**A SYSTEMATIC REVIEW FOR SUDDEN CARDIAC DEATH IN HYPERTROPHIC CARDIOMYOPATHY PATIENTS WITH MYOCARDIAL FIBROSIS: A CMR LGE STUDY**

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Background: Hypertrophic cardiomyopathy (HCM) is a genetic disease that affects the cardiac sarcomere, resulting in myocardial hypertrophy and disarray. Affected patients have a predisposition for malignant ventricular tachyarrhythmias and, consequently, sudden cardiac death (SCD). In single center studies, late gadolinium enhancement (LGE) defined fibrosis has been linked to the substrate for VT/VF. However, despite innumerable investigations, SCD has not been definitely attributable to LGE. Explanations for this are believed to be related to insufficient statistical power.

Methods: We performed an electronic search of MEDLINE, PubMED and CMR abstracts published between Jan01-Mar11. Key search terms: HCM, LV fibrosis, SCD and LGE. Studies were screened for eligibility based on inclusion criteria: referral for CMR exam with LGE for HCM; and follow-up for incidence of VT/VF and SCD. Relevant studies were summarized. The following data were abstracted: socio-demographic information; study design; incidence of reported VT/VF; SCD. Categorical variables were evaluated between pt groups via Chi-square test.

Results: A total of 64 studies were initially identified. Of these, 4(6.3%) were identified and included (n=1,063 pts, range 202-24) Three prospective and 1 retrospective studies were included. LGE was detected in 59.6% of pts.

Mean follow-up was 43±14mo. Age 22-90yrs (63%male). As expected, the presence of myocardial fibrosis was associated with VT/VF (χ2=6.5, p<0.05; OR 9.0(95% CI 1.2-8.7). Moreover, myocardial fibrosis strongly predicted SCD (χ2=6.6,p<0.05;OR .3

(95% CI 1.2-.7).

Conclusions: Despite single center CMR studies, LGE has consistently predicted VT/VF while prediction of SCD has remained paradoxically unlinked. Although the lack of studies meeting our criteria limited our ability to perform a comprehensive meta-analysis, we have been able to demonstrate for the first time that LGE-defined fibrosis is a predictor of SCD in patients with HCM. This observation now demands a multi-center RCT for confirmation but supports the consideration of a primary indication of AICD implantation in HCM with LGE.