Repair for Congenital Mitral Valve Stenosis

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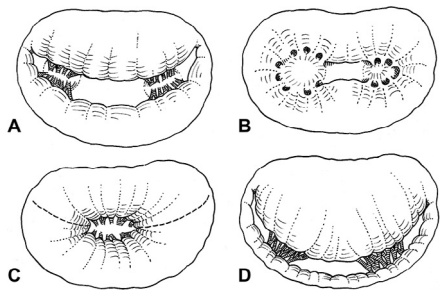
We report the techniques and long-term outcome of mitral valve (MV) repair to correct congenital mitral stenosis in children. Between 1986 and 2014, 137 children (mean age 4.1 ± 5.0, range 1 month-16.8 years) underwent repair of congenital mitral stenosis (CMS). In 48 patients, CMS is involved in Shone's anomaly. The typical congenital MS (type I) was seen in 56 patients. Hypoplastic MV (type II, n = 15) was associated with severe left ventricular outflow tract abnormalities and hypoplastic left ventricular cavity and muscle mass. Supravalvar ring (type III, n = 48) ranged from a thin membrane to a thick discrete fibrous ridge. Parachute MV (type IV, n = 10) have 2 leaflets and barely distinguishable commissures, but all chordae merged either into 1 major papillary muscle or asymmetric papillary muscles—1 dominant and the other minuscule. Hammock valve (type IV, n = 8) appeared dysplastic with shortened chordae directly inserted into the posterior left ventricular muscle mass. MV repair was performed using commissurotomy, chordal division, papillary muscle splitting and fenestration, and mitral ring resection, each applied according to the presenting morphology. During the 28-year follow-up period, 23 patients underwent repeat MV repair and 3 underwent MV replacement after failed attempts at repeat repair. At 1 and 15 years postoperatively, freedom from reoperation was 89.3 ± 5.1% and 52.8 ± 11.8%, and cumulative survival rates were 92.3 ± 4.3% and 70.3 ± 8.9, respectively. Mortality unrelated to repair accounted for 9 (20%) deaths. Long-term functional outcome of MV repair in children with CMS is satisfactory. Repeat repair or replacement may be deemed necessary during the course of follow-up.

Keywords: congenital mitral stenosis, parachute-like asymmetric mitral valve, parachute valve, hammock valve, hypoplastic mitral valve, Shone's anomaly, mitral valve repair

Introduction

Congenital mitral stenosis (CMS) is complex[1] and is typically associated with other congenital heart diseases. It is rarely found in isolation.[2,3] The associated congenital heart lesions may hide, or be hidden by, the mitral valve (MV) stenosis, which is exemplified in Shone's anomaly,[4-7] wherein the additional left heart obstructive lesions complicate its management. As such, surgeons are faced with what optimal strategy to offer to patients with CMS, especially in severe cases wherein medical or interventional therapy entails significant hemodynamic compromise.

Over the years, surgical treatment has been focused on a conservative approach, which provides relief of mitral stenosis, albeit for a short duration, depending on its severity and anatomic substrate as well as associated hemodynamically significant cardiovascular anomalies. Interventional treatments, such as percutaneous transcatheter balloon mitral valvuloplasty,[8] are utilized for medically refractory CMS, the goal of which is to reduce left ventricular inflow obstruction and left atrial (LA) pressure, hopefully



Appearance of a normal mitral valve (A) in comparison with stenotic mitral valves: parachute-like asymmetric valve (B), parachute valve (C), and hammock valve (D).

Central Message

For patients with congenital mitral stenosis, a repair strategy using techniques tailored to the presenting morphology demonstrated that repair can be performed in this population with satisfactory long-term survival and freedom from repeat repair and replacement. In patients with successfully repaired valves, adequate mitral valve function is maintained over a long time.

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producing lasting relief but minimally improving symptoms and delaying MV replacement until the patient is older and larger. This is true for isolated mitral stenosis. However, this approach is not optimal when there are associated left ventricular outflow tract (LVOT) obstructive lesions, such as coarctation of aorta and subaortic stenosis, wherein creation of a widely patent and competent left ventricular inflow leads to concerns about the inadequate loading of the left ventricle, which creates an impedance to left ventricular ejection, decreasing cardiac output and inability to sustain postoperative hemodynamics.[9,10] In infants and young children, surgical MV repair improves mitral leaflet mobility and provides adequate and effective inflow area; however, it may not be long lasting and there may be a need for eventual valve replacement.

With the advent of excellent diagnostic and imaging modalities, providing optimal guidance to assess the MV and surgically relevant anatomy, as well as the concomitant obstructive lesions, considerable improvements in perioperative management and outcome have been seen in this group of patients.

This study reports our institutional experience on surgical strategy to correct CMS, the operative results, and the long-term functional outcome in infants and children.

Patients and Methods

The institutional review board approved this retrospective or prospective study and waived the need for patient consent.

Between June 1986 and July 2014, 137 infants and children (mean age 4.1 ± 5.0, median 2.9, range 1 month-16.8, years) underwent surgical correction of CMS (Table 1). Medical records including preoperative evaluations, operative notes, and follow-up data were reviewed. All patients were in modified Ross/New York Heart Association functional class III.

Forty-three of the 58 patients with type I CMS underwent previous balloon mitral valvuloplasty, with 12 having a repeat intervention before the definite surgical procedure.

In 48 patients, CMS is involved in Shone's anomaly (Fig. 1), with multiple left ventricular inflow and outflow tract obstructive lesions (Table 1). In this group, it is very difficult to define which of the associated cardiac lesions is the predominant cause of symptoms. Relief of mitral stenosis unmasks any existing LVOT lesions. In our early years of experience, approach in this group is multistage, that is, treat the presenting lesions as they are unmasked. Later on, the advent of modern imaging modalities made it possible to diagnose all the other concomitant obstructions. Thus, we preferred single-stage surgery, wherein the optimal strategy is governed by the morphology of each obstructive lesion and favored MV repair on all patients with transmitral gradient >5 mm Hg, which, in this population, were underscored.

Follow-up outpatient records were provided by written correspondence from the referring physicians. No patients were lost to follow-up.

Anatomical Evaluation of MVs

These 137 children and adolescents with CMS were submitted to a complete two-dimensional echocardiographic examination before surgery, at the time of discharge from hospital and in a series of follow-ups.

Although most patients showed complex structural abnormalities in each of the valvular components (leaflets, chordae, papillary muscles), CMS was defined according to Ruckman and Van Praagh's classification (Table 1).[1] Thickened and rolled leaflets, short chordae tendineae, partial or complete obliteration of interchordal spaces by fibrous tissues, underdeveloped papillary muscles, and commissural fusion (type I, typical congenital MS, Fig. 2A) were seen in 56 patients. Hypoplastic MV (type II, Fig. 2B), described as small MV orifice, shortened chordae tendineae and small papillary muscles, was seen in 15 patients. This form of MS was associated with severe LVOT abnormalities, in all cases. Also seen was underdeveloped left ventricular cavity and muscle mass.

Supravalvar mitral ring (type III, Fig. 2C) was seen in 48 patients; this is described as a circumferential ridge of connective tissue that originates at the LA wall overlying the MV leaflets and frequently attached to the annulus. Variable in thickness and extent, it ranged from a thin membrane to a thick discrete fibrous ridge. The membrane was often adherent to the anterior MV leaflet. Adhesion to the valve impaired leaflet mobility. This was associated with variable abnormalities of the MV subvalvar apparatus.

Parachute MV (type IV, Fig. 2D), seen in 10 patients, has the usual 2 leaflets and commissures; however, all chordae tendineae are merged into 1 major papillary muscle, instead of being inserted into 2 papillary muscles. The valve naturally was deformed, and the chordae were short and thick; this, coupled with their convergent papillary insertion, allowed restricted leaflet mobility, thus creating a stenotic MV as the leaflets were closely apposed, greatly reducing the effective mitral orifice area. The only functional communication between the left atrium and the left ventricle was through the interchordal spaces. In aggregate, these spaces did not allow free egress of blood from the left atrium.

Parachute-like asymmetric MV (type IV, Fig. 3) has a large dominant papillary muscle directly fused with the leaflets, absence of chordae, and presence of only fenestrations. However, the other papillary muscle is very small with just few short chordae, causing an asymmetric location of the valve orifice.

Hammock valve (type IV, Fig. 4), defined as dysplastic with shortened chordae directly inserted in a muscular mass of the posterior LV wall resulting in tethering of both leaflets, was seen in 8 patients. The valvar orifice is partially obstructed by intermixed chordae and abnormal papillary muscles, characteristically implanted underneath the posterior leaflet. The chordae tendineae of the anterior leaflet cross the orifice toward the posteriorly implanted papillary muscles, producing the hammock appearance. In extreme cases, the hammock valve contains a fibrous diaphragm with scattered holes that allow the blood to flow from the left atrium to the left ventricle.

The left-sided obstructive lesions encountered were coarctation of the aorta (n = 48), subaortic stenosis (n = 35), and hypoplastic aortic arch (n = 5). Associated cardiac anomalies were patent ductus arteriosus, atrial septal defect, ventricular septal defect, and vascular ring (Table 1).