

Effect of hypokinetic transformation on the clinical phenotype and functional capacity in hypertrophic cardiomyopathy

Y. Wasserstrum¹, E. Itelman¹, R. Barriaes-Villa², X. Fernandez-Fernandez², Y. Adler¹, D. Lotan¹, R. Klempfner¹, N. Shlomo¹, A. Sabbag¹, D. Freimark¹, L. Monserrat², M. Arad¹

¹Sheba Medical Center, Ramat Gan, Israel; ²University Hospital Complex A Coruña, A Coruña, Spain

Funding Acknowledgement: Type of funding source: None

Background: Advanced hypertrophic cardiomyopathy (HCM) may be complicated by a dilated hypokinetic transformation. Reduced left ventricular ejection fraction (HFrEF) has been described in terms of specific risks of morbidity and mortality, and specifically in terms of increased risk for fatal arrhythmias. Nevertheless, recent publications have casted doubt regarding the role of arrhythmia in non-ischemic HFrEF and questioned the role of primary prevention strategies in these cases.

Methods: We've reviewed clinical characteristics of 883 patients age ≥ 40 , diagnosed with HCM who were evaluated in the cardiomyopathy clinic in two tertiary medical centers in Israel and Spain.

Results: Forty-five patients (5%) suffered from hypokinetic transformation. They were younger at diagnosis (median 32 [IQR 24–55] vs. 49 [35–60], $p < 0.001$), had a lower body-mass index ($28.4 [\pm 4.7]$ vs. $26.0 [\pm 3.9]$, $p < 0.001$), and suffered more from strokes (19% vs 6%, $p < 0.001$). They had lower had a lower NYHA class ($p = 0.001$) and lower exercise capacity

($7.3 [4.5–10.8]$ vs. $9.6 [6.7–12.0]$ METS, $p < 0.001$). Patients with hypokinetic HCM had higher rates of pacemaker and implanted defibrillator (ICD) implantations (41% vs 11%, $p < 0.001$) and (43% vs 13%, $p < 0.001$) respectively. These patients had a higher incidence of sustained ventricular tachyarrhythmias (14% vs 2%, $p < 0.001$). Among patients who had an ICD, patients suffering from hypokinetic transformation had received more appropriate ICD therapy (27% vs 12%, $p < 0.001$). These patients received more heart transplantations (13% vs 1%, $p < 0.001$), and had a trend for higher incidence rate of Sudden cardiac death (6% vs 2% $p = 0.06$) and a higher 5-year mortality rates (21% vs. 5%, $p < 0.001$).

Conclusions: HCM patients suffering from hypokinetic transformation have lower functional and exercise capacities, are more likely to suffer from ventricular tachyarrhythmias and experience appropriate ICD therapy, and undergo heart transplantation. They also have a significantly lower 5-year survival.

