

SURGICAL CORRECTION OF OBSTRUCTIVE FAMILIAL HYPERTROPHIC CARDIOMYOPATHY.

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Background: HOCM is genetically and clinically heterogeneous myocardial disease. An important clinical predictor of premature death is malignant family history. We studied a severe case of HOCM by searching for mutations in β -cardiac myosin heavy chain gene (MYH7) as a candidate. A missense mutation in codon 606 of MYH7 was identified as cause in a nuclear family with a malignant history (mother and son were affected). Surgical correction of LV obstruction (LVOT and mid-cavity) was approached by myectomy from the right side of the IVS. This is an alternative to the Morrow technique which does not allow to perform left-ventricular resection of mid-ventricular parts of the septum in HOCM patients with severe hypertrophy.

Methods: We present a method of excising severely hypertrophied IVS tissue causing obstruction of LVOT and the left mid-ventricular cavity by applying right- rather than left-ventricular myectomy. Access to the septum was through longitudinal incision of the RV conal part. Myectomy was performed by partial removal of hypertrophic tissue anterior to the Lancisi muscle. Both seriously affected carriers of the codon 606 mutation in MYH7 were treated this way.

Results: Follow-up studies in these 2 patients were done for 17 (mother) and 12 (son) months. The mean echocardiographic intraventricular gradient in the left ventricles after surgery decreased from 81/95 to 7/12 mmHg (mother and son, resp.). Septal thickness (by echocardiography) was reduced from 31/33 to 16/18 mm (mother and son, resp.). Follow-up echocardiography showed further a marked reduction of left atrial size. An increase in LV and RV filling fraction was shown by magnetic resonance imaging. Both patients were in sinus rhythm.

Conclusion: RV myectomy of the IVS is an effective technique for surgical treatment of familial HOCM in cases of massive left ventricular hypertrophy which includes obstruction of the mid-cavity in addition to the outflow tract.