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Left atrial size is an important predictor of morbidity in patients with latent obliterative hypertrophic cardiomyopathy.

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Background: Subaortic obstruction in HCM may be classified as obstruction at rest or latent (provokable). Although echo characteristics of hypertrophic cardiomyopathy (HCM) and latent obstruction (LO) have been studied, there is limited information on long-term morbidity in patients presenting with LO.

The aim of this study was to analyze predictors of morbidity in patients with LOHCM following in a tertiary referral center.

Methods: A retrospective study of 125 patients (73% men) with LOHCM diagnosed from 1975 to 2002 was performed. Inclusion criteria were: unexplained left ventricular hypertrophy with no significant outflow gradient (LVOTGR) at rest, increasing to >30 mmHg by pharmacological provocation, documented by echo (n=65) or cardiac catheterization (n=60). Symptoms, clinical findings, mortality and cardiovascular morbidity were analyzed.

Results: The mean age at presentation was 45.2 ± 16.1 years. At baseline the mean LVOTGR at rest was 7 ± 8 mmHg and 65 ± 25 mmHg after provocation, the mean left atrial diameter was 40 ± 6 mm, the mean septal thickness 18.6 ± 4.2 with hypertrophy limited to the basal 1/3 of septum in 71 patients (57%), and to the proximal 2/3 in 30%. Morbid events occurred in 59 of 127 patients consistent with cardiovascular morbidity of 46.5%. The probability of event-free survival for patients with LOHCM was 51±6% at 15 years of follow-up. Sixteen patients (13%) had one or more morbid events at the initial presentation, most frequent event being AF (n=13), CHF (n=3), MI (n=2) and/or cerebrovascular event (n=2). Two independent predictors of all cardiovascular morbidity were identified by a multivariate Cox regression analysis: left atrial enlargement at baseline HR 2.2 (95% CI 1.3 – 3.4), and a higher age at diagnosis HR 1.03 (95% CI 1.001 – 1.044).

Conclusion: The majority of patients with LO have less extensive hypertrophy and a more favorable prognosis than other types of HCM. However, in the presence of left atrial enlargement and older age at presentation LO has significant cardiovascular morbidity and mortality.

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Evaluation of subendocardial ischaemia by strain Doppler echocardiography in patients with left ventricular outflow tract obstruction.

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Background: The purpose of the present study was to assess the subendocardial wall function using tissue Doppler imaging (TDI) and strain rate imaging (SRI) in patients with congenital left ventricular outflow (LVOT) obstruction.

Methods: We studied with TDI and SRI 19 pts aged 11-31 years with congenital left ventricular outflow (LVOT) obstruction.

Results: Tissue Doppler imaging and strain rate echocardiography provide a unique insight in diastolic and systolic function. The systolic strain was reduced in both groups but myocardial strain rate was significantly lower in patients with LOHCM. This finding may explain the higher prevalence of atrial fibrillation in patients with LOHCM.

Conclusions: The presence of atrial fibrillation in patients with LOHCM may be attributed to impaired myocardial function.

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Mid-systolic septal deceleration - a new sign of left ventricular outflow tract obstruction obtained by colour-coded tissue Doppler echocardiography.

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Diagnosis of hypertrophic obstructive cardiomyopathy (HOCM) is based on the identification of asymmetric septal hypertrophy, mitral leaflet systolic anterior motion (SAM) and a systolic LVOT gradient. Many patients present with no significant resting gradient, but develop significant obstruction after provocation manoeuvres or inducedextrasystoles. Doppler evaluation of the LVOT gradient during such manoeuvres remains technically challenging, in particular in difficult cases with suboptimal Doppler angles and concomitant mitral regurgitation. We describe a new non-invasive sign for LVOT obstruction obtained by colour-coded tissue Doppler echocardiography (TDI).

Clinical Case: Septal longitudinal motion was studied by TDI in a 69-year old HOCM patient during transcoronary alcohol ablation of septal hypertrophy (TASH). Invasive hemodynamics showed only a small LVOT gradient at rest (Figure,left), but a significant post-extrasystolic LVOT gradient, which was associated with an abrupt mid-systolic deceleration (MSSD) pattern in the simultaneously acquired basal septal TDI velocity trace (Figure,middle). Immediately after TASH, both the LVOT gradient and the simultaneously recorded MSSD pattern were significantly reduced (Figure,right). A similar MSSD pattern was observed in 5 additional consecutive HOCM patients with severe LVOT obstruction, but in none of 10 patients with pure aortic valve stenosis.

Conclusions: The presence of an abrupt mid-systolic septal deceleration pattern in the TDI velocity trace is associated with severe, dynamic LVOT obstruction. It may constitute a new diagnostic tool for gradient characterisation and may help to monitor HOCM therapy.