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## Cephalad Resuspension of Aortic Leaflet of the Mitral Valve in Hypertrophic Obstructive Cardiomyopathy

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**A young child with hypertrophic obstructive cardiomyopathy, who was reoperated on for recurrent left ventricular outflow tract obstruction, underwent myectomy with patch augmentation of the ventricular septum. Cephalad resuspension of the aortic leaflet of the mitral valve resulted in abolishment of residual obstruction occurring secondary to its systolic anterior motion.**

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**H**ypertrophic obstructive cardiomyopathy (HOCM) is a primary hypertrophy of the myocardium associated with a small left ventricular cavity, increased systolic function, impaired diastolic function, and a variable, dynamic obstruction across the left ventricular outflow tract. Systolic anterior motion (SAM) of the mitral valve leaflets usually contributes to the obstruction. The mechanism of SAM consists of closure of the mural leaflet against the body of the aortic leaflet at approximately the junction of its mid- and free-edge third (rather than between the free-edge third and the free edge as in the normal heart). The free-edge portion of the aortic leaflet beyond the point of coaptation angulates on the remainder of the leaflet in a cephalad direction toward the aortic orifice. As a secondary event, the higher pressure below the aortic leaflet then forces it further into the left ventricular outflow tract. As a result, the left ventricular ejection is rapid and, of necessity, occurs predominantly in early systole. The degree of SAM and the diameter of the left ventricular outflow tract at the site of SAM correlate with the severity of the obstruction [1]. Systolic anterior motion is temporally related to both the peak of

the left ventricular outflow gradient and cessation of flow in the ascending aorta [2]. Residual SAM after repair of HOCM can pose major problems. We report such a case and describe a modified surgical technique to abolish SAM.

A 5-year-old boy with HOCM in whom a septal myectomy had been performed 3 years previously at another hospital was referred with a recurrent gradient across the subaortic outlet of 75 mm Hg. Echocardiographically, there was residual hypertrophy of the ventricular septum at the level of the free edge of the aortic leaflet of the mitral valve in its open position, with marked SAM of the valvar leaflets.

At reoperation, during cardiopulmonary bypass with aortic and bicaval cannulation, a transverse aortotomy was made with extension toward the commissure between the left and right aortic valve leaflets. A second longitudinal incision was made in the right ventricular infundibulum. Exposure through both incisions facilitated the incision and extensive resection of the hypertrophied area of the left ventricular septum. Subsequently, the ventricular septum and the right ventricular infundibulum were augmented with glutaraldehyde-treated autologous pericardial patches. The aortotomy was closed. A transesophageal echocardiogram after the patient had been weaned from cardiopulmonary bypass demonstrated a satisfactory reduction in ventricular septal width and marked enlargement of the subaortic area. Nonetheless, there was still severe SAM of the mitral valve, with a 45 mm Hg gradient across the outflow tract. The mural leaflet was seen to coapt with the midsegment of the aortic leaflet. Cardiopulmonary bypass was reinstated, a right atriotomy made, and the interatrial septum incised at a distance of approximately 2 cm parallel to the annulus of the septal leaflet of the tricuspid valve (thereby carefully avoiding the area of the atrioventricular node) (Fig 1). The aortic leaflet of the mitral valve, with the exception of its lateral edges, was detached from the membranous and muscular atrioventricular septal areas and resuspended in a cephalad position with a continuous 5-0 polypropylene suture (Prolene; Ethicon, Inc, Somerville, NJ). The maximum elevation at the midsegment of the leaflet was 5 mm.

Repeat echocardiography after weaning from cardiopulmonary bypass demonstrated closure of the mural leaflet against the aortic leaflet between the free edge and the central segment with absence of SAM. Simultaneous pressure measurements in the left ventricle and aortic root demonstrated a systolic gradient of 12 mm Hg. The further postoperative course was uncomplicated. At 18 months' follow-up, there has been no recurrence of obstruction within the subaortic outflow tract.

### Comment

Infants and children presenting with symptomatic HOCM usually represent the severe end of the spectrum,

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with marked left ventricular hypertrophy, frequent congestive heart failure, and a relatively high incidence of sudden death. Based on the substantial risk of sudden death and the potential for noncompliance with long-

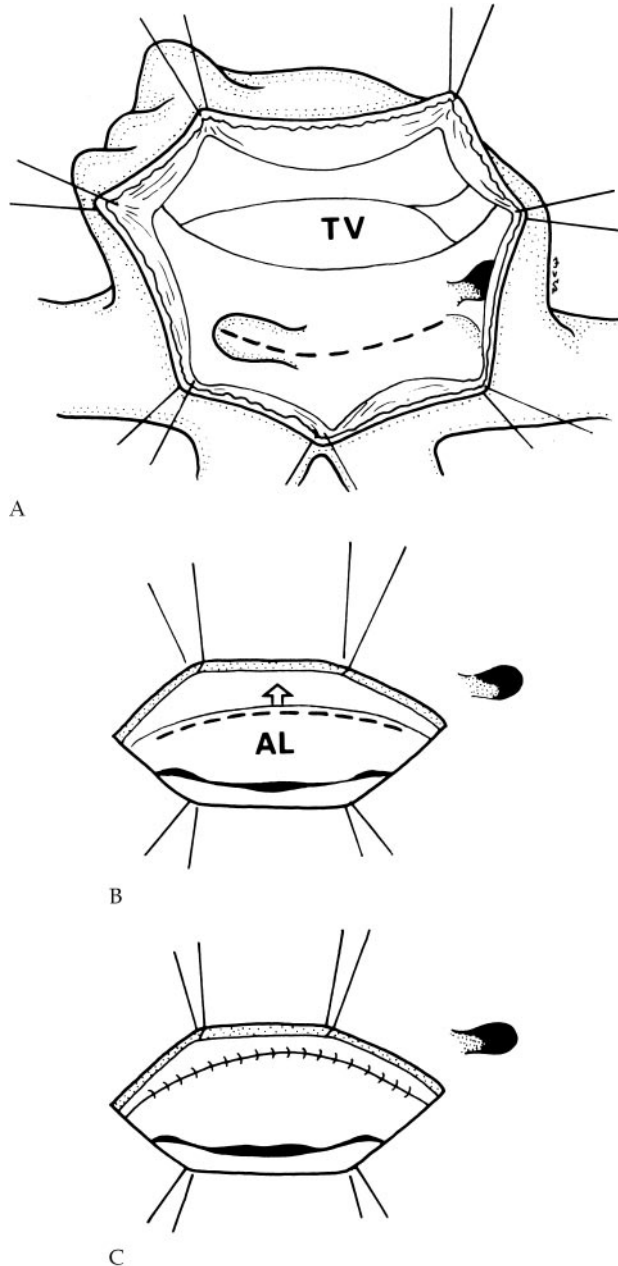


Fig 1. Cephalad resuspension of aortic leaflet of the mitral valve. (A) Through a right atriotomy, starting at the fossa ovalis, the interatrial septum is incised parallel to the septal leaflet of the tricuspid valve (TV). (B) The mitral valve is exposed by application of traction sutures on the incised interatrial septum. Dashed line indicates incision of aortic leaflet (AL) at its insertion. The aortic leaflet is elevated (arrow) with the exception of its lateral edges. (C) Resuspension of the aortic leaflet at a higher level results in enlargement of the subaortic space and transposition of the zone of coaptation of the aortic leaflet with the mural leaflet closer to the free edge of the aortic leaflet.

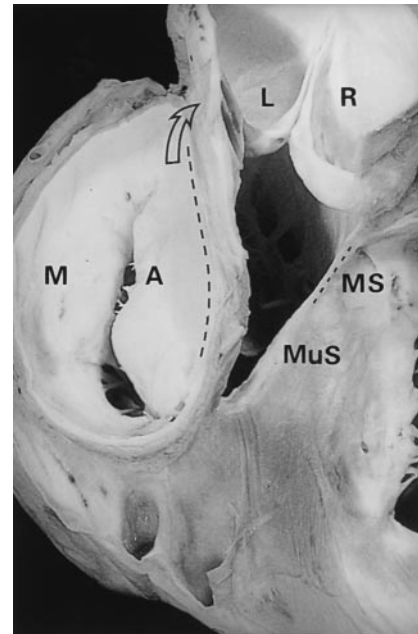


Fig 2. Wedging of posterior extension of the subaortic outflow tract between the axis of opening of the mitral valve and plane of the atrioventricular septum in the normal heart. The noncoronary sinus of Valsalva has been removed. Dashed line indicates the incision in the aortic leaflet (A) of the mitral valve. Cephalad resuspension of the aortic leaflet (arrow) enlarges the subaortic space. (Dotted line = location of membranous septum [MS]; L = left sinus of Valsalva; M = mural leaflet; MuS = muscular septum; R = right sinus of Valsalva.)

term medical therapy, the management of such patients is difficult. Moreover, there is little evidence that long-term survival is positively affected by treatment with calcium-channel blockers or  $\beta$ -adrenergic receptor-blocking drugs. The only potentially effective drug for lowering the risk of sudden death in HOCM is amiodarone. Unfortunately, toxicity of amiodarone (interstitial pneumonitis, atrioventricular block, and left ventricular dysfunction) is related to the dose and duration of treatment, and long-term administration in young patients seems ill advised if other effective therapies are available.

Recently, dual-chamber pacing has been proposed as an alternative to surgical treatment for relief of symptoms in patients with HOCM [3]. Although this novel therapy has been reported to lead to clinical improvement in some patients, there is concern about the observed deterioration of ventricular diastolic function and the increase in left atrial pressure.

A variety of surgical procedures have been advocated for relief of subaortic obstruction secondary to HOCM, including (extended) left ventricular septal myectomy, if necessary, combined with a modified Konno procedure, or, in adult patients, replacement of the mitral valve [4-7]. In infants and young children with HOCM and associated aortic valvar disease, we have performed the combined resection of left ventricular septal myocardium

and a Ross-Konno operation, thereby augmenting the ventricular septum with a flap of infundibular muscle that is adherent to the pulmonary valve autograft.

In the presented case, despite extensive resection and additional patch augmentation of the hypertrophied septum, SAM persisted, thereby generating residual subaortic obstruction. In analogy to plication of the aortic leaflet of the mitral valve [7], cephalad resuspension reduces its redundancy, thereby transposing the zone of coaptation with the mural leaflet closer to the free edge of the aortic leaflet, with resulting abolishment of SAM. A secondary beneficial effect of the resuspension maneuver (unlike plication), in analogy to that reported in atrioventricular canal with absent or restrictive interventricular communication and associated obstruction of the subaortic outlet [8], is the creation of an enlarged subaortic space. This is based on the fact that resuspension results in a wider angle between the axis of opening of the mitral valve and the plane of the ventricular septum, thereby lessening the degree of wedging of the posterior extension of the subaortic outflow tract (Fig 2). The risk of creating mitral valve regurgitation by the resuspension maneuver is low because the aortic-mitral curtain is a stationary fibrous structure and the proximal half of the aortic leaflet, which is hinged from this curtain, is not very mobile, with only the distal half moving during the cardiac cycle [9].

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## Absent Pulmonary Valve Syndrome With Interrupted Aortic Arch

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A surgically treated case of absent pulmonary valve syndrome associated with type B interrupted aortic arch is presented. The presence of a restrictive ductus arteriosus promoted the development of a collateral circulation between ascending and descending thoracic aorta, allowing the child to remain clinically stable after birth.

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Chavers [1] in 1847 first described congenital absence of the pulmonary valve. Since then many cases of absent pulmonary valve syndrome (APVS) have been reported. The lesion has rarely been described in isolation and more often is associated with various congenital heart defects, usually tetralogy of Fallot. We report on a patient with APVS and interrupted aortic arch, and analyze the role the restrictive ductus arteriosus may have played.

A fetal echocardiogram at 21 weeks' gestation demonstrated dextrocardia, situs solitus, and concordant atrioventricular and ventriculoarterial connection. There was a large ventricular septal defect, pulmonary stenosis and regurgitation, and dilatation of the main pulmonary artery consistent with APVS (Fig 1). Pulsed-wave Doppler echocardiography suggested a pressure drop across the ductus, from pulmonary artery to descending aorta.

A male infant weighing 2.6 kg was delivered at term. Pulses were palpable and there was no discrepancy between upper and lower limb pressures. In addition to the antenatal findings, echocardiography demonstrated type B interrupted aortic arch, but with pulsatile flow in the descending aorta. The ductus arteriosus remained restrictive, with a continuous left-to-right flow and a peak. Doppler velocity of 3 m/s. As the baby was asymptomatic and his circulation was not ductus dependent, an elective operation was postponed to allow for a period of growth. After a few months, a remarkable discrepancy between head circumference (90th centile) and body

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