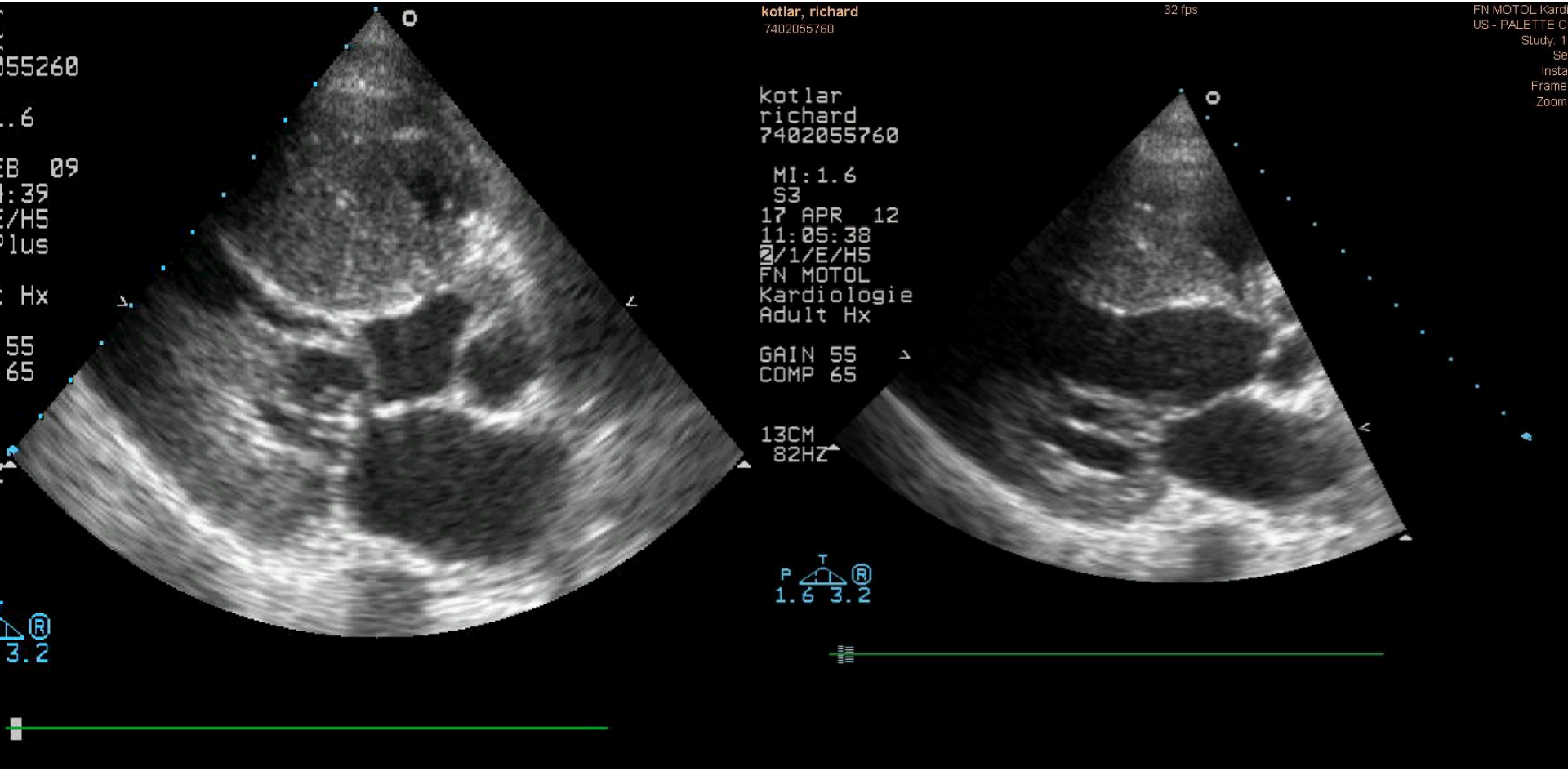
(Table 1). The 30-day procedure-related mortality rate was 0.4% identical to the five surgical centers reported by Maron et al. [4].

Int J Cardiol 2016

ASA vs MYE
General remarks

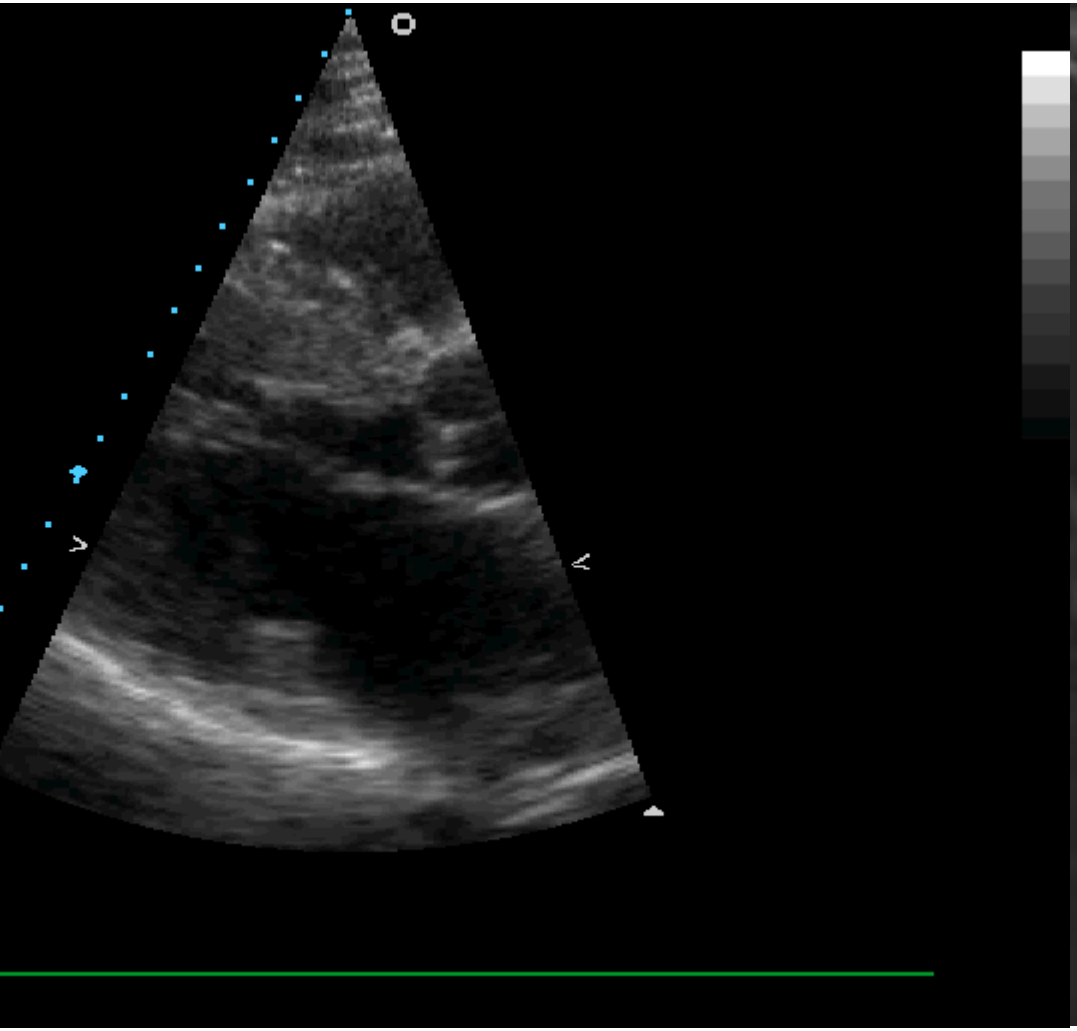
- Among the most important trends in current medicine are a **specialization of physicians and a centralization of medical care**. This trend deepens differences between results achieved in small- and high-volume centers.
- **The more complex the therapeutic goal, the bigger is the gap** between the safety and the efficacy of performed procedures in high- and low-volume centers.
- This fact again emphasizes the important caveat that **replication of excellent surgical results** achieved at the Mayo or Cleveland Clinic **is not possible at lower volume centers.**

Each patient needs optimal therapeutic option and experienced physician/center

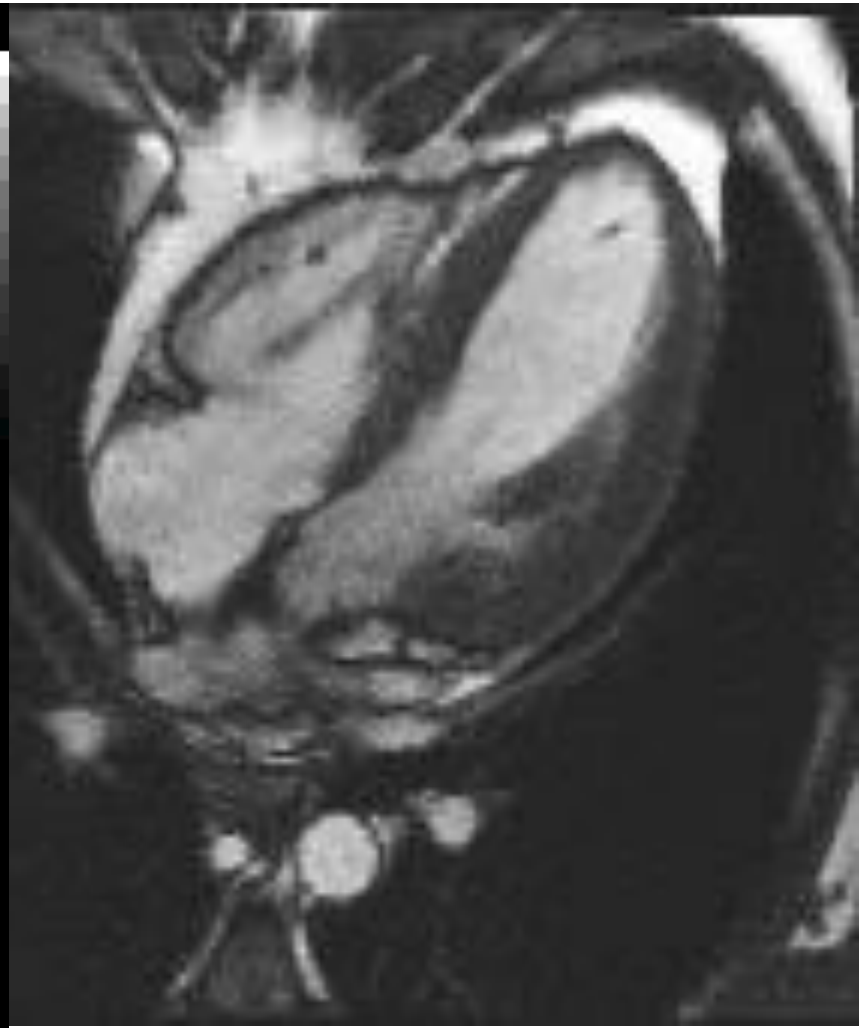


Morrow procedure

Each patient needs optimal therapeutic option and experienced physician/center



TTE: ASA



CMR: ASA

Therapeutic alternatives

Table. Lesion-Specific Treatment Options for Structural Abnormalities in Patients With Hypertrophic Cardiomyopathy and LV Outflow Tract Obstruction

Structural abnormality	Treatment options
LV hypertrophy	
Basal and asymmetrical septal	Transaortic myectomy
	Alcohol septal ablation
Midventricular	Transapical myectomy
Apical	Transapical myectomy
Mitral leaflet elongation	Mitral valve replacement
	Plication
	Other repair including Alfieri-type stitch
Papillary muscle adhesion, displacement and abnormal attachments	Division of papillary muscle
	Release of abnormal attachments

Song JK. Circ J 2014

Take-home messages

- **In dedicated centers are both ASA and MYE safe and effective.**
- We need more "real-world" data from less specialized hospitals (and should be taken into account when writing medical guidelines).
- **In smaller European countries and less developed health-care settings might be difficult to achieve a high-volume competence**
- We need tailored therapy for each patient.

- **<u>1% RULE: Procedure-related mortality rate in septal reduction therapy should not exceed 1%.</u>**