Commentary: Pulmonary Hypertension and Survival in Hypertrophic Cardiomyopathy: A Predictor or a Surrogate?

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Word Count: 454/500

Conflict of Interest: Author S.H. is a consultant for Encare EIAS system. There are no other conflict of interest related to this manuscript

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Central Picture Legend: Sameer A. Hirji MD (left), Borami Shin (middle), Sary Aranki MD (right)

Central Message: Pulmonary hypertension is present in almost half of the patients with hypertrophic cardiomyopathy, and is associated with worse survival.

Management of hypertrophic cardiomyopathy (HCM) has evolved over the last several decades aided by refinements in risk stratification tools that tailor treatment strategies. The innovations in surgical techniques provide durable benefit, and improvements in medical therapies help to minimize overall morbidity. (1) In appropriately selected patients, surgical (septal) myectomy has become an effective, low-risk operation with anatomical (based on hemodynamic parameters) and clinical benefits (based on patient symptoms). Nonetheless, several key questions still remain unanswered including the optimal timing of surgery for severe symptoms despite optimal medical therapy (Class I guidelines), or those with severe pulmonary hypertension (PH), left atrial dilatation with associated risk of atrial fibrillation, severe mitral regurgitation, or severe heart failure (Class IIb guidelines). (2)(3)

We read with great interest the paper by Ahmed et al.(4), which attempts to answer the above question and raise awareness of the prognostic value of preoperative PH in the context of optimal timing of intervention and long-term survival. To their credit, the authors utilized their extensive 30-year surgical experience with HCM to determine the prevalence and impact of PH in patients undergoing surgical myectomy. They found that PH was present in almost half of the
patients, and those with at least mild PH had worse survival compared to those without PH. The correlation between PH and mortality seems valid, and despite the underlying patient selection biases (in terms of those who undergo surgery), these findings provide some food for thought: First, PH appears to be a predictor of worse survival, and second, surgical myectomy may potentially provide greater benefit to patients if offered earlier in their clinical course rather than wait till progression of symptoms as highlighted in the guidelines. This aggressive approach may halt the irreversible pulmonary endothelial remodeling that may be associated with long-term right ventricular dysfunction and worse survival. (5)

Given the high incidence of myocardial fibrosis and sudden cardiac death in this patient cohort (6), it remains paramount that clinicians perform systematic, thorough and nuanced investigations to provide optimal treatment in the current era of precision medicine. While PH appears to be an important additional variable to assist with patient selection, it remains to be determined whether PH is a consequence of HCM or a surrogate for worsening cardiac dysfunction. This study doesn’t delineate between the two but perhaps using either a contrast-enhanced cardiovascular magnetic resonance imaging with late gadolinium enhancement (LGE) or myocardial biopsy may have provided further guidance in determining ideal candidates and timing for surgical myectomy (7) in the absence of large scale data or randomized clinical trials.

References


