Surgical strategies and outcomes in patients with supra-annular mitral ring: a single-institution experience

John W. Brown*, Mark Ruzmetov, Mark D. Rodefeld, Mark W. Turrentine
Section of Cardiothoracic Surgery, Indiana University School of Medicine, Indianapolis, IN, USA

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Abstract

Objective: Supra-annular mitral ring (SMR) is a rare developmental abnormality of the supra-valvular area of the mitral valve, which produces a variable degree of obstruction to left ventricular (LV) filling. Management of SMR remains an important therapeutic challenge. SMR constitutes a small but inadequately described subset that has a relatively good outcome with appropriate management. Methods: Between 1983 and 2009, 27 patients with SMR underwent surgical treatment directed to the stenosing SMR. The demographic and clinical features, diagnostic modalities and the surgical management were studied retrospectively. Of these, 92% of patients had associated SMR with other congenital heart disease. The membranous component of the SMR was completely excised in all patients. Results: There were no early deaths; however, there were five late deaths. The 20-year survival rate was 82%. By univariate analysis, young age (<1 year; p = 0.02) and Shone’s anomaly (p < 0.001) were risk factors for late mortality. The median follow-up duration was 6.5 years (range: 3 months to 20 years). Three patients required re-operation for recurrent LV outflow tract (OT) obstruction (all with Shone’s anomaly), resection of sub-aortic membrane (n = 2) and the Ross–Konno procedure (n = 1). Additionally, one patient required MV replacement. Freedom from re-operation for LV outflow and/or inflow obstruction was 88% at 20 years. At latest follow-up, only two patients had peak trans-mitral gradients >5 mm Hg related to supra-valvular obstruction. Conclusion: SMR is an uncommon lesion, which can be safely and effectively managed by surgical resection. While co-existent mitral valvular lesions are usual, they are not typically severe in degree, as evidenced by the generally benign postoperative outcome following resection of SMR.

Keywords: Congenital heart disease; Paediatric; Re-operation; Survival

1. Introduction

Supra-annular mitral ring (SMR) is a rare developmental abnormality of the supra-valvular area of the mitral valve (MV) producing obstruction to left ventricular (LV) filling [1–3]. SMR should be differentiated from cor triatriatum sinister, since the embryological origin, morphology and the surgical implications are different in these two malformations. Supra-annular mitral ridge is an extremely rare entity which may mimic classical SMR, but is one anomaly which should be distinguished by the general rule that surgical resection is contraindicated. Although cases of isolated SMR have been reported sporadically [1,4,5], SMR is more commonly associated with other obstructive lesions in the left heart, such as valvular stenosis of the MV, parachute MV or mitral arcade, sub-aortic membrane or fibromuscular tunnel, aortic valve stenosis with or without bicuspid aortic valve, transverse arch hypoplasia or discrete coarctation of aorta [6,7], and less commonly with anomalies in the right heart, such as pulmonary stenosis or tetralogy of Fallot [8]. Classically, SMR has been described as one of the four specific features of Shone’s complex [6].

Increasing awareness of this condition as well as refinements in diagnostic techniques will allow more frequent diagnosis of this condition [7]. The identification of this subset of patients is important, however, because of the relatively better prognosis with appropriate management. There is paucity of information in the literature about the pathology, diagnosis and surgical management of this entity, and this retrospective analysis of a series of 27 patients, managed at our institution, attempts to address some of these issues.

2. Materials and methods

Between January 1983 and May 2009, 27 patients were operated upon in our institution for SMR causing significant mitral stenosis (MS), either in isolation or in association with other cardiac anomalies. The mean age of the patients was 5123, USA. Tel.: +1 317 274 7150; fax: +1 317 274 2940.

* Corresponding author. Address: Section of Cardiothoracic Surgery, Indiana University School of Medicine, 545 Barnhill Dr., EH 215, Indianapolis, IN 46202-5123, USA. Tel.: +1 317 274 7150; fax: +1 317 274 2940.
E-mail address: jobrown@iupui.edu (J.W. Brown).

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of 8 mm Hg or a postoperative mean diastolic gradient during follow-up greater than a mean diastolic gradient of 8 mm Hg across the mitral apparatus was considered significant. Peak velocity of the tricuspid regurgitant jet was measured to calculate systolic right ventricular pressure. Associated cardiac defects were methodically sought. Cardiac catheterisation data were reviewed, and the mean diastolic trans-mitral gradient and pulmonary artery pressure were recorded. Angiograms were reviewed for other heart defects. A favourable outcome was defined as lack of progression of the stenosis during follow-up greater than a mean diastolic gradient of 8 mm Hg or a postoperative mean diastolic gradient of ≤5 mm Hg [3].

Anatomical evaluation was done in all patients by echocardiography or cardiac catheterisation and confirmed at operation. All but two of the 14 patients with Shone’s complex had an abnormal MV and presented with at least two other LV outflow tract (LVOT) obstructive lesions. None of the 13 patients without Shone’s complex had MV anomalies. Not all of the left heart obstructive lesions were severe enough to require intervention at the time of this report.

2.1. MV assessment

The MV was systematically assessed in all the patients using trans-thoracic echocardiography. The leaflets, the chordae and the papillary muscles were viewed and the size of the mitral annulus was measured, and the z-score calculated. The mean trans-mitral gradients ranged from 10 to 30 mm Hg (mean: 15.2 ± 5 mm Hg). The patient with the isolated SMR had the largest gradient of 30 mm Hg.

In all patients, the SMR was circumferential and attached to the left atrium slightly above the mitral annulus (supra-annular SMR).

2.2. Associated defects

Associated cardiac defects were diagnosed in 25 patients; the remaining two patients had an isolated SMR. All associated cardiac anomalies are shown in Table 1. Shone’s anomaly was diagnosed in 14 of the 27 patients (52%). In 11 of the remaining patients, other cardiac defects included tetralogy of Fallot or double-outlet right ventricle (DORV; n = 5), partial atrioventricular communication (n = 3), perimembranous ventricular septal defect (n = 2) and total anomalous pulmonary venous return (n = 1). A bicuspid aortic valve was noted in nine (33%), a ventricular septal defect was noted in 14 (52%) and atrial septal defect in 5 of the 27 patients (19%). Non-cardiac anomalies were noted in 4 of the 27 patients (15%): pyloric stenosis (n = 1), biliary atresia (n = 1), cleft lip with palate (n = 1) and Pierre–Robin syndrome (n = 1).

In the group with Shone’s anomaly (n = 14), the most prevalent left-sided obstructive lesion was coarctation of the aorta in 11 patients (11/14, 79%), and bicuspid aortic valve which was present in nine of the 14 patients (64%) with associated valvular aortic stenosis, noted in two of these patients (3/14, 21%). A mildly hypoplastic transverse aortic arch (diameter: <4 mm) was present in five patients (5/14, 36%), diffuse or discrete sub-aortic stenosis due to a fibromuscular tunnel or membrane in eight patients (8/14, 57%) and supra-valvular aortic obstruction in one patient (1/14, 7%). In the 14 patients with Shone’s anomaly, the MV anomalies were defined according to the classification of Ruckman and Van Praagh [9]. In seven patients (7/14; 50%), the morphology of the MV included mitral stenosis, fused chordae, single papillary muscle and parachute MV apparatus. Congenital mitral stenosis consisting of thickened leaflets, partial or complete obliteration of interchordal spaces and a variable reduction in interpapillary distance, were present in five patients (5/14; 36%), and two patients had normal MVs.

Table 1

<table>
<thead>
<tr>
<th>Associated anomalies (25 patients)</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect</td>
<td>14 (52%)</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>12 (45%)</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>9 (33%)</td>
</tr>
<tr>
<td>Subaortic stenosis</td>
<td>7 (26%)</td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>5 (19%)</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>5 (19%)</td>
</tr>
<tr>
<td>Mitral stenosis</td>
<td>4 (15%)</td>
</tr>
<tr>
<td>Partial atrioventricular communication</td>
<td>3 (11%)</td>
</tr>
<tr>
<td>Bicuspid aortic stenosis</td>
<td>3 (11%)</td>
</tr>
<tr>
<td>Hypoplastic transverse aortic arch</td>
<td>2 (7%)</td>
</tr>
<tr>
<td>Total anomalous pulmonary veins return</td>
<td>1 (4%)</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
</tr>
</tbody>
</table>

![Fig. 1. The technique of supra-annular mitral ring resection.](https://academic.oup.com/ejcts/article-abstract/38/5/556/416731)
2.3. Surgical technique

After standard midline sternotomy, bicaval cannulation, initiation of standard cardiopulmonary bypass, topical cooling, antegrade cold-blood potassium cardioplegia and systemic hypothermia to 28 °C, the MV was approached through the intra-atrial septum (n = 7; in patients with concomitant right-sided cardiac anomalies) or left atrium (n = 10). Morphological abnormalities of the MV approach were systematically assessed (Fig. 1a). The full extent of the SMR was delineated. The initial incision is made anteriorly (above the anterior leaflet of the MV and directed towards the aortic—mitral fibrous continuity) and extended to the junction of the membrane and the MV annulus (Fig. 1b). This initial incision is directed away from the possible location of the left circumflex coronary artery. Care is taken to protect the MV during the resection (Fig. 1c).

In all patients, MV leaflets were then thoroughly inspected for any perforations, and competency of the valve was assessed by instillation of saline into the LV cavity. The mitral annulus was also sized with appropriately sized dilators. The associated cardiac anomalies were then corrected if they were clinically significant. All patients had intra-operative trans-oesophageal echocardiography to assess the adequacy of the surgical repair.

2.4. Statistical analysis

Measured and calculated data are expressed as mean ± standard deviation (SD). Comparison between two groups was performed using the unpaired t-test. The Kaplan—Meier product-limit method was used for analysis of survival and freedom from re-operation. Multiple regression analysis was performed as conditional backward stepwise proportional hazards regression. In the analysis of risk factors for mortality, freedom from re-operation variables with significance levels of 0.1 in univariate analysis were submitted to a multivariate logistic regression model. Early mortality was defined as death during initial hospitalisation or within 30 days of operation. Any deaths later than that were defined as late mortality. A p-value of <0.05 was considered significant. Specific statistical software SPSS for Windows version 10 (SPSS Inc., Chicago, IL, USA) was used for data analysis.

3. Results

3.1. Mortality

There were no early deaths; however, there were five late deaths. The causes of late deaths are shown in Table 2. The 20-year survival rate was 82%. Overall survival was 89% at 1 year, 85% at 5 years and 82% at 10 and 20 years (Fig. 2). By univariate analysis, young age (<1 year; p = 0.02) and Shone’s anomaly (p < 0.001) were risk factors for mortality. Of the late surviving patients (n = 22), two were in the New York Heart Association (NYHA) class II, and the remaining 20 are in class I.

3.2. Follow-up

The median follow-up duration was 6.5 years (range: 3 months to 20 years). Three patients required re-operation for recurrent LVOT obstruction (all with Shone’s anomaly): resection of sub-aortic membrane (n = 2) and Ross—Konno procedure (n = 1). Additionally, one of those patients required MV replacement. Freedom from re-operation for LV outflow or inflow obstruction was 88% at 20 years. There are no significant risk factors for re-operation. At latest follow-up, only two patients had peak trans-mitral gradient >5 mm Hg related to supra-valvular obstruction.

### Table 2

<table>
<thead>
<tr>
<th>Cause</th>
<th>No. of patients</th>
<th>Death months postoperatively</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low cardiac output</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>DIC</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Bacterial endocarditis</td>
<td>1</td>
<td>96</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>1</td>
<td>2.5</td>
</tr>
<tr>
<td>Sudden/unknown</td>
<td>1</td>
<td>24</td>
</tr>
<tr>
<td>Total</td>
<td>5</td>
<td></td>
</tr>
</tbody>
</table>

Fig. 2. Kaplan—Meier curve of survival in patients with supra-annular mitral ring.

### Table 3

<table>
<thead>
<tr>
<th>Year</th>
<th>Number of patients</th>
<th>Age (mean)</th>
<th>Type</th>
<th>Follow-up time (mean)</th>
<th>Mortality</th>
<th>Redo</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tulloh and colleagues [16]</td>
<td>1995</td>
<td>23</td>
<td>0.7 years</td>
<td>Supra all</td>
<td>58 months</td>
<td>6 (26%)</td>
</tr>
<tr>
<td>Konstantinov and colleagues [5]</td>
<td>2004</td>
<td>13</td>
<td>NA</td>
<td>Supra all</td>
<td>55 months</td>
<td>0</td>
</tr>
<tr>
<td>Collison and colleagues [7]</td>
<td>2006</td>
<td>15</td>
<td>27 months</td>
<td>Supra all</td>
<td>30 months</td>
<td>1 (7%)</td>
</tr>
<tr>
<td>Toscano and colleagues [3]</td>
<td>2009</td>
<td>13</td>
<td>36 months</td>
<td>Supra (5); intra (8)</td>
<td>90 months</td>
<td>0</td>
</tr>
<tr>
<td>Brown and colleagues [current]</td>
<td>2009</td>
<td>27</td>
<td>3.9 years</td>
<td>Supra all</td>
<td>6.5 years</td>
<td>5 (19%)</td>
</tr>
</tbody>
</table>
4. Discussion

SMR, also known as supra-valvular mitral stenosis, supra-valvular ring of the left atrium and supra-valvular stenosing ring, was first described by Fisher in 1904, when the ridge of connective tissue above the MV was likened to the diaphragm of the microscope [10]. Classically, SMR was described as one of the four specific features of Shone’s complex [6]. In his original description, Shone reported eight patients who had multiple obstructive lesions in the left heart. Although only two patients exhibited all four features, SMR was a common finding in all eight patients. Thus, SMR has been regarded as an essential element of Shone’s complex. It is widely recognised, however, that only a small proportion of cases of SMR is associated with classical Shone’s complex. In our experience, Shone’s anomaly was seen in 52% (14 of 27).

SMR should be differentiated from cor triatriatum sinister, since the embryological origin, morphology and the surgical implications are different in these two malformations. The embryological origin of SMR is unclear, but it has been postulated that this condition results from incomplete division of the endocardial cushion tissue and is fibrous [4]. SMR is differentiated from cor triatriatum, which is believed to be a result of incomplete absorption of primary pulmonary vein during the 5th embryonic week and consists of fibromuscular tissue [11]. In cor triatriatum, the communication of proximal and distal chambers is usually a single hole in the membrane, which is further above the MV and proximal to the left atrial auricle.

The first surgical correction of SMR was described by Lynch and colleagues in 1962 [12]. The study showed that surgical correction of this anomaly can be performed successfully, leading to excellent late clinical results. This finding has been confirmed by our study. Four of our deaths were in patients with Shone’s anomaly, which itself is a complex disease process with a guarded prognosis.

The cardiac anomalies associated with SMR have not been well described. From our study, we propose that the associated cardiac anomalies can be grouped broadly into two categories: SMR associated with ventricular septal defect (VSD; including tetralogy of Fallot) and SMR associated with LVOT pathologies, especially sub-aortic membrane, bicuspid aortic valve and coarctation of aorta (Shone’s anomaly). In the first scenario, this is important in the context of patients with VSD and turbulence across the MV in which an SMR needs to be ruled out. In the second scenario, in patients with multi-level left heart obstructions, an SMR should be excluded to prevent residual defects, as has been reported previously [13].

Isolated occurrence of the SMR was first reported by Chung and colleagues [14]. In our series, only two patients had isolated SMR (8%). Yaidesevar and colleagues [15] identified associated rheumatic MV abnormalities in three cases and there exists a possibility for the ridge to develop as a post-inflammatory phenomenon.

Previous studies suggested that late outcomes are excellent, and the need for re-intervention is uncommon [1,5,7]. Our operative experience with SMR compares favourably with other series [1,3,5,7] (Table 3). Furthermore, the SMR did not recur after surgery. In all of the cases of SMR, ring resection led to good-to-excellent long-term results. Toscano and colleagues [3] found that predictors of poor surgical outcome were mitral annulus hypoplasia at birth and anomalous sub-valvular apparatus. Tulloh and colleagues [16] reported that age <18 months was also associated with a poor surgical outcome. We did find that young age (<1 year; p = 0.02) and Shone’s anomaly (p < 0.001) were risk factors for mortality.

5. Conclusion

SMR is a rare developmental abnormality, which restricts LV filling and causes pulmonary hypertension. It is rarely seen as an isolated anomaly and is more usually associated with other left heart obstructive lesions (Shone’s anomaly) in up to 50% or in VSD or DORV in another 40%. Surgical recurrence is the treatment of choice along with treatment of the associated lesions. The risk factors for early mortality include age <1 year and/or associated Shone’s complex.

References

Appendix A. Conference discussion

Dr Y. d’Udekem (Melbourne, Australia): As you mentioned, supramitral ring is rare and still a poorly delineated lesion. There are fewer than 150 cases described in the literature, and its description in these different reports varies greatly.

I take as an example the fact that the association with Shone’s anomaly varies between 15 and 80%; therefore, any information on the disease is very valuable.

My first question concerns the patient population. Are you aware of any additional patients who may have been diagnosed with a disease, with a supramitral ring, but were not offered any surgery?

Dr Brown: No. If they had a gradient — I suppose they could have had a gradient smaller than 8 mm, and a decision was made not to operate on them. But all the patients in our retrospective series that had this diagnosis and had a gradient greater than 8 were included.

Dr d’Udekem: My second question concerns the rate of recurrence. All the reports in the literature insist on the fact that the results are very good, and there is no recurrence. But I think that the reason why they insist so much is because there is actually recurrence of the disease, and we suffered some recurrence in Melbourne.

So you virtually encountered no recurrence which is a bit different to our experience. Do you think this is related to a more radical surgical technique, or did you observe some morphologic recurrence, a beginning of a new membrane, which did not get a re-operation because the gradients through the mitral valve were acceptable?

Is that the reason why you specified a gradient of 8 mm Hg as being satisfactory rather than, for example, 5 mm Hg?

Dr Brown: In all of our postoperative patients, the gradient was less than 5 mm Hg. Preoperatively, 8 mm Hg was what we considered to be significant to warrant the operation, but in all of our patients, the late postoperative gradient was 5.

And for reasons that are not terribly clear to me, we haven’t seen a significant recurrence. We are fairly radical when we resect these membranes. And it’s hard to believe that we have not had recurrence, but that’s what our data shows.

Now, you might ask if the 5 patients who had late deaths had some evidence of recurrence? We were not, however, able to appreciate a significant gradient in any patient in our analysis.

Dr A. Corno (Liverpool, UK): What did you learn from this study? Is there anything in the decision-making management that you will do differently in the future that you can suggest to us?

Dr Brown: The most important aspect of this lesion to the surgeon is a good preoperative assessment of the mitral inflow in all patients. In some patients with partial AV septal defect or in patients with isolated VSD, you might not open the left atrium to look for a supra-annular mitral ring unless the pre-op echo demonstrated a significant restriction to mitral valve inflow. I give credit to our cardiologists who tell us that there is something in addition to the VSD or partial canal or tetralogy that we need to look at, and so we pay attention to this area in those patients.

In Shone’s complex, you’re always looking at the mitral inflow and looking at the mitral valve, so it’s hard to miss in the Shone’s complex group. But in the non-Shone’s group, you can easily miss it if you’re not looking for it.

The lesson to be learned is to be sure you assess the mitral inflow in all patients before the operation; otherwise, you may overlook a ring that will come back to haunt the patient and you, and you’ll have to reoperate on that patient.

Dr G. Stellin (Padova, Italy): In your experience, there is less than 50% incidence of underlying mitral valve dysplasia underneath the membrane. My question is: Have you included those cases with a very complex mitral valve dysplasia in association with a very small membrane on top of the mitral valve annulus?

My second question is, have you excluded, in this experience the so-called cor triatriatum sinister?

Dr Brown: Yes. We did eliminate the cor triatriatum patients, of course. And, yes, in the Shone’s complex group, 12 of the 14 with Shone’s complex had abnormal mitral valves. And so there were only 2 of the Shone’s that didn’t have an abnormal mitral valve.

So, yes, if we dictated that we resected the membrane, we included those patients in the analysis.

Dr Stellin: Okay.