

Surgical relief of left ventricular outflow obstruction in pediatric hypertrophic cardiomyopathy: The need for a tailored approach



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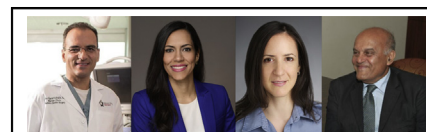
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Central Message

A personalized tailored approach to the operative procedure of relief of LVOT obstruction using sophisticated tools and preoperative imaging may be key to a successful operation.

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Hypertrophic cardiomyopathy (HCM) is extremely heterogeneous with regard to both genetic origins and phenotypic expressions.^{1,2} The age of onset of the disease has been recently identified as a major determinant of prognosis, phenotype, and association with syndromic features. This has led to the recognition of pediatric HCM as a specific disease entity¹ requiring specific management. Left ventricular outflow tract (LVOT) obstruction is a defining feature,³ which when severe requires surgical myectomy usually through a transaortic approach. In contrast to adults, the performance of this operation in children is more challenging and requires the use of specific approaches.

The article by the Paris group in this issue of the *Journal*⁴ is a welcome addition to the literature because it outlines some of these challenges and reports the long-term results of the use of the modified Konno operation in 79 children with the most severe form of HCM varying in age between 1 month and 18 years. The overall results are excellent except in infants with Noonan's syndrome with biventricular obstruction. Perhaps delaying the operation to a minimum age could improve the results. The authors conclude that the modified Konno technique should be considered an alternative approach to transaortic septal myectomy in high-risk patients with extensive hypertrophy and associated right heart obstruction, and in younger patients with a small aortic orifice. A similar surgical approach to HCM septal myectomy through the right ventricle has been recently described for simultaneous relief of right and left ventricular outflow obstruction without perforating the septum⁵ in a tailored technique.

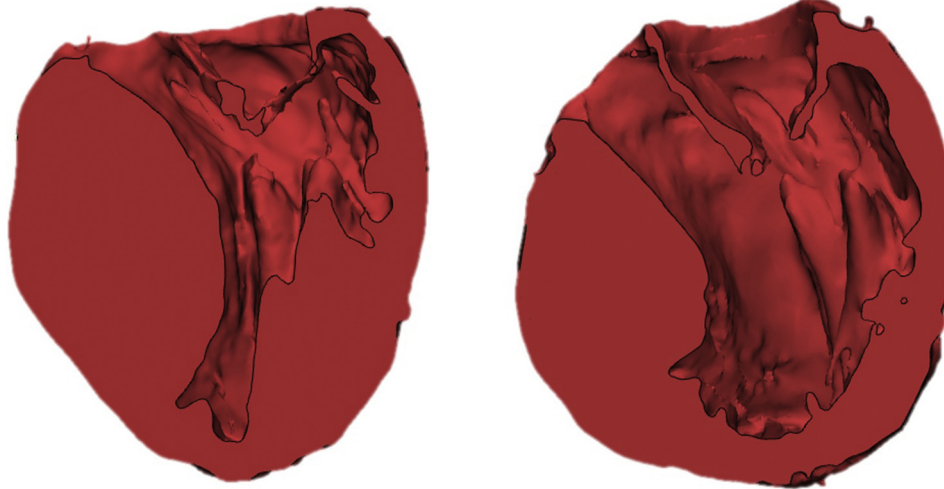
The LVOT performs extremely sophisticated functions, which depend on its complex geometry and dynamism.⁶⁻⁸

Changes in the structure and function of the LVOT in HCM are a dynamic process involving most if not all of its component parts, can affect other parts of the left and right ventricle, and are more severe in pediatric patients. The nature and degree of the changes need to be defined preoperatively to enable a personalized surgical approach to restore the sophisticated functions of the LVOT without injuring the surrounding vital structures. The procedure should be tailored to deal with all the varied components of the obstruction, including fibrous trigones, accessory tissues, and papillary muscle fusion. Planning the personalized operation requires a series of tests. Preoperative multimodality imaging, 3-dimensional modeling (Figure 1), and intraoperative transesophageal echocardiography are essential to define the specific abnormalities to be dealt with and assess the repair. The use of currently available and emerging imaging modalities, such as diffusion tensor magnetic resonance imaging,⁹ could be applied to define the deranged pattern of myocardial trabeculae and possibly fiber orientation, in individual patients, to further enhance the precision of the operation. In addition, computer simulation with 3-dimensional modeling of the ventricle coupled with application of virtual operations with more detailed analysis of the possible effects on the hemodynamics¹⁰ and the kinetics of the myocardium may be of value in further refinement of the operation.¹¹ The development of instruments to enhance visibility and precision inside the ventricular cavity could be of assistance during operation.

Left ventricular wall structure

Pre-operative

Post-operative



3D anatomical model reconstructed for HOCM patient pre and post operative from End-diastolic phase CT images

FIGURE 1. Preoperative and postoperative 3-dimensional segmentation of the left ventricular wall in a 13-year-old patient with HCM, showing complete relief of outflow obstruction, the site of the septal myectomy, considerable reverse remodeling of the rest of the ventricular wall, and normalization of the left ventricular volume at end diastole. Segmentation by Mohamed Nagy, Biomedical Engineering and Innovation Laboratory, Aswan Heart Centre. 3D, 3-dimensional; HOCM, hypertrophic cardiomyopathy; CT, computed tomography.

Tailoring the surgical approach on the basis of the individual patient's anatomy using advanced technology may be key in the success of the operation and personalized medicine.

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