PRESENTER DISCLOSURE INFORMATION

Dr. Goldenberg Receives Research Grants from:

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PREVENTION OF SUDDEN CARDIAC DEATH IN PATIENTS WITH HYPERTROPHIC CARDIOMYOPATHY

Ilan Goldenberg, MD

Professor of Cardiology

Heart Center, Sheba Medical Center, TelHashomer, Israel and

University of Rochester Medical Center, Rochester NY, USA

OUTLINE

- Risk of SCD
- Risk factors for SCD in HCM
- Recommendations for the prevention of SCD
- Gaps and knowledge and limitations

BACKGROUND

- A minority of clinically recognized patients with HCM have increased risk for SCD (1% per year)
- ICDs offer the only effective means of preventing SCD in HCM patients

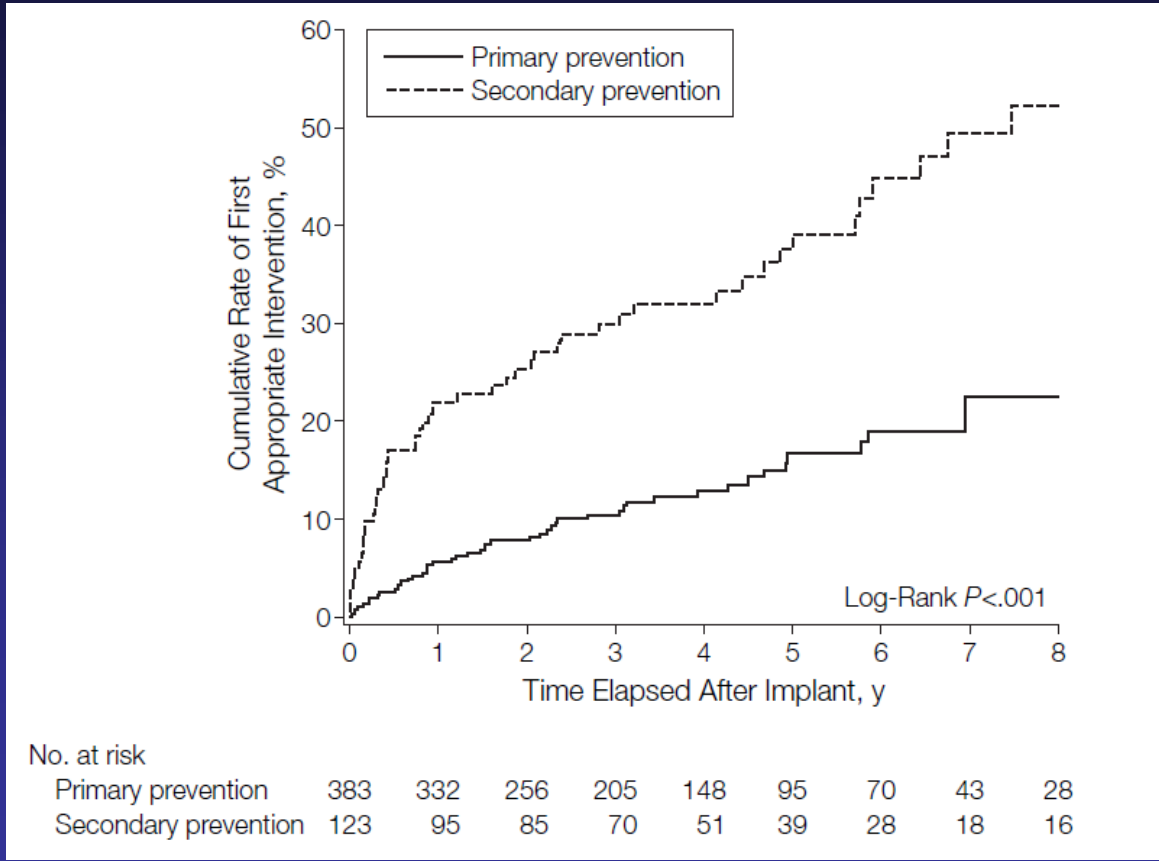
BACKGROUND

- Appropriate selection of primary prevention ICD therapy limited by:
  - Variable definitions for risk markers
  - Relative infrequency of HCM and SCD
  - Sparse clinical data
  - Morbidity associated with early ICD implant.
  - Use of surrogate markers for SCD
  - Do not consider reduced risk of SCD by age

RISK MARKERS FOR SCD IN HCM

SECONDARY PREVENTION

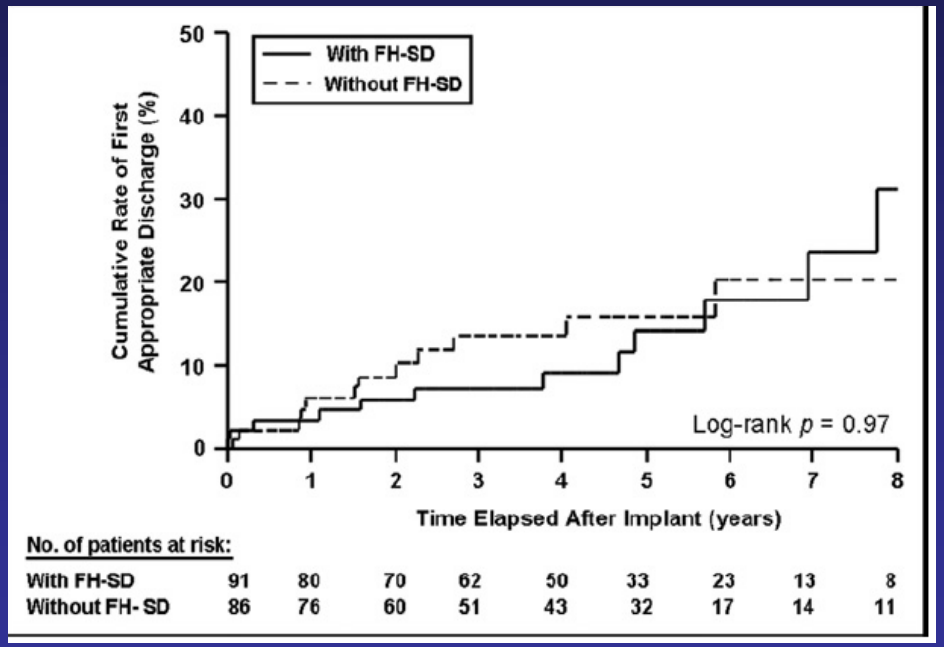
- History of:
  - VF
  - Sustained VT
  - SCD



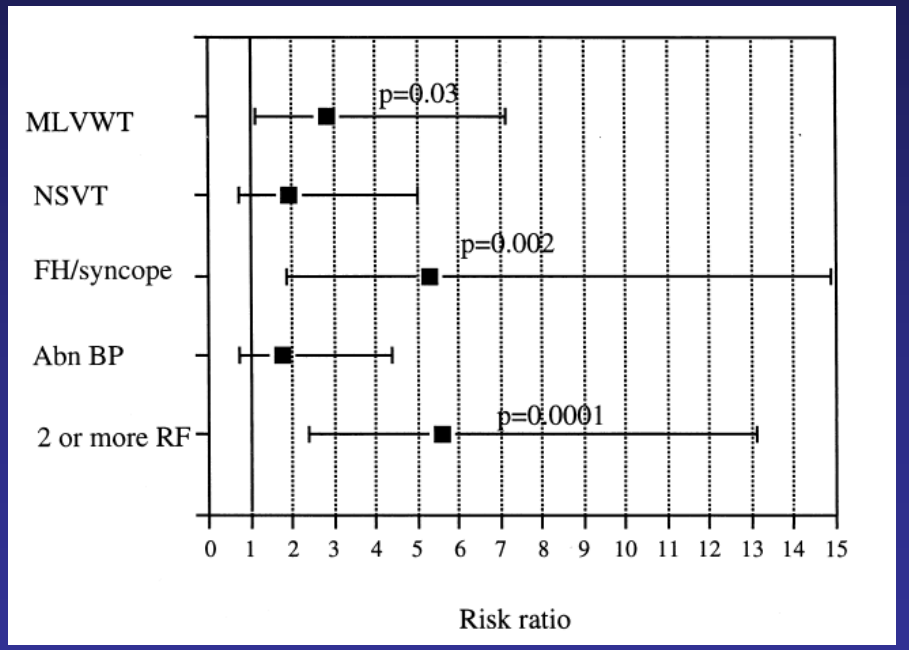
Maron et al. JAMA, 2007

FAMILY HISTORY OF SCD

Limited and conflicting data:



Bos JM, et al. AJC, 2010

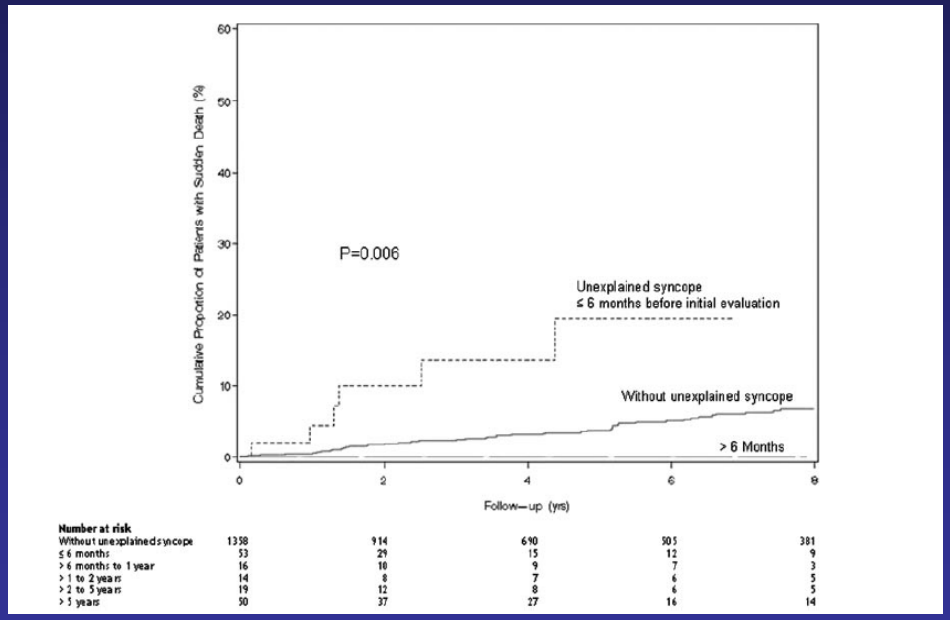


Elliott PM et al. JACC 2000

SYNCOPE

- Multifactorial etiology
- Requires a careful clinical history

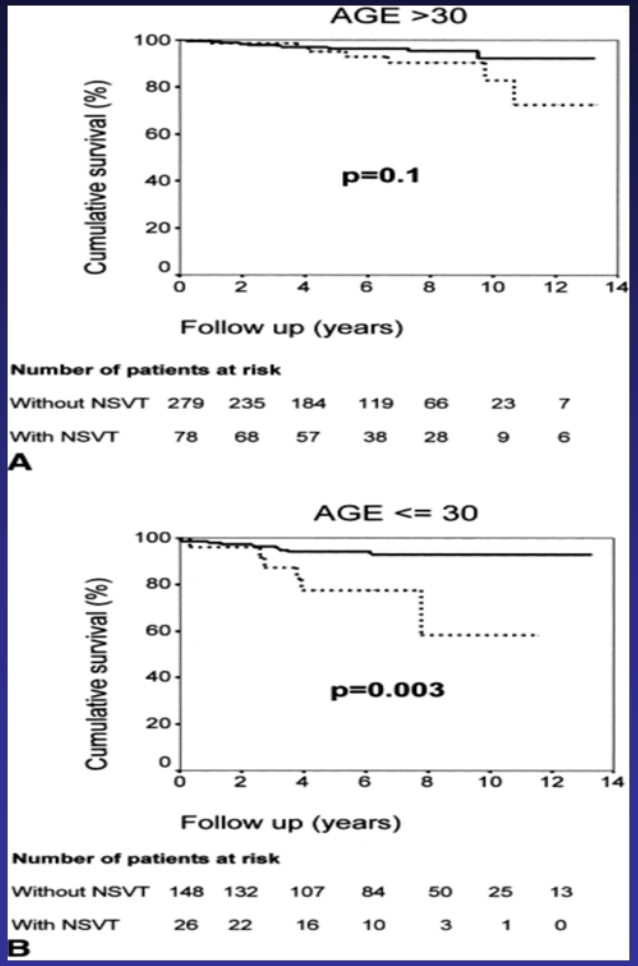
- *Predominant risk associated with recent unexplained syncope:*



Spirito, et al. Circulation, 2009

NONSUSTAINED VT

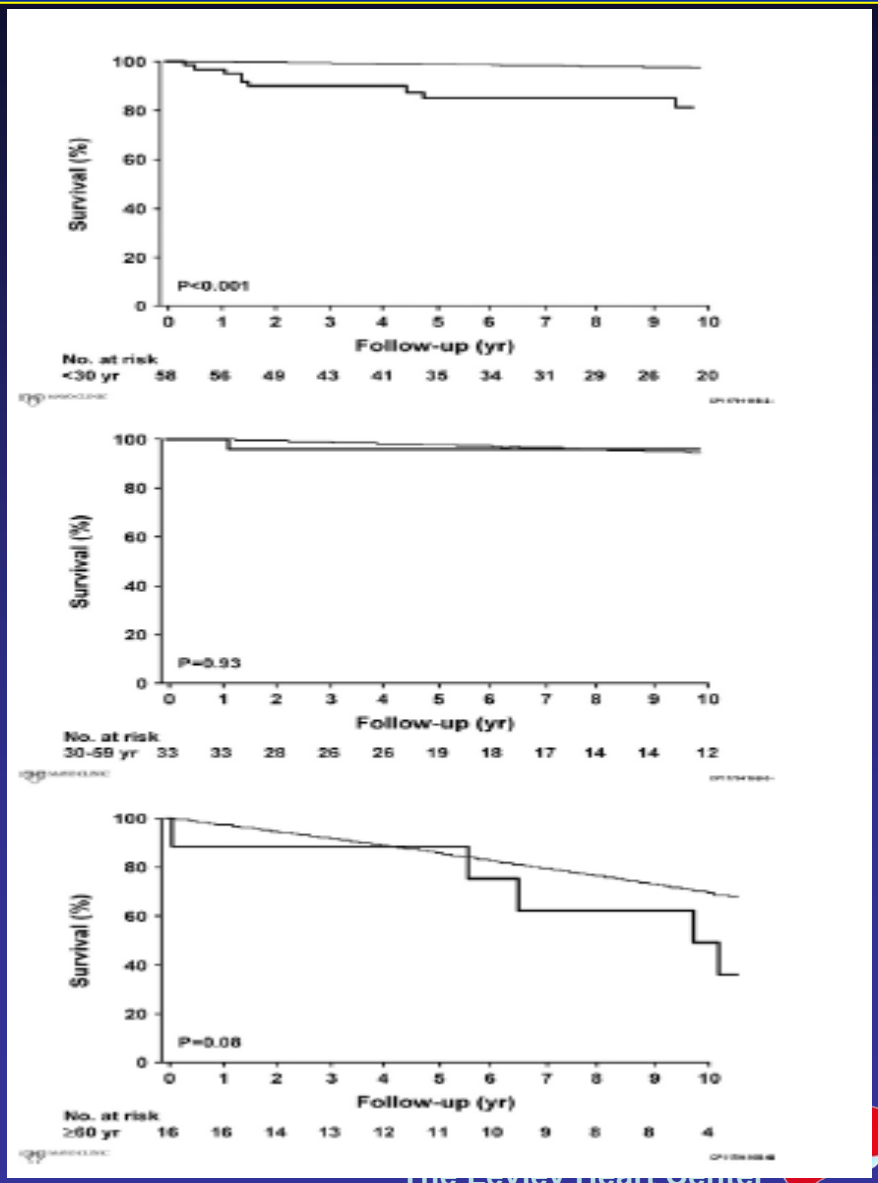
- Conflicting data from 5 studies
- One study showed age-dependent association with SCD



Monserrat et al. JACC, 2003

MAX. LV WALL THICKNESS

- Arbitrarily defined at ≥ 30 mm
- Risk increases in a linear fashion
- *Associated risk is age-dependent*



Sorajja et al. J Am Col Echo, 2006

Sheba Medical Center
Tel Hashomer

B.P. RESPONSE TO EXERCISE

- Defined as effort associated:
  - Failure to increase by at least 20 mm Hg
  - Drop of at least 20 mm Hg
- Occurs in up to a third of HCM pts

- Univariate association with SCD shown in several studies

ADDITIONAL POSSIBLE RISK MODIFIERS

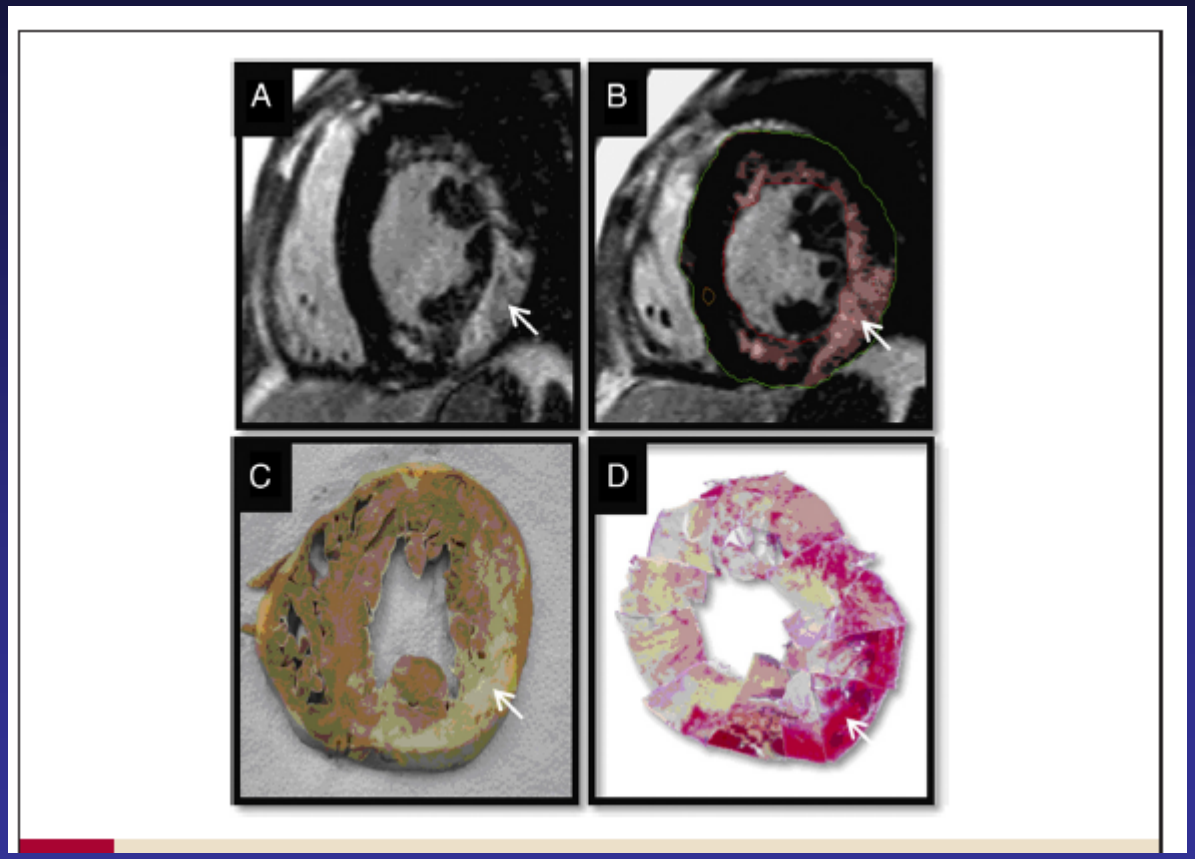
LVOT OBSTRUCTION

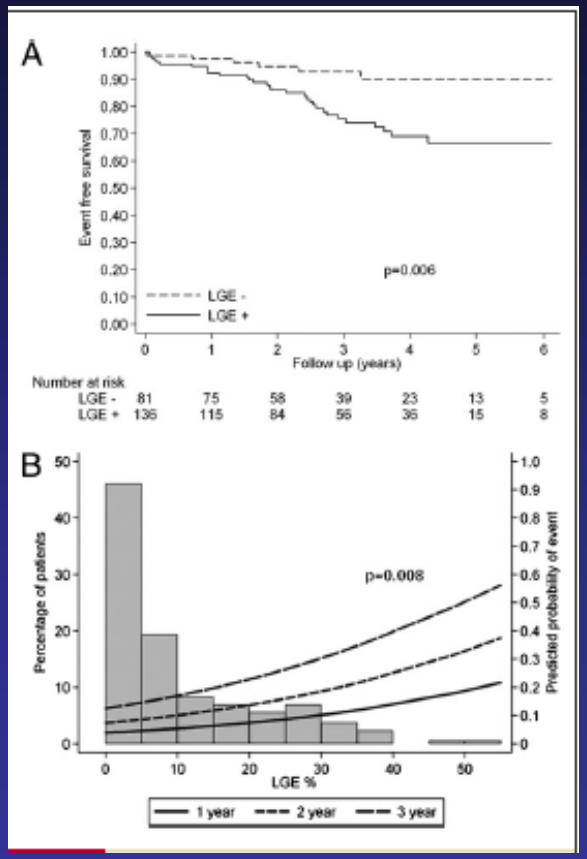
- ≥ 30 mm Hg obstruction suggested to be associated with increased risk for SCD
- Low risk of SCD following surgical myomectomy
- Conflicting data
- Dynamic obstruction limits this variable as a risk modifier

LGE-CMR

- Represents fibrosis or scarring
- Limited data suggest association with SCD
- Limitations:
  - Common in HCM pts
  - Lack of consensus on appropriate imaging protocols or threshold

LGE-CMR





O'Hanlon et al. JACC 2010

LV APICAL ANUERYSM

- 2% develop a thin-walled LV apical aneurysm associated with regional scarring
- Increased risk for adverse clinical events during follow-up:
  - Progressive heart failure
  - Possibly SCD

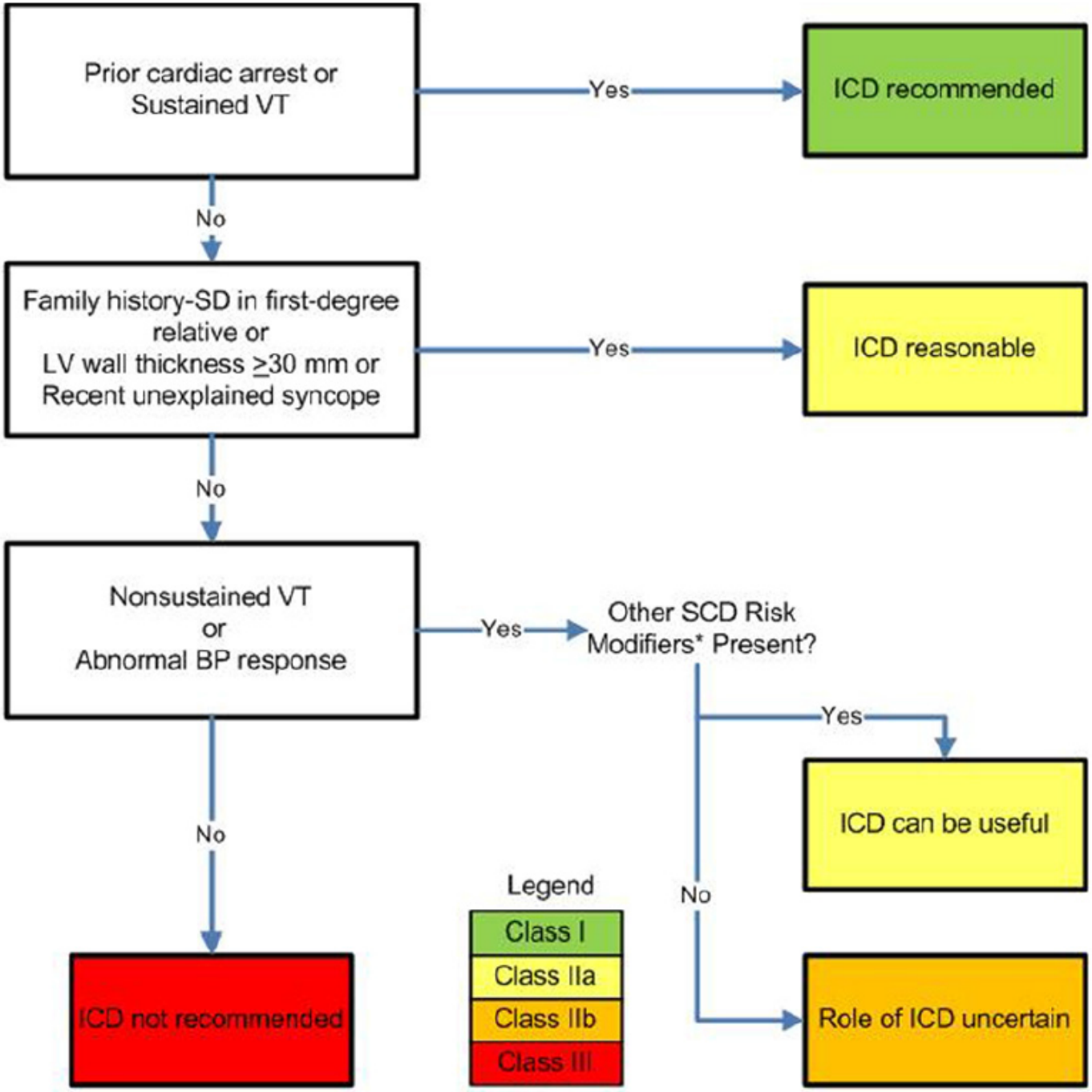
GENETIC MARKERS

- Currently routine mutational screening offers little prognostic information:
  - Some “malignant” mutations found to have a lower rate of clinical risk factors than “benign” mutations
  - Many are novel within specific families

SUMMARY: RISK FACTORS

- Low positive predictive value (10% - 20%)
- High negative predictive value (85% - 95%)
- The majority of pts with ≥ 1 risk factors will not experience SCD
- Cumulative number of risk factors not shown to correlate with risk

2011 ACCF/AHA GUIDELINE FOR THE DIAGNOSIS AND TREATMENT OF HYPERTROPHIC CARDIOMYOPATHY


Prior cardiac arrest or Sustained VT
Yes
ICD recommended
No
Family history-SD in first-degree relative or
LV wall thickness ≥30 mm or
Recent unexplained syncope
Yes
ICD reasonable
No
Nonsustained VT
or
Abnormal BP response
Yes
Other SCD Risk Modifiers* Present?
Yes
ICD can be useful
No
Role of ICD uncertain
No
ICD not recommended
Legend
Class I
Class IIa
Class IIb
Class III

CONCLUSIONS

- Current data even regarding established (guidelines-based) risk markers for SCD in HCM are limited and conflicting
- Decision needs to be individualized:
  - Age
  - Strength of the risk factor
  - Risk-benefit of lifelong ICD therapy
- There is a need for a contemporary risk stratification approach in HCM

THANK YOU