The mitral valve in hypertrophic cardiomyopathy – an echocardiographic approach

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In the recent past, the mitral valve was referred to as the cornerstone of any echocardiographic examination. Changes in its echocardiographic pattern were considered, in many cases, to be specific for the diseases in which they were described. Among such diseases was hypertrophic cardiomyopathy.

Three abnormalities were successively described in the M-mode echocardiograms of patients with the obstructive form of hypertrophic cardiomyopathy. The first was a prolonged mitral valve-septum contact, with a slow diastolic closing slope – the E-F slope, whether measured along the first (E-FO) or the second (FO-F) components (Edler, 1967). The second abnormality was a systolic anterior movement attributed to the anterior leaflet. The third was a thickening of the septum, out of proportion compared with the opposite posterior wall – asymmetric septal hypertrophy.

Both the echocardiographic mitral valve abnormalities and asymmetric septal hypertrophy were initially considered to be specific for the disease but later this was questioned. In fact the diagnostic value of echocardiographic mitral valve abnormalities in the spectrum of hypertrophic cardiomyopathy was greatly reduced when it was recognized that even in their absence the study of left ventricular walls allowed identification of many cases of hypertrophic cardiomyopathy, generally with asymmetrical septal hypertrophy, but occasionally with concentric hypertrophy. Furthermore, systolic anterior motion was described, though rarely, in conditions other than hypertrophic cardiomyopathy (Maron & Epstein, 1980).

On the other hand, it was also recognized that asymmetric septal hypertrophy could occur in other entities and that mitral valve abnormalities could be present in the absence of asymmetric septal hypertrophy or even left ventricular hypertrophy detectable by M-mode echocardiography (Mintz et al., 1978). When examining 1009 consecutive adult patients, we found that among 84 (8.3%) having asymmetric septal hypertrophy, only 30 (2.9%) had hypertrophic cardiomyopathy; 2 patients, in whom the only echocardiographic abnormalities present were mitral valve abnormalities, had familial hypertrophic cardiomyopathy (Madeira et al., 1978).

Despite obvious limitations, changes in the mitral valve echocardiographic pattern, namely systolic anterior motion, are highly specific. The prevalence of true systolic anterior motion is very low in the general population and among cardiac patients without hypertrophic cardiomyopathy (Maron et al., 1981). Therefore it retains an important diagnostic value, though only for the obstructive form of the disease.

Apart from its usefulness in diagnosing hypertrophic (obstructive) cardiomyopathy, the study of echocardiographic mitral valve abnormalities – both systolic and diastolic – permits the prediction of a gradient and to a certain extent the type of distribution and degree of septal hypertrophy. There is now substantial data supporting the concept that narrowing of the left ventricular outflow tract, caused by septal thickening and by the anteriorly displaced mitral valve, is the basis for the dynamic obstruction leading to systolic anterior motion, due to the Venturi effect (Gilbert et al., 1980). Our data, presented in Figure 1, from a study of 38 patients with hypertrophic cardiomyopathy – 12 with resting obstruction, 13 with latent obstruction and 13 with no obstruction – shows that patients with resting obstruction are those who have a thicker septum, in whom the mitral valve is significantly more anteriorly placed and who have a larger atrium, possibly indicating the presence of mitral regurgitation.

Further indirect evidence that narrowing of the outflow tract by the septum interferes with the normal dynamics of the mitral valve can also be found from the analysis of its echocardiographic diastolic pattern (Figure 2). Whereas in non-obstructive hypertrophic cardiomyopathy the diastolic closing slope (E-F) is an almost uniphasic line (see Figure 2, top left), in the obstructive form there is a prolongation of E-FO time (see Figure 2, top right and bottom). The E-FO time is a substantial part of the rapid diastolic filling period (Kalmanson et al., 1975; Vignola et al., 1977), and echocardiographically it represents the ring movement (Zaky et al., 1968), after which the valve semi-closes due to vortices formed between the anterior leaflet and
the septum (Madeira et al., 1974). It seems reasonable to assume that narrowing of the left ventricular outflow tract (partly due to septal thickening) interferes with the formation of vortices thus delaying diastolic closing of the mitral valve and therefore prolonging E-FO time. Accordingly we found a positive correlation ($r = 0.80$) between septal thickness and E-FO time – the thicker the septum, the longer this rapid diastolic filling period, judged by the mitral echogram.

Two-dimensional data are in accordance with these assumptions. The analysis of septal thickness (long-axis plane) in our 38 patients has shown that there is a variability in the distribution of septal hypertrophy (Figure 3), which may predominate in the upper septum (basal dominant), in the lower septum (distal dominant), be equally distributed, or sieged at the apical zone. Of our 12 patients with resting obstruction, 8 had basal dominant hypertrophy, 4 had equally distributed hypertrophy and none had distal dominant hypertrophy; of the 13 patients with latent obstruction, 4 had basal dominant hypertrophy, 4 had equally distributed hypertrophy and 5 distal dominant hypertrophy; finally, of the 13 patients without obstruction, none had basal dominant hypertrophy, 4 had equally distributed hypertrophy, 7 had distal dominant hypertrophy.
Figure 2  The mitral valve echogram in hypertrophic cardiomyopathy, non-obstructive (top left) and obstructive (top right). Notice (bottom) the prolonged E-FO time. MVO = mitral valve opening.

trophy and 2 apical hypertrophy. Therefore, none of the patients with no obstruction had basal dominant septal hypertrophy and none of the patients with resting obstruction had distal dominant hypertrophy.

In conclusion, echocardiographic abnormalities of the mitral valve play an important role in establishing the diagnosis of hypertrophic cardiomyopathy by separating obstructive from non-obstructive cases and in predicting the outflow gradient. It seems from the analysis of diastolic abnormalities of the mitral valve echogram, combined with the study of septal hypertrophy by M-mode and two-dimensional methods, that basal septal hypertrophy, narrowing the left ventricular outflow tract, plays a decisive role in the genesis of obstruction.
Figure 3 Two dimensional (long-axis plane) echocardiograms of 3 relatives with hypertrophic cardiomyopathy. a – basal dominant septal hypertrophy; b – distal dominant septal hypertrophy; c and d – diastolic and systolic frames from a case of apical hypertrophy.

References


