Septal Myectomy and Myotomy Attenuate Left Ventricular Hyper-Contractility in a Child with Hypertrophic Obstructive Cardiomyopathy

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Introduction

Hypertrophic obstructive cardiomyopathy (HOCM) in children is a rare disease which can be treated surgically by - septal myectomy/myotomy. It has been recognized as an effective method in relieving left ventricular outflow tract (LVOT) obstruction. However, we hypothesized that the effect of septal myectomy/myotomy is not achieved so much through the structural widening of the LVOT as through the reduction of septal hyper-contractility. In order to test this hypothesis, echocardiographic Vector Velocity Imaging (VVI), using a speckle tracking method, was used in a pediatric HOCM surgical case.

Case Presentation

A 7-year-old boy was referred to our hospital for surgical treatment. He was diagnosed with severe HOCM with asymmetric septal hypertrophy (ASH) when he was 2 months old. Restriction of daily activity and full medication failed to improve his NYHA functional status III, as demonstrated by maximally recorded BNP of 2984 pg/ml (at age 5 years), necessitating surgical intervention.

Laboratory blood and urine data showed no abnormalities except a high BNP value of 1780 pg/ml. His chest x-ray demonstrated cardiomegaly with a 58% cardiothoracic ratio. His electrocardiogram demonstrated left
ventricular (LV) hypertrophy with strain-type ST segment depression. His echocardiogram demonstrated LV diastolic dimension (LVDd), 24 mm; LV systolic dimension (LVDs), 13 mm; LV ejection fraction (LVEF), 85.5%; interventricular septal wall thickness (IVSt), 29 mm; posterior wall thickness (PWt), 13 mm; peak LVOT pressure gradient (PG), 124 mmHg; mild grade 1 aortic valve regurgitation (AR); moderate grade 2 mitral valve regurgitation (MR); and typical systolic anterior movement (SAM) of the anterior leaflet. Contrast-enhanced magnetic resonance imaging (MRI) with gadolinium confirmed chronic myocardial enlargement (Fig. 1a and 1b).

The heart and ascending aorta were exposed via median sternotomy. Cardiopulmonary bypass was established in the standard fashion, the proximal aorta was crossclamped, and cardiac arrest was induced with antegrade infused modified St. Thomas solution. A proximal aortotomy was made. A specially designed scalpel composed of a no.11 surgical blade tip vertically held with a needle holder was used for septal muscle columnar resection (15 mm length, 15 mm depth, 50 mm height) from 10 mm below the right coronary cusp attachment to near the apex. The surgeon’s index finger was used to perform additional blunt crushing of the resected portion to disrupt the muscle bundle continuity and lessen the contractile force.

Intraoperative transesophageal echocardiography using a 7 MHz transducer (V5M, Acuson Sequoia c512, Siemens Medical Solutions USA Inc., Mountain View, CA) was performed. Peak LVOT PG dramatically dropped from a preoperative value of 180 mmHg to 7.6 mmHg after cessation of cardiopulmonary bypass. Postoperative VVI analysis was performed (Syngo software, Siemens Medical Solutions USA Inc., Mountain View, CA). The pre and postoperative calculated results were maximum twist, 17.9° and 7.9° (Fig. 2a); and minimum untwist, 75.2°/sec and 87.3°/sec (Fig. 2b).

His postoperative course was uneventful. Postoperative transthoracic echocardiography demonstrated IVSt, 29 mm; PWt, 10 mm; LVDd, 26 mm; LVDs, 19 mm; LVEF, 53.0%; peak LVOT PG, 20 mmHg; SAM; and trivial MR. Postoperative MRI demonstrated reduced LVOT muscle mass. He was discharged on postoperative day 10.

Pathology of the resected septal muscle demonstrated disarrayed cardiac muscle fibers, interstitial vacuolization, and interstitial fibrosis, findings compatible with the diagnosis of HOCM.

Discussion

The incidence of pediatric HOCM is relatively rare, with a reported frequency of approximately 0.47 per 100000 children, compared to a relatively high incidence of non-obstructive hypertrophic cardiomyopathy (HCM) (approximately 2 in 1000 young adults). Although the clinical and hemodynamic spectra of ASH in childhood are broad, deterioration in clinical condition or sudden death is reported to be relatively common once overt typical clinical symptoms appear. Symptomatic HOCM patients refractory to medical treatment can be treated effectively with septal myectomy, and postoperative long-term survival is considerably improved compared to the natural history of this disease.
In patients with HOCM, severe asymmetric septal hypertrophy not only causes moderate narrowing of the LVOT but also induces significant septal hyper-contraction. The latter causes LV posterolateral cardiac and papillary muscles to move upward (i.e., toward LVOT), resulting in SAM of the mitral leaflet, and thus making the LVOT narrow further in systole and causing valvular regurgitation.

The central technique of the Morrow operation (conventional septal myectomy) was dilating the LVOT by partially removing the septal muscle. We believe attenuation of hypertrophied septal muscular contractile power is at least equally, if not more, important. This concept led to our new surgical technique of not only cutting but also crushing the septal musculature by using the surgeon’s index finger. We used VVI analysis to evaluate perioperative LV contractility. The method is useful for evaluating LV twisting and untwisting, which are thought to represent cardiac systolic and diastolic function, respectively. LV twist was calculated by determining the difference between apical and basal rotation angles. As the angle of LV twist increases, LV contractility also increases. LV untwist was calculated as rotation angle/second. It may be a useful diastolic function marker or may even serve as a therapeutic target for improving diastolic function that can be impaired in hypertension and LV hypertrophy.

The data acquired in the present case showed that maximum LV twist decreased from 17.9°, preoperatively, to 7.9°, postoperatively, demonstrating that myotomy/myectomy using our resection/crush method rendered the excessive septal contractile power to nearly normal (cf. normal twist: 7.1 ± 3.8°). This coincided with the global LVEF reduction from 85.5% to 53.0%. However, the change in LV untwist (75.2°/sec to 87.3°/sec) was not significant (normal untwist value: 59.4 ± 23.4°/sec), demonstrating that the procedure did not affect diastolic function. Postoperative mitral valve SAM and MR almost disappeared, which we believe is also a consequence of the attenuated contractile power of the hypertrophied muscle.

VVI analysis showed the extent of myocardial contractile power and the effectiveness of the myectomy/myotomy. VVI analysis was shown to be valuable, and continuing use of it is warranted.
Disclosure Statement

The authors report no conflicts of interest in the writing of this paper.

References