Long-term surgical outcome of mitral valve repair in infants and children with Shone’s anomaly†

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Abstract

OBJECTIVES: Having assembled information on a large series of patients with Shone’s anomaly with the longest follow-up, we studied the operative results and long-term outcome of mitral valve (MV) repair techniques performed to correct the left ventricular inflow tract lesions of this congenital anomaly.

METHODS: Between 1986 and 2011, 45 infants and children [mean age 5.16 ± 5.0 (median 3.9; range 2 months to 16.8 years] underwent surgical correction of Shone’s anomaly. Left ventricular outflow tract obstructive lesions consisted of coarctation of the aorta, found in 89%, subaortic stenosis due to fibromuscular hypertrophy in 71% and subvalvular membrane in 51%. Left ventricular inflow tract obstructions concentrated on the mitral valve included mitral ring in all cases and the gamut of small and narrowed mitral ori.

RESULTS: MV repair was performed using commissurotomy, division of chordae tendinae, papillary muscle splitting and fenestration, and resection of mitral ring, applied according to the presenting morphology in patients with either previously-corrected or concomitant correction of the left-sided obstructive lesions. Postoperative echocardiography showed absence of MV stenosis and immediate improvement of symptoms, except in a 3-month-old infant who died 18 days postoperatively due to myocardial failure. During the 23-year follow-up, 23 patients underwent repeat MV repair and one underwent MV replacement after failed attempts at repair. Mean duration of follow-up was 17.5 ± 1.5 years (range 6.4–22.7 years). Freedom from reoperation was 97.6 ± 2.4%, 89.3 ± 5.1%, 77.1 ± 7.2%, 72.0 ± 8.3% and 52.8 ± 11.8%, at 30 days, 1, 5, 10 and 15 years postoperatively, respectively. Mortality unrelated to valve repair accounted for nine deaths (20%). Age at operation, severity of mitral abnormalities and concomitant left ventricular outflow tract lesions proved to be independent risk factors for reoperation and mortality (P < 0.05).

CONCLUSIONS: Long-term functional outcome of mitral valve repair in children with Shone’s anomaly is satisfactory. Repeat MV repair and/or replacement may be deemed necessary during the course of follow-up.

Keywords: Shone’s anomaly • Mitral valve stenosis • Congenital heart anomalies • Mitral valve repair

INTRODUCTION

Forty-eight years after Shone and colleagues described the developmental complex of four potentially obstructive lesions—consisting of parachute mitral valve (MV), subvalvular mitral ring, subaortic stenosis and coarctation of the aorta—there have been four other published studies reporting their long-term operative outcomes [1-5]. Other reports on Shone’s anomaly (also known as Shone’s complex and Shone’s syndrome) were included in treatises dealing with congenital mitral stenosis (MS), such as that of Serraf and colleagues, who categorized 25 patients with Shone’s syndrome among their reported series of 72 infants with congenital MS, and other sporadic case reports describing the anomaly [6-9]. In this respect, there has been a tremendous paucity of information. This is because paediatric patients with Shone’s anomaly are a rare occurrence and a total of only 134 cases operated on with long-term outcomes have been specifically reported in the literature to date. Another reason could be that patients in this subset are generally considered a poor surgical risk and are often excluded from reports dealing with each of the individual disease components.
The finding in Shone's original description—that the extent of MV involvement seems to be the predominant factor determining outcome—is supported by the review of 30 cases by Bolling and colleagues, which represents the most comprehensive report to date and comprises patients with a multitude of anatomic variants and different management approaches [2]. The same observation was reported by Brauner et al. in their study of 19 cases, Brown et al. with their reported 27 cases and Ikemba et al., who published MV morphology and morbidity/mortality in 50 patients with Shone's complex [3-5].

Having collected data on a large series of Shone's anomaly with the longest follow-up, we analysed the operative results and long-term outcome of MV repair techniques performed to correct the left ventricular inflow tract lesions of this congenital anomaly.

**PATIENTS AND METHODS**

The Institutional Review Board approved this study and waived the need for patient consent.

**Patients**

Between June 1986 and March 2011, 45 infants and children with mean age 5.16 ± 5.0 years (median 3.9; range 2 months to 16.8 years) with multiple left ventricular inflow and outflow tract obstructive lesions (Table 1) underwent surgery at the Deutsches Herzzentrum Berlin. Their medical records were reviewed, including preoperative evaluations, operative notes and follow-up data. Subjects’ follow-up records were provided by both the Department of Congenital Heart Disease/Paediatric Cardiology and Department of Clinical Studies, Deutsches Herzzentrum Berlin, and by written correspondence from the referring physicians. No patients were lost to follow-up. The end of the follow-up study was March 2011.

These children were divided into age groups as follows: ≤ 1 year: n = 12; mean age 0.4 ± 0.27 months (median 0.32; range 0.02–0.96 months); 1–5 years: n = 19; mean age 3.5 ± 1.4 years (median 3.6; range = 1.1–4.8 years); >5–10 years: n = 8, mean age 8.6 ± 2.0 years (median 8.6; range 6.2–9.2 years) and >10 years: n = 6, mean age 13.2 ± 1.6 years (median 15.2, range 11.2–16.8 years). All patients were in modified Ross functional Class III (n = 31) or the comparable NYHA Class III (n = 14). The obstructive lesions of Shone’s anomaly are shown in Table 1. Echocardiograms were taken peripheratively and serially during follow-up.

**Anatomical evaluation of mitral valves**

All 45 children and adolescents 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. All had abnormal MVs with mitral ring and at least two other left heart obstructive lesions, severe enough to require intervention. MV anomalies were defined according to Ruckman and Van Praagh's classification of MS [10]. 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) were seen in 24 patients. Hypoplastic MV (Type II), described as small MV orifice, shortened chordae tendineae and small papillary muscles, was seen in five patients. This form of MS was associated with severe left ventricular outflow tract (LVOT) abnormalities in all cases. Supravalvular mitral ring (Type III) was seen in 45 patients This is described as a circumferential ridge of connective tissue that originates at the left atrial 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 subvalvular apparatus. Parachute MV (Type IV) was seen in eight patients, described as having the usual two MV leaflets and commissures but all chordae tendineae, instead of inserting into two papillary muscles, merged into one major papillary muscle. Naturally, the valve 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, since 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.

The most prevalent left-sided obstructive lesion was coarctation of the aorta, seen in 40 patients (89%). Additional left-sided lesions included bicuspid aortic valve, seen in 17 (38%) with aortic valve stenosis. Hypoplastic aortic arch was seen in eight (18%), diffuse subaortic stenosis due to fibromuscular hypertrophy in 32 (71%) and discrete membrane in 23 (51%).

Associated cardiac anomalies were ventricular septal defect (11; 24%), atrial septal defect (15; 33%), patent ductus arteriosus (28; 62%) and vascular ring in three (7%).

**Haemodynamic evaluation**

Systolic pressure gradients in the area of subaortic obstruction ranged from 18 to 100 mmHg (mean 65.2 ± 13.7 mmHg) and
coarctation gradients ranged from 25 to 80 mmHg (mean 53.2 ± 13.7 mmHg). Eleven patients presented with symptoms of congestive heart failure and three, with associated ventricular septal defect, had significant pulmonary hypertension (mean pulmonary artery pressures, at rest and during exertion, of 32 and 47 mmHg, respectively, with pulmonary to systemic flow ratio [Qp/Qs] >1.5).

A gradient ≥5 mmHg across the MV was considered significant and was observed in all 45 children (100%): mean value was 20.8 ± 4.7 mmHg; range 8–30 mmHg.

**Echocardiographic evaluation of mitral stenosis**

Mitral stenosis was quantified by measurement of the MV orifice area (cm²) and mean resting end-diastolic gradient (mmHg) and was graded as zero (4–6 cm², 0 mmHg); grade I (mild: 2–4 cm², <5 mmHg); grade II (moderate: 1–2 cm², 5–10 mmHg); or grade III (severe: <1 cm², >10 mmHg). Preoperatively, 34 patients had moderate MS (grade II) while 11 had severe (grade III) MS.

**Surgical technique**

Forty-five patients underwent a total of 367 procedures to repair the left ventricular inflow and outflow tract obstructive lesions (Tables 2 and 3) including the repair of associated anomalies (Table 4).

A total of 141 procedures were performed for obstructive MV lesions. Indications for MV repair were abnormal MV with <2 cm² orifice area, mean resting end-diastolic gradient of >5 mmHg, presence of mitral ring, and associated left ventricular outflow obstructive lesions. Haemodynamic criteria included pulmonary artery pressure of >25 mmHg at rest and >30 mmHg on exertion. Increased left atrial- to left ventricular pressure gradient was an indication for repeat MV surgery.

Our approach to the left ventricular inflow- and outflow tract obstructive lesions is single-stage surgery whenever possible. Single-stage repair was done in 23 patients because all obstructive lesions and intracardiac defects that were deemed significant were diagnosed all at one time. However, this did not preclude eventual reinterventions when necessary. Staged repairs were done in 22 patients, on whom the first operations were usually those that addressed the left ventricular outflow tract, followed by interventions that were carried out as it was established that the other concomitant lesions appeared sufficiently haemodynamically significant to warrant surgery.

The majority of patients in our series presented initially in the neonatal period with coarctation of the aorta (n = 12) and in the 1–5 year old group (n = 19), it is the predominant outflow obstructive lesions. The neonatal coarctation cases presented with severe symptoms, which could have masked the other intracardiac pathology until repaired.

In general, we first treated the most distal obstructive lesions (usually coarctation).

There was no discrimination in this study population. All 45 children, on whom MV surgery was performed at any stage in

| Table 2: Surgical procedures performed on the left ventricular outflow tract |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|-----------------|
| Surgical procedures             | Number of procedures | 1st intervention | 2nd intervention | 3rd intervention | Total |
| Repair of coarctation of aorta   | 40               | 12              | 4               | 56              |
| Enlargement of hypoplastic aortic arch | 5         | 1              |                 | 6               |
| Septal myectomy                  | 32               |                 |                 | 32              |
| Resection of subvalvular membrane| 23               | 7              |                 | 30              |
| Konno–Rastan aortoventriculoplasty | 3           |                 |                 | 3               |
| Aortic valve commissurotomy      | 13               | 8              | 3              | 24              |
| Aortic valve dilatation          | 9                | 7              | 2              | 18              |

*Ross procedure (n = 5), aortic valve replacement (n = 2), ascending aortic replacement (n = 1); aortic valve replacement (n = 3); aortic valve repeat dilatation (n = 5), aortic valve replacement (n = 1); aortic valve replacement (n = 2).

| Table 3: Surgical procedures performed on the left ventricular inflow tract, the mitral valve, in 45 patients |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|-----------------|
| Surgical procedures             | Number of procedures | Primary MV repair | Repeat MV repair | 3rd intervention | Total |
| Resection of mitral ring        | 45               | 12              |                 | 57              |
| Repair of parachute valve       | 8                | 1              | 1*             | 10              |
| Commissurotomy                  | 32               | 10             |                 | 42              |
| Papillary muscle division       | 20               | 5              |                 | 25              |
| Chordal division                | 4                | 3              |                 | 7               |

*MV replacement.

| Table 4: Concomitant procedures performed |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
| Procedure                        | n (%)           |
| Ligation of patent ductus arteriosus | 28 (62.2)     |
| Closure of atrial septal defect  | 15 (33.3)       |
| Closure of ventricular septal defect | 11 (24.4)     |
| Resection of vascular ring       | 3 (6.7)         |
their disease, had significant transmitral gradient that necessitated intervention. Since there were no patients with hypoplastic-, small- or even borderline left ventricle, all patients underwent biventricular repair.

MV repair was performed through a median sternotomy under cardiopulmonary bypass and moderate systemic hypothermia. Antegrade intermittent cold crystalloid cardioplegia with topical hypothermia was used for myocardial protection. Through a left atriotomy along the interatrial groove, the mitral annulus, leaflets, chordae tendinae and papillary muscles were exposed and meticulously inspected to determine the precise nature of the lesion. Leaflet coaptation was assessed with a forceful transvalvular injection of saline with a bulb syringe. Using a nerve hook, the coaptation of the anterior and posterior leaflet was assessed for the presence of sufficient tissues along the coaptation plane. The valve orifice area was assessed with a Hegar dilator and, more recently, with a Ziemer-Hetzer valve sizer (Fehling Instruments GmbH, Germany). The nomogram published by Rowlatt et al. was helpful in determining the normal valve diameter for a specific body surface area [11].

Various repair techniques were employed, according to the presenting valve morphology causing MS.

In children with fused commissures, commissurotomy was carried out on both the anterolateral and posteromedial commissures. Division of chordae tendinae and division and splitting of papillary muscles were performed in patients with short, fused and matted chordae.

‘Parachute valve’ has the usual two MV leaflets and commissures, but all the chordae tendinae cluster into one major papillary muscle. It often presents as a funnel-type structure with some distinct fibrous lines at the sites of commissural fusion. The most appropriate site for leaflet-splitting incisions was defined on both sides from the common papillary muscles towards the ‘assumed’ trigones (commissurotomy and fenestration, Fig. 1A and B). These incisions were extended into the body of the papillary muscle, which was split towards its base, assuring sufficient thickness of both new papillary muscle ‘heads’ (Fig. 1 C–E) [12].

The degree and extent of incision, commissurotomy and fenestration were measured using a Hegar dilator and, more recently, a Ziemer-Hetzer valve sizer, as determined by the minimum acceptable age-related MV diameter [11].

Sharp dissection of the fibromembranous mitral ring was required to initiate the resection. It is very important to remove all components of the ring. Since the ring is usually within the

Figure 1: Parachute valve: (A) pre-repair; (B) commissurotomy and fenestration; (C) and (D) commissurotomy and splitting of the papillary muscles; (E) completed repair.
mobile portion of the leaflet, precautions were taken to avoid injuring the leaflet body itself when dissecting the ring off. Saline injection through the valves and intraoperative transesophageal echocardiography (TEE) were routinely performed to assess the adequacy of repair. In all the repair strategies we employed, the minimal final MV opening area in children should not be less than 10% below the norm, according to body surface area [11]. No mitral insufficiency ensued in 37 patients (82%) from the techniques employed, while eight (18%) had trivial or mild insufficiency. Regardless of the underlying pathology and techniques used, no patient was discharged from the hospital with more than mild stenosis/insufficiency. Postoperative transthoracic echocardiography was carried out annually, or as clinically indicated on the basis of symptoms. The degree of MS was estimated by means of standard echocardiographic measurement techniques. Assessment of MV function was divided by mean transmitral diastolic pressure (leaflet thickness, commissural fusion, valve pliability and morphology of the subvalvular apparatus).

Statistical analysis

All data were analysed using SPSS for Windows, version 16.0 statistical software (SPSS Inc., Chicago IL, USA). Data are expressed as absolute and percentage frequency values and continuous data as mean ± standard deviation, median and range, as appropriate. Repeated measurements of echocardiographic variables (MV area (cm²), annulus size (cm) and anterior leaflet area (cm²)) were analysed, in serial timelines and in relation with somatic growth, by running Friedman tests. Z-scores were calculated and numbers of values greater than 2 were counted. To facilitate comparison between preoperative and follow-up MV dimensions, z-values were computed as follows: z = (measured value minus mean value of normal controls), divided by SD of normal controls. To adjust for age, body size and growth-related changes in MV dimensions, linear measurements were indexed to the square root of body surface area (BSA⁰·⁵). Normal growth was defined as a lack of z-score change between early and late follow-up. Failure of growth of any dimension would be detected as a statistically significant decline in z-score. Freedom from reoperation and cumulative survival rates were analyzed according to Kaplan-Meier estimates, with 95% confidence interval (CI), and Cox proportional hazard regression methods, to identify the risk factors for reoperation and mortality. A value of P ≤ 0.05 was considered significant.

RESULTS

Morbidity

Four patients required permanent pacemaker implantation for complete heart block, performed within 30 days following the operation. These were the patients who also underwent concomitant closure of ventricular septal defects. There was no occurrence of other complications.

Early mortality

Early death occurred in a 3-month old infant who underwent urgent single-stage surgery for all the obstructive lesions of Shone’s anomaly with associated pulmonary hypertension (mean pulmonary artery pressure of 32 mmHg), ventricular septal defect and patent ductus arteriosus. Postoperatively, he had mild residual MS (MV orifice area of 2 cm² and mean resting end-diastolic gradient <5 mmHg). The left atrial pressure was 12 mmHg. He suffered from low-output cardiac failure, haemodynamic instability and hypotension 34 h postoperatively and was placed on an extracorporeal membrane oxygenator. Eventually he developed capillary leak syndrome and died on the 18th postoperative day.

Late mortality

Eight late deaths occurred among the 44 patients who were discharged from the hospital. A 2-month-old infant with typical congenital MS, who underwent repeat MV repair with concomitant repair of coarctation of the aorta and repeat septal myectomy 4 months later, died from heart failure a year after repeat surgery. A patient with parachute valve, who was 2 years old at the time of the initial MV repair, underwent repeat repair 5 years later. He subsequently underwent MV replacement 2 years afterwards but died 8 years after the replacement. Six patients died from a non-cardiac event at 1 (n = 1, episodes of seizures), 3 (n = 1, end-stage renal failure), 5 (n = 2, end-stage renal failure and pneumonia), 8 (n = 1, vehicular accident) and 13 (n = 1, unknown) years postoperatively. Cumulative survival rate was 97.6 ± 2.4%, 92.3 ± 4.3%, 83.8 ± 6.1%, 75.7 ± 7.8% and 70.3 ± 8.9%, at 30 days, 1, 5, 10 and 15 years postoperatively, respectively (Fig. 2A). Overall survival rates by age group during a mean duration of follow-up of 17.5 ± 1.5 years (range 6.4–22.7 years) are shown in Fig. 2B.

Operative outcome of mitral valve repair

Change in functional class. There was a significant improvement in functional class postoperatively (P < 0.001) and this was sustained until the late follow-up period (Fig. 3A).

Echocardiographic assessment

Change in severity of mitral stenosis. Absence of MS (mean MV orifice area 5.2 ± 0.8 cm² without mean resting end-diastolic pressure gradient) was noted after MV repair (P < 0.001). In the course of follow-up, 14 patients had developed significant MS (mean MV orifice area 2.3 ± 0.5 cm² and mean resting end-diastolic pressure gradient 8.7 ± 1.3 mmHg) warranting repeat intervention (Fig. 3B).

Mitral valve area and leaflets. The increase in MV area—which is substantial—from 21.65 ± 7.07 mm² to 26.12 ± 8.4 mm² [z-score -1.9 ± 1.8] (P = 0.000), implied MV growth over a mean period of 13.62 ± 2.37 years at 96% CI. The anterior mitral leaflet area, measured to be 24.88 ± 6.1 mm² as compared to a preoperative area of 22.29 ± 5.9 mm² [z-score -1.7 ± 2.0] (P = 0.000), had also grown.

Annular diameter. Planimetry of the annular area revealed a mild but significant increase in the growth of the annulus. The mean annular size increased to 35.76 ± 14.8 mm² from 32.94 ± 14.8 mm² [z-score -1.7 ± 2.1] (P = 0.000).
Figure 2: Cumulative survival: (A) overall; (B) stratified by age group.

Figure 3: Comparison of preoperative, postoperative, early and late follow-up: (A) functional class, (B) degree of mitral stenosis (MS). Freedom from reoperation: (C) overall; (D) stratified by age group.
All calculated z-scores at 1 year and 10 years after MV repair were normal when indexed to body surface area (z-score range -2.0 ± 2.0). As body surface area increased, there was an associated rise in MV dimensions. The mean atrial diameters and mass, which was 75.6% ± 18.1% of that predicted normal for body surface area after the first year and 94.6% ± 5.5% of that predicted normal after 10 years, suggests MV growth as appropriate for somatic growth.

Resection of mitral ring. Twelve patients (27%) who underwent primary resection of mitral ring had to undergo repeat resection. These were the patients with the membranous variety, which may not have been detected during the initial resection, since not only was the membrane adherent to the anterior MV leaflet but some tissue components remained proximal to the posterior mitral leaflet. These were also the patients with Type I congenital MS with fused commissures and thickened leaflets (n = 10); two had hypoplastic MV (Type II), of whom one had emphasized, shortened chordae and another had miniature papillary muscles. Along with the resection of the fibromembranous ring, they also underwent commissurotomy, chordal division and papillary muscle splitting. The latest echocardiographic evaluation of these patients showed absence of MS.

Follow-up

Freedom from reoperation after MV repair. Mean duration of follow-up was 17.5 ± 1.5 years (range 6.4–22.7 years). Freedom from reoperation was 97.6 ± 2.4%, 89.3 ± 5.1%, 77.1 ± 7.2%, 72.0 ± 8.3% and 52.8 ± 11.8%, at 30 days, 1, 5, 10 and 15 years postoperatively, respectively (Fig. 3C). Stratified, based on age groups at the time of initial repair, freedom from reoperation was 100% at 30 days in all age groups. In the <1 year olds, freedom from reoperation was 95.56% at 1 year and was sustained until the late follow-up period. Repeat MV repair was performed mostly in the 1–5 year age group until 10-year follow-up. In the ≥10 year-old group, it was noted that there was no repeat MV surgery 5 years after the initial MV repair (Fig. 3D). We performed only one MV replacement, and this was on a patient with parachute valve (see section on late mortality).

Thirty-two repeat MV procedures were performed on 14 patients, which were all related to restenosis and repeated MV dysfunction. These were performed mostly for Type I MS and hypoplastic MV.

Table 5 shows the multivariate analysis of perioperative risk factors for reoperation and mortality. In this series, the types of MS (Type I congenital MS with fused commissures and thickened leaflet [P = 0.004], hypoplastic MV (Type II) with emphasized shortened chordae and miniature papillary muscles [P = 0.001], supravalvar mitral ring [P = 0.002] and parachute valve [P = 0.003]), the associated pulmonary hypertension [P = 0.001] and the left ventricular outflow tract lesions [P = 0.00] are significant risk factors. However the repair techniques, as well as staged surgery, proved to be non-influential to the outcome in this study population.

**DISCUSSION**

This study comprises an institutional series of surgical treatment of children with Shone's anomaly, with emphasis on MV repair. The formidable surgical challenge presented by these patients is amplified not only by the co-existence of restrictive and often surgically unfavourable MV morphology with other obstructive lesions at an early age, but also primarily because of their small size and their immature—and enormously fragile—leaflet tissues. The optimal surgical approach to patients with Shone's anomaly is governed by the morphology of each obstructive lesion. We favoured an aggressive approach to MV pathology, i.e. we performed MV repair on all patients with transmitral gradient >5 mmHg, which were evident in this population.

The majority of patients in this study presented with coarctation of the aorta as the predominant obstructive lesion, similar to the findings of other investigators [1–4]. The concomitant MV lesions were mostly haemodynamically and clinically significant at the time of coarctation repair.

Mitrail involvement seen in this series does not entirely encompass the features of Shone's anomaly as originally described [1]. The MV morphology conforms to the range of congenital MV anomalies reported by the pathological studies of Ruckman and...
Van Praagh and that of others, as well as by Shone himself [1, 10, 13, 14]. Although mitral ring is present in all 45 patients, parachute valve comprises only 17.7% of the MV lesions. The chordae were short and thickened and there is considerable variability in the position of papillary muscles. Mostly, the papillary muscle was centrally located. We did not see both papillary muscles with chordae tendinae attached to only one, as reported by Brown and colleagues [4]. In this instance, this malformation is what Oesthoek and colleagues called ‘parachute-like asymmetric MV’ when they observed that, among their 129 autopsy specimens, only one had a true parachute MV and 29 had parachute-like valves [13]. We believe that the parachute valves, associated with significant subaortic stenosis, were amenable to repair because the main obstructive mitral element was subvalvular. These patients presented with coarctation of the aorta and severe subaortic obstruction were effectively treated by septal myectomy or resection of the subaortic membrane, when present. Outcomes in this subgroup are related to the degree to which MS can be relieved. In these cases, adequate repair becomes a balance between residual stenosis and induced insufficiency, which could be precisely assessed with intraoperative TEE.

Mitral ring, a characteristic feature, is the most prevalent cause of MS in this disease [6, 14]. Its recurrence, though rare, has been described [6, 15, 16]. Our experience in 12 patients who underwent repeat resection of mitral ring may not be absolutely described as recurrence, because during the repeat resection we found that the rings were mostly of membranous type, completely adherent to the MV leaflet, which may have been difficult to dredge up during the first intervention, owing to absence of a clear line of distinction from the leaflet to which the ring was attached, so that safe dissection was doubtful. We might possibly have missed this membranous part of that fibrous ring, having been overly cautious not to damage the leaflet. The operative experience of Toscano and colleagues was excellent, without any recurrence in their series of 25 patients, with reinterventions done for residual MS from abnormal subvalvular apparatus or valvular hypoplasia [17].

The other MV lesions in our series were mostly the typical congenital MS (Type I) seen in 53% of patients and hypoplastic MV (Type II)—the miniature of a normal MV—seen in 11% in this series. We found this form of MS to be associated with severe LVOT abnormalities in all cases.

The typical congenital MS was easy to repair. Simple commissurotomy, chordal division and splitting the papillary muscles created an effective orifice area, providing relief of the stenosis. We did not see the need to touch the thickened and rolled leaflets and left them alone.

Determining the approach to the hypoplastic MV is equally challenging. It was a dilemma to be faced with a minuscule leaflet and subvalvular apparatus, in addition to the extremely delicate and underdeveloped—if not insufficient—tissues, particularly in infants. Repair is the only available option. With meticulous inspection, especially of the subvalvular apparatus, we were convinced that conscientious division of chordae tendinae—to provide adequate interchordal spaces—and scrupulous fractionation of the papillary muscles—to provide a fairly adequate functional size and length—could provide a functional MV, albeit not long-lasting. This delays the need for valve replacement, with all its drawbacks in the face of the complete unavailability of a prosthesis suitable for this age group. Even when the primary repair result is not optimal, time is gained for repeated repair until a definitive, adult-size prosthesis can be implanted. Because repair is often temporary and/or ineffective, MV replacement is undertaken as a last option when repair fails [18]. We agreed with several reports [3, 4, 6, 19, 20] including some of ours [12, 21, 22] which reported that long-term results of valve repair are superior to replacement in patients with congenital MV anomalies.

The use of modified surgical repair techniques, tailored according to the presenting MV morphology, and the routine use of intraoperative echocardiography made it possible for us to perform repairs with confidence. Brauner and colleagues reported that the failed MV repair procedures in their series were performed before the availability of TEE and hence it is assumed that this outcome is preventable [3]. We are fortunate to have employed this diagnostic modality since its introduction.

Because of the presence of multilevel left ventricular inflow and outflow tract obstructive lesions, decision-making regarding single-ventricle vs biventricular repair in infants is crucial and failed valve repair can lead to pulmonary hypertension and eventual removal of a single ventricle pathway. In this series, the decision to carry out a biventricular repair was straightforward, since there were no patients with hypoplastic-, small- or even borderline left ventricle; hence this has not been the main concern. Likewise, whether the MV should be repaired or replaced has been a matter of controversy. Since the MV lesion is not isolated, there has not been any concern about the inadequate loading of the left ventricle, which creates an impedance to left ventricular ejection, decreasing cardiac output and inability to sustain postoperative haemodynamics. Creation of a widely patent and competent left ventricular inflow, which is the MV, leads to normalization of postoperative pulmonary pressures and adequate ventricular workload. The results of repair approaches used in this group are encouraging and we believe that this group gained maximally from repair.

The single-stage operative approach did not prove to have a significant positive effect on long-term outcome in these patients, even in terms of reoperation, since relief of MS unmasked any existing left ventricular inflow tract lesions. Similar to the report of Serraf and colleagues, we found no difference in reoperation rate between those who underwent single- or multi-staged approaches [6]. However, because of the complex nature of the disease, those patients undergoing single-stage repair are expected to have a high incidence of reintervention on the mitral or aortic valve or the aortic arch [23].

We did not aim for an anatomical restoration of the MV; rather we aimed for restoration of functional competency of whatever valve apparatus is involved.

We are not able to compare the different types of MV repair to find out whether one is better than another, because these techniques have been modified with the passage of time: hence none could be recognized as an independent risk factor. The reoperation rate for every age group was varied and different factors were involved, such as the type of lesions the patient first had. Our results have shown that the risk factors determining outcome are largely based on the type of MS (Table 5).

Whatever the type of left ventricular inflow tract lesion, and however it is presented, we believe that, even in infants and very young children, an aggressive repair approach to MV surgery is satisfactory.

In this series, repeat MV repair was mostly performed in the 1–5 year age group because of residual mitral dysfunction, which could have been greatly intensified when the other left ventricular outflow tract obstructive lesions became pronounced following the initial relief of MS.
Left ventricular outflow tract obstructive lesions in this series have been subvalvular, valvular and supravalvular. Subaortic stenotic lesions are rarely present at birth and are usually progressive, albeit at a variable rate [24]. We have resected discrete membranous lesions, which eliminated the obstruction without any recurrence. Ventricular septal myectomy was performed to eliminate the long segments of fibromuscular tunnel lesion that represent a less common but more severe form of subaortic stenosis. Residual postoperative gradients were present and we have a 52.1% recurrence rate. We found that a Konno-Rastan aortoventriculoplasty effectively eliminated subaortic and aortic valvular obstruction in three patients, without recurrence. In patients with recurrent and severe aortic valve stenosis that was deemed no longer amenable to repair, the Ross procedure provided excellent valve competence. This procedure may eventually emerge as the best option for definitive repair of LVOT obstructive lesions in children with Shone’s anomaly.

In all these MV lesions in Shone’s anomaly, successful repair allowed for valve growth relative to body growth without the need of anticoagulation [25]. It is important to note that all patients were in modified Ross/NYHA functional Class I with normal growth and development.

CONCLUSION

It is apparent that the diversity and complex nature of left ventricular inflow and outflow tract obstructions in children with Shone’s anomaly make it necessary to tailor the surgical interventions and management to each individual patient. Despite high surgical risk, late outcome in the majority is favourable.

In this study, we have demonstrated that an aggressive functional repair approach to the MV and relief of the left ventricular outflow tract obstruction led to long-term event-free survival in these children. Repair strategy has been challenging, especially in hypoplastic valves in infants. Outcomes in this population are related to the degree to which MS can be relieved.

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APPENDIX. CONFERENCE DISCUSSION

Dr E. Belli (Le Plessis-Robinson, France): You present a rather favourable outcome associated with this subgroup of patients which has a bad reputation. Shone’s anomaly associates multilevel left heart obstruction in the presence of a supramitral ring, which is in fact almost always connected to the valvular tissue, and it may be better to call it mitral ring. After reading your manuscript, I once again had the impression that the presence of a mitral
ring can be considered an anatomical factor associated with a more favourable outcome when we are talking about multilevel left heart obstructions, so-called Shone or Shone-like anomalies.

I have one additional comment. During the video, you were talking about commissurotomy but one would think that those were all created without commissure. They are not normal valves which are stenotic. So we cannot call it commissurotomy, maybe commissure creation or something like that. On the other hand, reoperation after conservative mitral valve surgery should be considered as a common event and not a complication or a failure of the first procedure.

Dr Delmo Walter: Yes, I acknowledge that.

Dr Belli: You focused on the mitral valve, but my question is more on the Shone’s complex. In the light of your experience, would you prefer to address all obstructive lesions without looking at whether they are significant or not, or will you remain more conservative and treat only the obstruction which justifies the surgical procedure in Shone’s anomaly?

Dr Delmo Walter: I think that all the obstructive lesions should be corrected, the left ventricular inflow obstruction and the left ventricular outflow obstruction, because if you correct only the left ventricular outflow, then you have an inadequate loading of the left ventricle and there will be impedance to left ventricular ejection, which will decrease your cardiac output and, to sustain postoperative haemodynamics, you have to have a widely patent and competent left ventricular inflow, which is the mitral valve. So I think both should be corrected, if possible, when you have diagnosed Shone’s anomaly. But most of the time, as in our previous experience, some lesions were corrected first before we were able to diagnose that the patient had Shone’s anomaly, because some lesions, like coarctation of the aorta, mask the left ventricular inflow lesions. I think it’s better to do it, of course, if you are able to diagnose it, in a single-stage repair.

Dr Belli: Let’s give an example: a 3-week old patient with coarctation and a mitral gradient mean of 7.5 mmHg and you have to do a procedure. There is no evidence of supramitral ring. That can happen very often, as you know. The ring can be an intraoperative finding, which satisfies the surgeon when he sees it. So what would you do with this 3-week-old child with some MS and coarctation?

Dr Delmo Walter: But 7.5 is already a severe gradient in children, which tells us that a significant stenosis is present. It’s a severe gradient, so I will do mitral valve repair at the same time as the repair of coarctation of the aorta. That should improve the outcome for such a patient.

Dr Belli: The last question, do you have selection criteria for the patients with borderline left ventricle, eventually suitable for univentricular repair versus biventricular? This is discussion which is missing in your study. What is your policy on this type of patient?

Dr Delmo Walter: In this series of Shone’s anomaly in 45 patients, we didn’t see any borderline left ventricle. We don’t have any experience with borderline left ventricle in Shone’s anomaly.