Cardiovascular **Magnetic Resonance** in Hypertrophic Cardiomyopathy Martin S. Maron, MD, FACC, FHRS **Medical Director** Hypertrophic Cardiomyopathy Center **Tufts Medical Center** Boston, MA Chanin T. Mast HCM Center

Tufts Medical Center SEPTAL ABLATION SEPTAL MORRISTOWN, NJ ENT GENETICS ALCOHOL SEPTAL ABLATION SEPTAL MYECTOMY MULTI IOMYOPATHY MEDICAL MANAGEMENT GENETICS ALCOHOL SEPTAL AB APPROACH HYPERTROPHIC CARDIOMYOPATHY MEDICAL MANAGEMENT

Morristown Medical Center



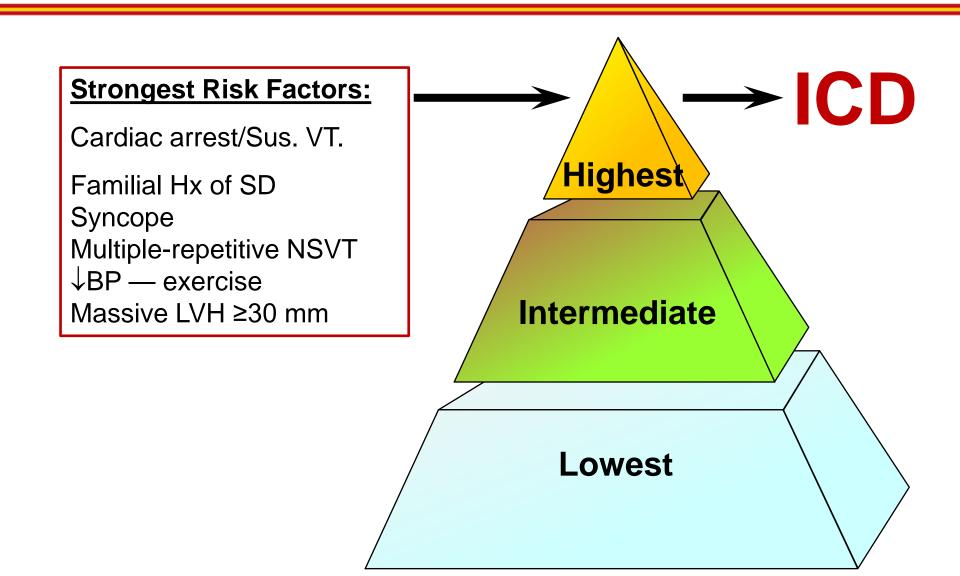
DISCLOSURE

• Gadolinium is FDA off-label use for CV imaging

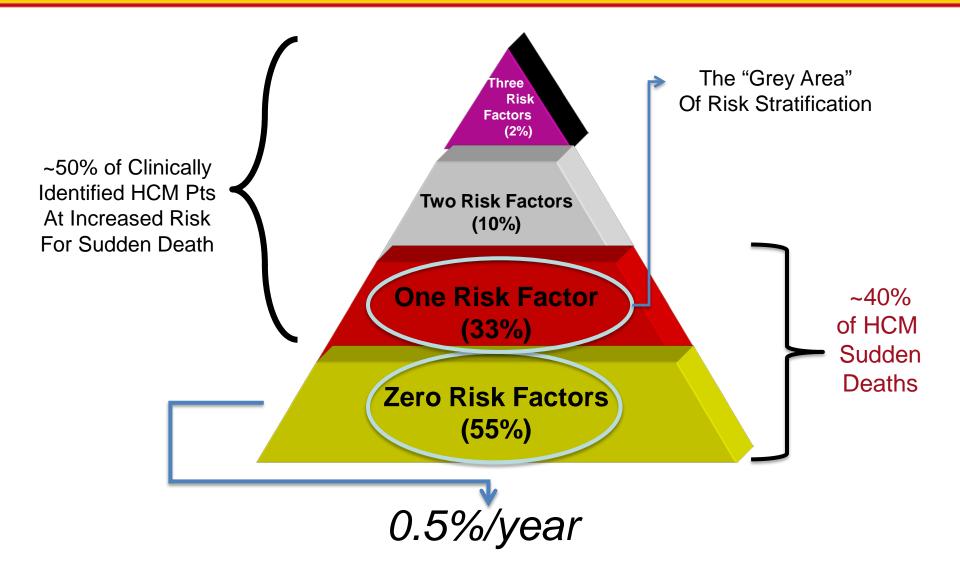


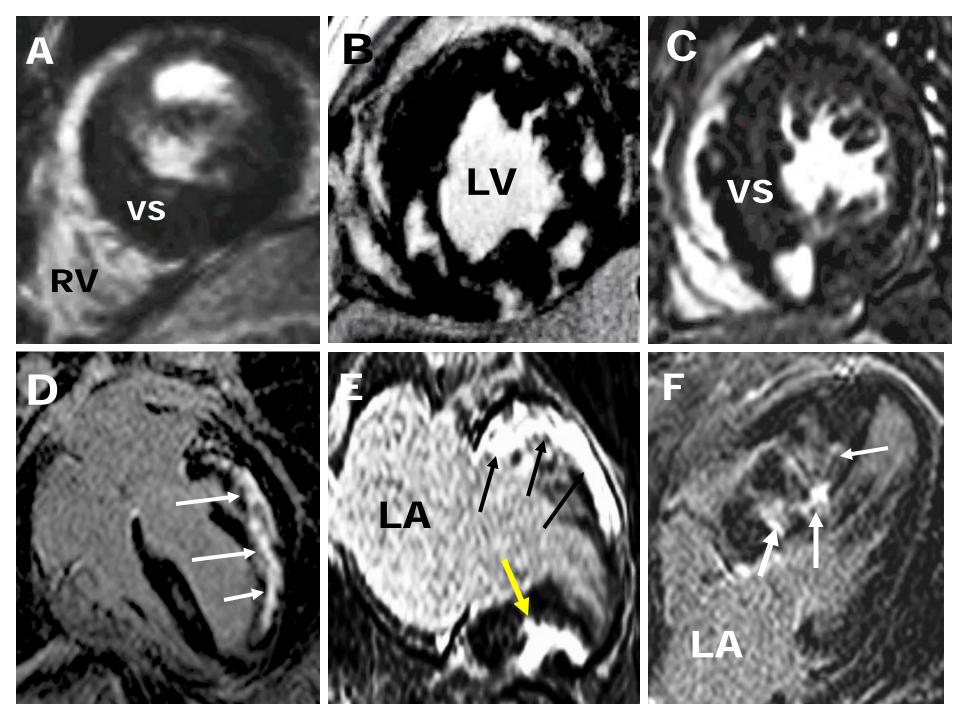


Strongest Risk Factors

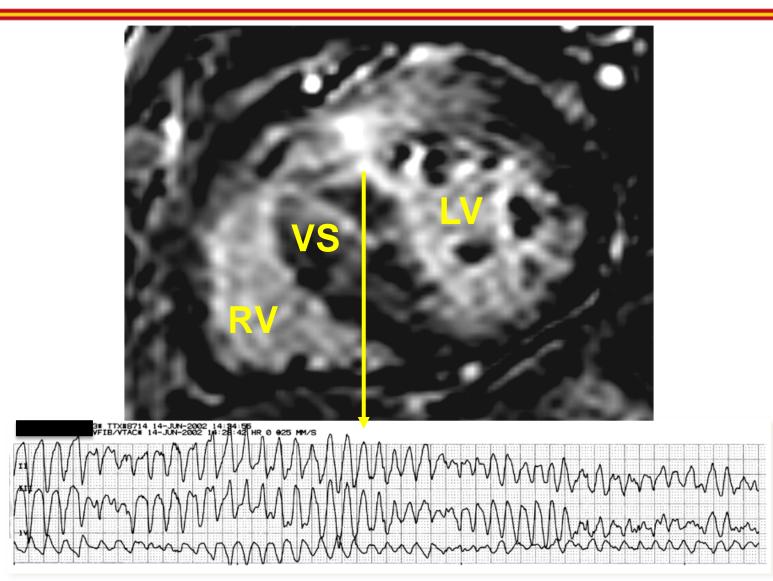


Challenges of Risk Stratification in HCM

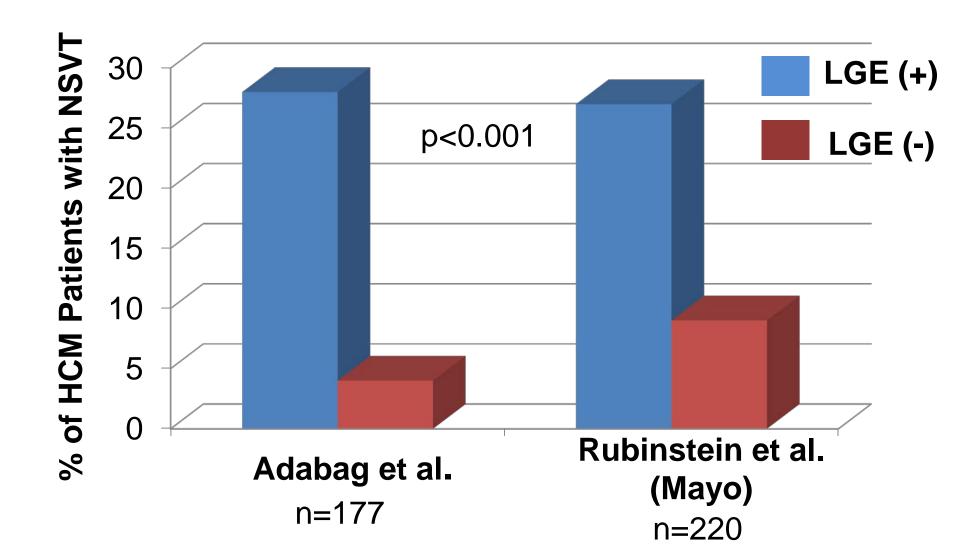




Foci for Ventricular Arrhythmias?



Holter NSVT and Presence of LGE

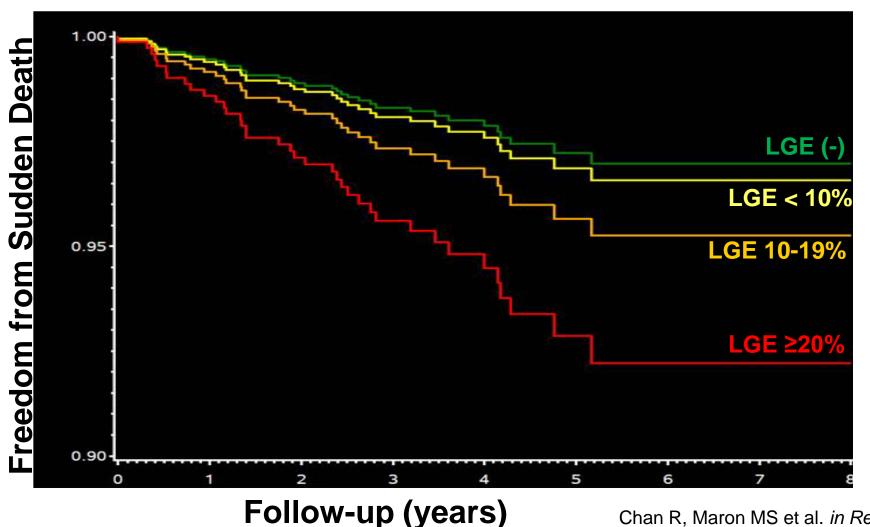


LGE for Prognosis in HCM Multicenter Study

- Tufts Medical Center, Boston, MA
- Minneapolis Heart Institute
- Toronto General Hospital, Canada
- Azzendia Carregia, Florence, Italy
- Bologna, Italy
- Pisa, Italy
- Rome, Italy
- **PERFUSE CMR Core Laboratory**



Relation Between Sudden Death and Extent of LGE in 1293 HCM Patients



Chan R, Maron MS et al. in Review

Sudden Death Event Rates in HCM Patients without Conventional Risk Factors

% LGE	Adjusted HR	Est. 5-year Event Rate(%)
0%	1.0	2.5
5%	1.3	3.2
10%	1.6	4.0
15%	2.0	5.0
20%	2.6	6.3
25%	3.2	8.0
30%	4.2	10.0
40%	6.7	15.5

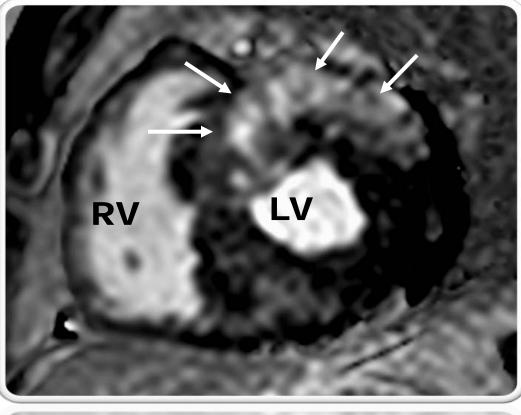
Chan R, Maron MS et al. in Review

Improvement of Sudden Death Prediction with Addition of %LGE to Risk Model

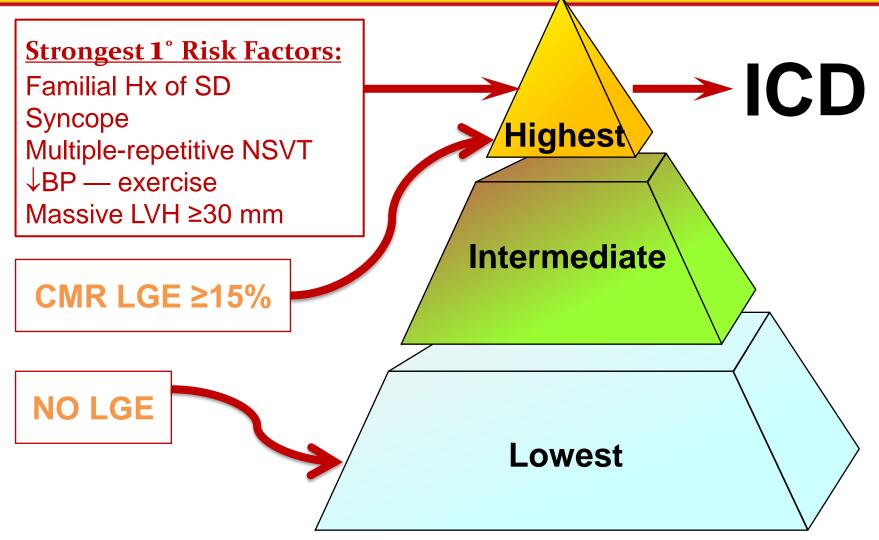


When Extensive LGE Counts

32 year-old man with Sudden Death Asymptomatic (Max LV WT = 26 mm) Normal Ejection Fraction No Traditional Risk Factors RF LGE = 22% of the LV Mass



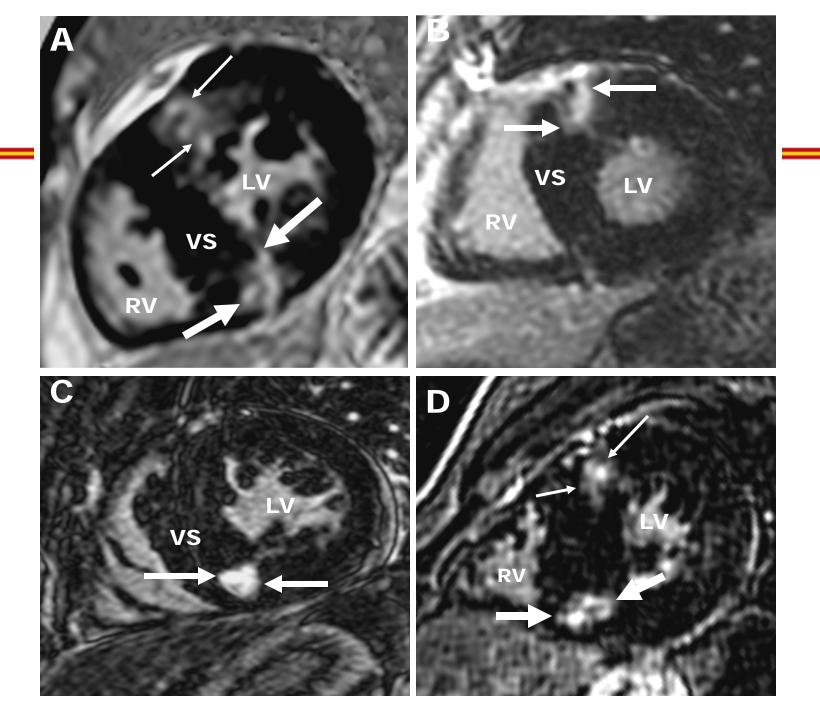




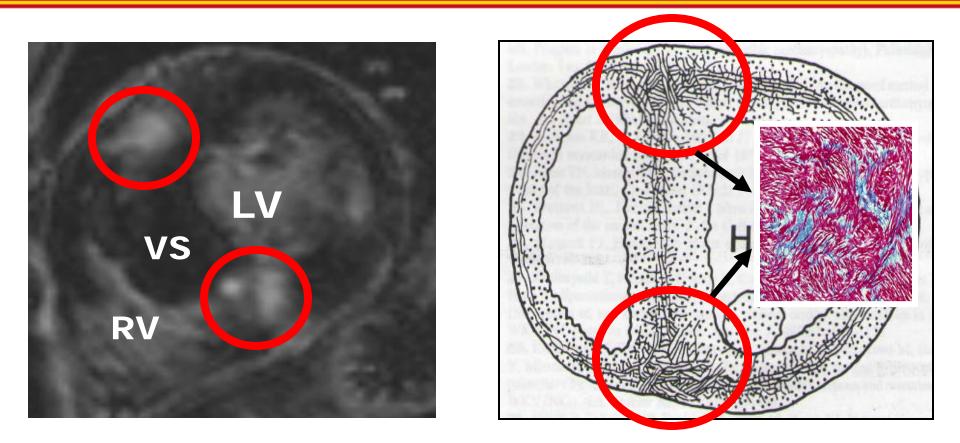
Chan R, Maron MS et al. in Review

Risk Factors

Risk of SD is about the Amount of LGE...<u>Not</u> its Pattern or Location

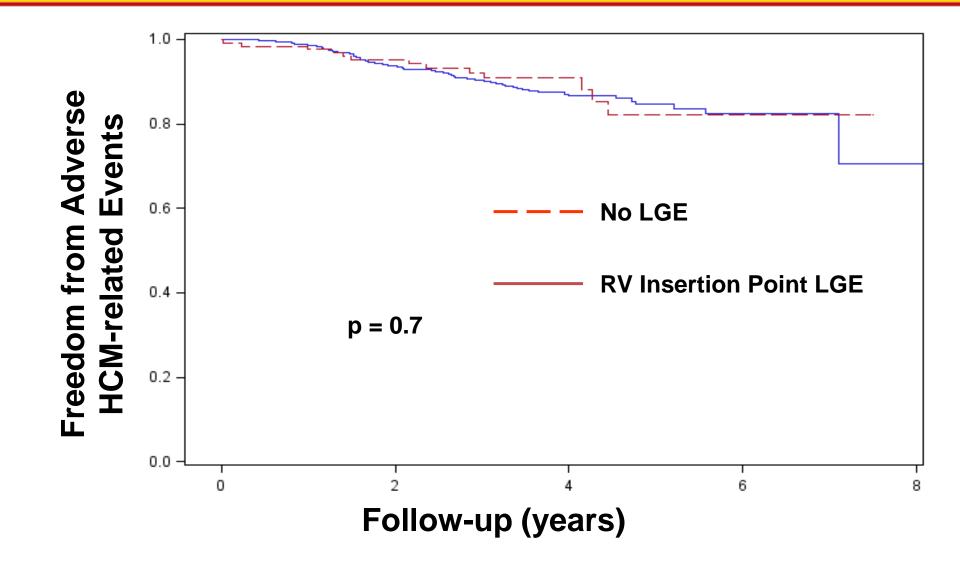


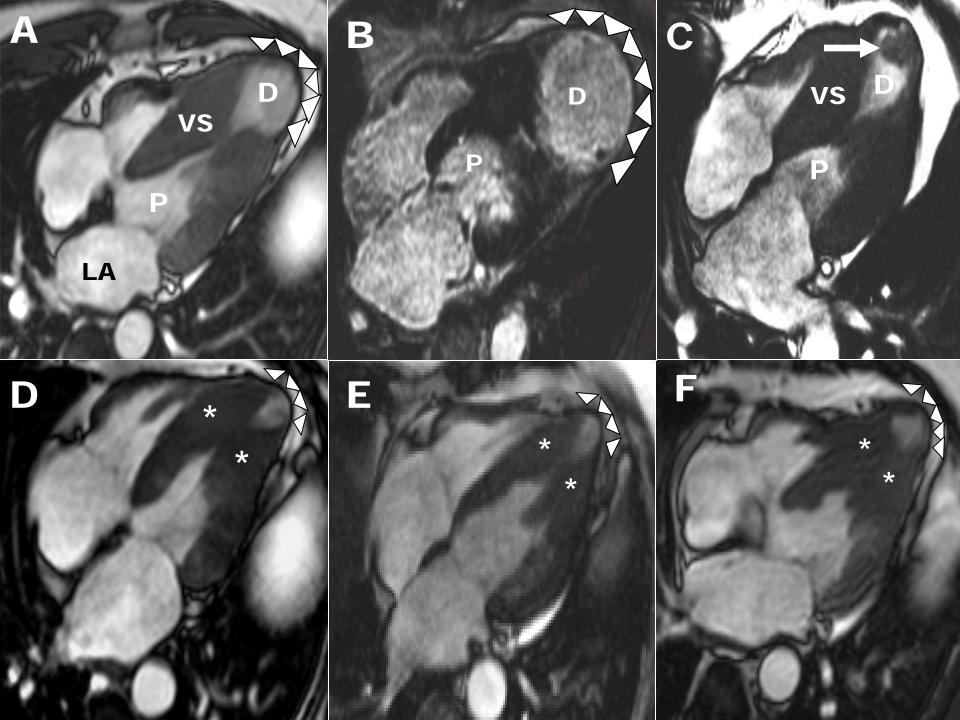
RV Insertion Point LGE in HCM = Expanded Extracellular space



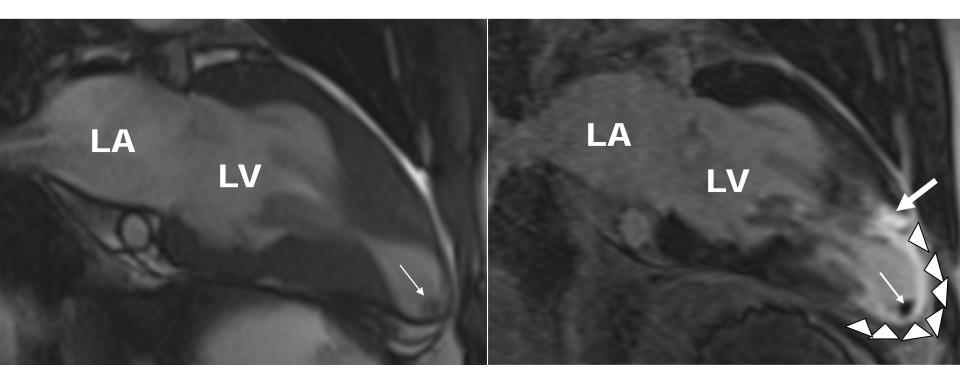
Not Likely Replacement Fibrosis

Sudden Death Risk in Patients with RV Insertion Point LGE Only



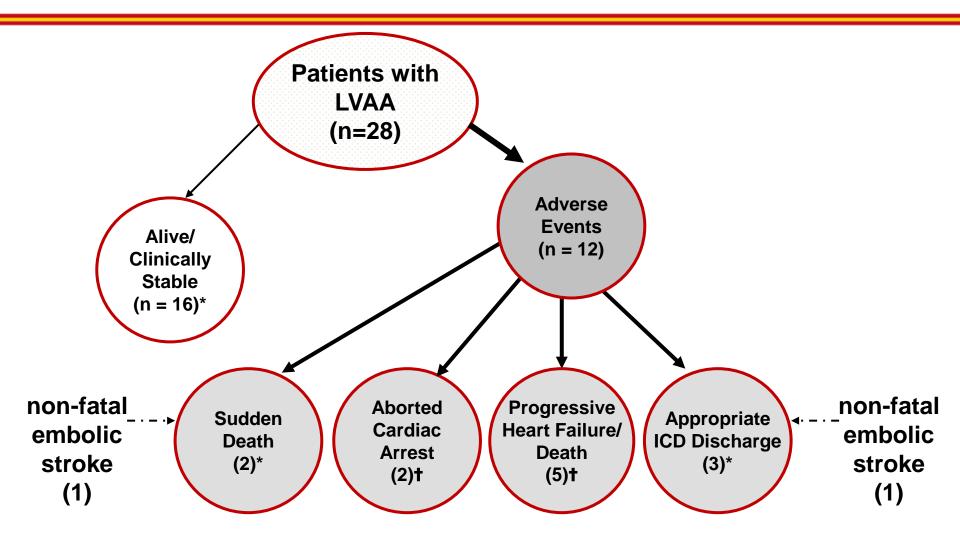


LV Apical Aneurysm with Thrombus



Maron MS et al. Circulation 2009

Cardiovascular Event Rate = 11%/yr



Maron MS et al. Circulation 2009

CMR in HCM

- Particularly well suited to characterize the diverse phenotypic expression of this complex disease...
- Superior to echo for HCM diagnosis...ie., anterolateral wall, apex
- Management strategies for invasive septal reduction therapy...ie., mitral valve, anomalous insertion of papillary muscles
- Sudden Death Risk Prediction...Extensive LGE (≥15% of LV) identifying a novel subgroup of HCM patients at increased risk for SD that would not be considered without CMR and may now be candidates for ICD
- Absence of LGE associated with low risk....may serve to influence decision-making against ICD implants in "grey-zone" situations

Heart & Vascular Center



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