Operative Techniques in Thoracic and Cardiovascular Surgery

Valve-Sparing Konno and Hypertrophic Obstructive Cardiomyopathy in Children

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Introduction

Septal myectomy is considered to be the most appropriate surgical treatment for patients with hypertrophic obstructive cardiomyopathy (HOCM) and severe symptoms that are unresponsive to medical therapy.[1] Extended transaortic septal myectomy is the procedure of choice in most centers. It provides safe and effective relief of left ventricular outflow tract (LVOT) obstruction. A satisfactory procedure includes subaortic myectomy extended to the midventricular level and correction of anomalies of the mitral papillary muscles. Failure to achieve adequate septal resection or to correct associated mitral anomalies may lead to residual obstruction and the need for reoperation.[2] Adequate septectomy must be carried out apically far enough to prevent contact of the anterior mitral leaflet with the septum at the midventricular level. It is also essential to address adequately associated mitral valve anomalies, such as anomalous papillary muscle insertion directly onto the anterior mitral leaflet, extensive fusion of papillary muscles with the ventricular septum, and abnormal insertions of accessory papillary muscles or chordae tendineae onto the ventricular septum.

In the pediatric population with HOCM, LVOT obstruction may develop either in early infancy or, otherwise, later on during adolescence.[3] In both groups, severe LVOT obstruction may increase rapidly and a surgical approach is recommended because high LVOT gradient is recognized as a risk factor for sudden death. Extended transaortic septal myectomy provides satisfactory results, particularly in experienced centers.[1,4] However, the procedure is associated with an increased risk of residual or recurrent obstruction and of iatrogenic injury to the aortic and the mitral valves.[4] In young children, the operation may be technically challenging because of the difficulty of exposure of the intraventricular lesions through a small aortic orifice. Limited visibility may yield to inadequate resection at the midventricular level or inability to address mitral valve anomalies.

The modified valve-sparing Konno procedure, first described by Cooley and Garrett in 1986, is usually used to relieve tunnel or complex subaortic stenosis while preserving the aortic valve.[5,6] The modified Konno procedure provides excellent exposure of the LVOT, even in small children, thus allowing extensive muscular resection as well as correction of mitral valve anomalies. It is thus particularly indicated in severe forms of HOCM.[7] Since 1990, the modified Konno operation is our procedure of choice in children with HOCM.[8,9] Associated mitral valvuloplasty to correct severe systolic anterior motion of the mitral valve is performed in selected cases, using the retention plasty technique, as described by Delmo Walter et al.[10] Implantation of an epicardial cardiac defibrillator is also carried out, if indicated (history of syncope or sudden death presumably related to arrhythmia, very severe ventricular septal thickness, and a positive family history of sudden death) (Figs. 1-11).

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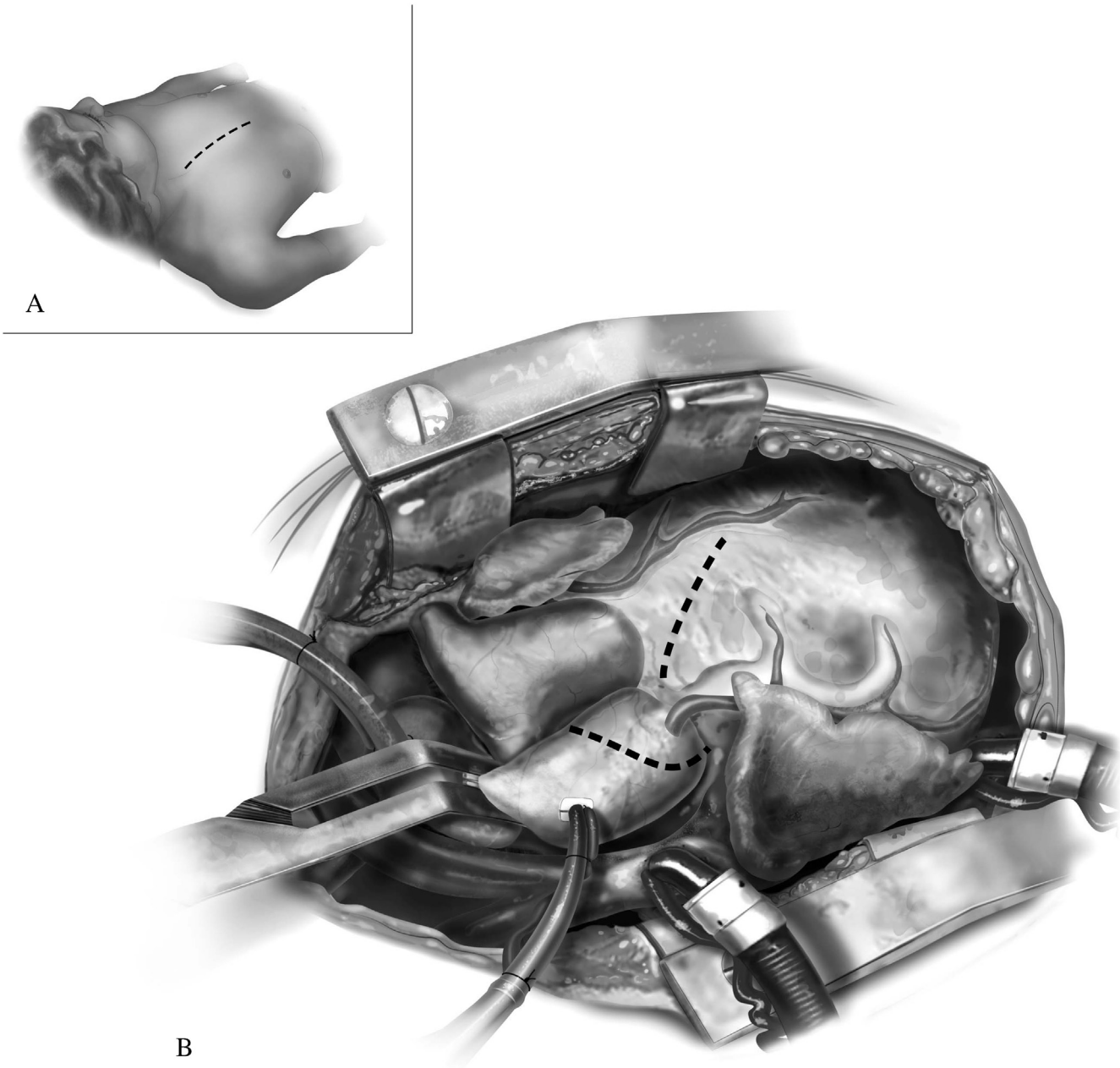

Figure 1 A median sternotomy is performed. Cardiopulmonary bypass is instituted using 2 caval cannulas and an ascending aorta cannula. The aortic cannula is inserted as distally as possible into the ascending aorta. A left vent is placed directly into the left atrium. Cardiopulmonary bypass is carried out in normothermia and using conventional hemofiltration. Because of the presence of severe myocardial hypertrophy, great care should be taken to achieve optimal intraoperative myocardial preservation. Multidose warm blood cardioplegia is our first choice. The initial infusion is given into the ascending aorta; reinfusion doses are administered every 10-12 minutes directly into the coronary ostia. Alternatively, cold histidine-tryptophan-ketoglutarate cardioplegia (Custodiol, Custodiol HTK, Köhler Chemie GmbH, Bensheim, Germany) can be used, particularly in small children, to avoid repeated cannulation of the coronary ostia.

The ascending aorta and the pulmonary trunk are dissected apart as near as possible to the aortic annulus. A diagonal incision is made in the aorta, extended under direct vision, deep into the noncoronary sinus, such as to gain perfect exposure of the aortic valve and the subaortic area. The right ventricle is opened transversely below the pulmonary valve, care being taken not to injure important coronary arteries.