Surgical management of complex and tunnel-like subaortic stenosis

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Received 6 September 1999; received in revised form 11 February 2000; accepted 29 February 2000

Abstract

Background: Relief of primary or secondary subaortic stenosis (SAS) remains a surgical challenge. Heart block, aortic valve regurgitation and recurrent obstruction have been persistent problems. Methods: Forty six patients who underwent surgery for complex and tunnel-like SAS between January 1990 and November 1998 were reviewed. In 45 of the 46 patients SAS developed following repair of a primary congenital heart defect and only one patient presented with de novo tunnel-like SAS. Fifteen of the 45 patients had undergone repair of double-outlet right ventricle (DORV) and the remaining 30 had undergone repair of a variety of defects. The median age at the time of surgery was 5 years. The modified Konno procedure was performed in 15 patients, Konno procedure in three, Ross–Konno procedure in two and resection of the conal septum in 12 patients. Five patients with DORV underwent replacement of the intraventricular baffle and two patients underwent an aortic valve-preserving procedure in conjunction with mitral valve replacement. Results: There were no deaths. None of the patients had an exacerbation of aortic regurgitation and none developed complete heart block. The median follow-up was 3 years (range 1 month–8.5 years). Two patients developed recurrent SAS defined as a gradient of 40 mmHg or greater diagnosed by transthoracic echocardiography. Freedom from SAS at 1, 3 and 5 years was 100, 94 and 86%, respectively. Conclusions: We favor the modified Konno procedure and conal resection to the Konno or the Ross procedure, since insertion of a prosthetic valve or homograft is avoided and aortic valve function is preserved. Excellent relief of tunnel-like SAS can be achieved without damage to the conduction tissue. © 2000 Published by Elsevier Science B.V.

Keywords: Subaortic stenosis; Modified Konno

1. Introduction

The surgical management of left ventricular outflow tract obstruction remains a challenge, particularly ensuring complete relief of the obstruction, preserving the aortic valve and avoiding heart block and recurrence of the obstruction. Subaortic stenosis (SAS) spans a spectrum of anomalies ranging from a simple membrane to tunnel-like obstruction. In the membranous type, excision of the fibrous tissue with or without myectomy often results in satisfactory relief of the obstruction [1]. However, simple excision of the fibrous ridge plus myectomy is often inadequate in the treatment of tunnel-like SAS [2,3]. Tunnel-like SAS can occur in isolation or it may be associated with ventricular septal defect (VSD), partial or complete atrioventricular canal defects, interrupted aortic arch, and coarctation of the aorta [4–6]. It may also manifest itself following repair of these conditions or after repair of double-outlet right ventricle (DORV) [7] or after the arterial switch operation for transposition of the great arteries.

In this study we have differentiated complex and tunnel-like SAS from membranous SAS by operative procedure. When the patient has required more than resection of a subaortic membrane with or without simple myectomy, they have been defined as having complex and tunnel-like SAS. The aim of this study was to examine the outcome in these patients.

2. Methods

A retrospective analysis of patients who underwent surgery for tunnel-like SAS between January 1990 and November 1998 was carried out. A total of 46 patients were identified. Of the 46 patients, 15 had undergone repair of DORV (Table 1). The primary congenital heart defects of the remaining 30 patients who had had previous surgery and subsequently developed SAS are shown in Table 2. Only one patient presented with de novo tunnel-like stenosis. The median age at the time of the first operation was 3 months (range 2 days–9 years) and the median age at the time of surgery for SAS was 5 years (range, 3-10 years). The median interval between these two operations was 4 years (range 1.5–8.5 years).
2.1. Patient characteristics

Twelve patients had undergone biventricular repair and three had undergone pulmonary artery banding for repair of DORV (Table 1). Two of the patients who had had pulmonary artery banding subsequently underwent the Fontan procedure and one patient had a bidirectional Glenn procedure performed at the time of conal resection for SAS. Of the 12 patients with biventricular repair, four had undergone the arterial switch operation. Eight patients had undergone intraventricular repair of DORV, one with associated right ventricular outflow tract obstruction which required pericardial patch augmentation. One patient had an associated supravalve mitral ring which required excision and one underwent repair of coarctation of the aorta.

Eight patients had undergone repair of type B interrupted aortic arch in the neonatal period. Five patients had undergone resection of subaortic membrane and had subsequently developed tunnel-like obstruction. The remainder had had primum atrial septal defect, common atrioventricular canal defect, coarctation of the aorta and VSD repaired, and one patient with the diagnosis of transposition of the great arteries IVSD/pulmonary stenosis had had a Rastelli procedure performed (Table 2).

Five patients had variants of Shone’s syndrome. Two underwent excision of supravalve mitral ring and repair of associated coarctation. One patient had a primum ASD with severe SAS. She was not referred for surgery until 9 years of age when she underwent replacement of the mitral valve with a mechanical prosthesis and closure of primum ASD with a pericardial patch. She was subsequently found to have a 65-mmHg gradient due to a tunnel extending from the aortic valve to the level of the mitral prosthesis. The latter two patients have previously been reported in detail with the method of repair [8].

2.2. Relief of subaortic stenosis: surgical considerations

2.2.1. Modified Konno procedure

A total of 15 patients underwent the modified preservation of the aortic valve. An oblique incision over the infundibulum of the right ventricle is made. This is extended to the level of the aortic annulus and just to the left extending towards the left/right commissure and a right-angle instrument is passed through the aortic valve to guide an incision into the infundibular septum. The incision is then enlarged in a leftward direction below the pulmonary valve. Extra

Table 1
Morphologic and surgical data in patients with double-outlet right ventricle

<table>
<thead>
<tr>
<th>Case no.</th>
<th>VSD</th>
<th>Associated lesions</th>
<th>Primary repair</th>
<th>Repair of SAS</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Subaortic</td>
<td>Valv. and subvalv. PS; straddling MV</td>
<td>IV repair</td>
<td>Excision of aneurysmal tissue of MV</td>
</tr>
<tr>
<td>2</td>
<td>Subaortic</td>
<td>–</td>
<td>IV repair</td>
<td>Conal excision and membranous resection</td>
</tr>
<tr>
<td>3</td>
<td>Subaortic</td>
<td>Subvalvular PS</td>
<td>IV repair; patch to RVOT</td>
<td>Baffle reconstruction</td>
</tr>
<tr>
<td>4</td>
<td>Subpulm</td>
<td>–</td>
<td>ASO</td>
<td>Resection of membr. tissue</td>
</tr>
<tr>
<td>5</td>
<td>Subaortic</td>
<td>Subvalvular PS</td>
<td>IV repair</td>
<td>Baffle reconstruction</td>
</tr>
<tr>
<td>6</td>
<td>Non-com</td>
<td>Straddling MV</td>
<td>PAB/BDG/Fontan</td>
<td>Conal excision</td>
</tr>
<tr>
<td>7</td>
<td>Subpulm</td>
<td>–</td>
<td>ASO</td>
<td>Baffle reconstruction</td>
</tr>
<tr>
<td>8</td>
<td>Non-com</td>
<td>Subvalvular PS; straddling MV</td>
<td>PAB/Fontan</td>
<td>Conal excision</td>
</tr>
<tr>
<td>9</td>
<td>Subpulm; T-B</td>
<td>–</td>
<td>ASO</td>
<td>Baffle reconstruction</td>
</tr>
<tr>
<td>10</td>
<td>Subpulm</td>
<td>Straddling TV</td>
<td>ASO</td>
<td>Membr. resection, myectomy</td>
</tr>
<tr>
<td>11</td>
<td>Doub-com</td>
<td>–</td>
<td>IV repair</td>
<td>Modified Konno</td>
</tr>
<tr>
<td>12</td>
<td>Subaortic</td>
<td>Supravalve mitral ring</td>
<td>IV repair; excision of ring</td>
<td>Excision of protruding septum</td>
</tr>
<tr>
<td>13</td>
<td>Non-com</td>
<td>Coarctation</td>
<td>IV repair; coarctation repair</td>
<td>Modified Konno</td>
</tr>
<tr>
<td>14</td>
<td>Non-com</td>
<td>–</td>
<td>IV repair</td>
<td>Baffle reconstruction</td>
</tr>
<tr>
<td>15</td>
<td>Non-com</td>
<td>Scimitar syndrome</td>
<td>PAB</td>
<td>BDG/Conal excision</td>
</tr>
</tbody>
</table>

* VSD, ventricular septal defect; SAS, subaortic stenosis; IV repair, intraventricular repair; MV, mitral valve; PS, pulmonary stenosis; RVOT, right ventricular outflow tract; subpulm, subpulmonary VSD; doub-com, doubly committed; non-com, non-committed; ASO, arterial switch operation; TV, tricuspid valve; PAB, pulmonary artery banding.

Table 2
Tunnel-like subaortic stenosis and associated lesions in 30 patients

<table>
<thead>
<tr>
<th>Primary lesion</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>TGA/VSD/PS</td>
<td>1</td>
</tr>
<tr>
<td>Type B, interrupted aortic arch</td>
<td>8</td>
</tr>
<tr>
<td>Shone’s variant</td>
<td>5</td>
</tr>
<tr>
<td>Membranous SAS</td>
<td>5</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>2</td>
</tr>
<tr>
<td>Common atrioventricular canal</td>
<td>2</td>
</tr>
<tr>
<td>TGA/common atrioventricular canal</td>
<td>1</td>
</tr>
<tr>
<td>Primum atrial septal defect</td>
<td>3</td>
</tr>
<tr>
<td>Coarctation/valvular aortic stenosis</td>
<td>3</td>
</tr>
</tbody>
</table>

* TGA, transposition of great arteries.
care is taken about resecting tissue from the inferior edge of the incision to reduce the risk of injury to the His bundle. At this point, any fibrous subaortic tissue is resected. A patch of glutaraldehyde-treated pericardium or Gore-Tex is sutured with interrupted pledgeted sutures to close the created VSD with all the sutures remaining on the right ventricular aspect of the septum. The incision in the free wall of the right ventricular outflow tract is then closed either directly or with a patch of pericardium or Gore-Tex (Figs. 1–3).

2.2.2. Konno procedure

Three patients underwent the Konno procedure with implantation of a prosthetic valve and use of a Dacron patch to enlarge the left ventricular outflow tract [9].

2.2.3. Ross–Konno procedure

Two patients underwent this operation, which is pulmonary autograft root replacement of the aortic root including the aortic valve with reimplantation of the coronary arteries and ventricular septoplasty. The right ventricular outflow tract was reconstructed with an allograft valved conduit. One patient was a 17-year-old who had had two aortic valvotomies and subaortic myectomies performed. The other was a 6-year-old who had undergone balloon valvuloplasty for critical aortic stenosis in the neonatal period and had subsequent surgical valvotomy performed.

2.2.4. Resection of the conal septum

Twelve patients underwent resection of the conal septum. The conal septum was observed as a ridge of muscle projecting into the anterior wall of the left ventricular outflow tract, and was excised as widely as possible without interfering with aortic valve integrity. Six patients had undergone biventricular repair of DORV and two had undergone the Fontan procedure. Three patients had undergone repair of interrupted aortic arch and one had undergone closure of VSD.

2.2.5. Resection of fibromuscular ridge and myectomy

Seven patients underwent this procedure. Two patients had undergone repair of primum atrial septal defect, three patients had undergone repair of common atrioventricular canal defect, one had closure of VSD and repair of hypoplastic aortic arch and coarctation of aorta, and one had undergone repair of coarctation of aorta. All of these patients had extensive fibromuscular ridges.

2.2.6. Removal and reconstruction of the intraventricular baffle

Five patients in the DORV group underwent baffle reconstruction. One further patient had aneurysmal parachute-like tissue arising from the anterior mitral valve leaflet into the subaortic area. He underwent excision of this tissue avoiding damage to the mitral valve.

2.2.7. Aortic valve preserving procedure and mitral valve replacement

This technique was used in the two patients with Shone’s variant. An aortic incision is directed towards the commissure between the left and noncoronary cusps. Then an oblique incision is made in the right atrial wall and is extended across the roof of the right atrium, across the atrial septum.
towards the left atrium meeting the aortic incision. After excision of the mitral valve, the aortic incision is extended across the junction of the non and left coronary cusps to expose the subaortic fossa. A triangular PTFE patch is used to reconstitute the aortic valve and to supplement the mitral annulus. A mitral prosthesis is sutured to the base of this triangle and the rest of the mitral annulus [8].

2.3. Statistical analysis

The Kaplan–Meier product-limit method was used to determine the estimated freedom from restenosis with 95% confidence intervals calculated by Greenwood’s formula.

3. Results

There were no early or late deaths. Follow-up was available in all patients, except for one patient who lives outside the United States. The median follow-up was 3 years (range 1 month–8.5 years). At the latest follow-up, all patients had transthoracic echocardiography performed.

3.1. Recurrent SAS

This was defined when at follow-up a gradient of 40 mmHg or more was measured across the left ventricular outflow tract with transthoracic echocardiography. Freedom from recurrent SAS at 1, 3 and 5 years was 100, 94 and 86%, respectively. Two patients had developed recurrent SAS. One patient who had undergone repair of primum atrial septal defect and subsequently underwent the modified Konno procedure was diagnosed to have a gradient of 50 mmHg by transthoracic echocardiography at 6 years following the modified Konno operation. He underwent cardiac catheterization where this gradient was measured to be 35 mmHg. The second patient had undergone biventricular repair of DORV and subsequently had had resection of subaortic membrane and myectomy performed. At 3 years’ follow-up a gradient of 40 mmHg was detected with transthoracic echocardiography. Cardiac catheterization was not performed in this case.

3.2. Redo surgery following relief of SAS

One patient with severe neonatal idiopathic hypertrophic subaortic stenosis who had undergone the modified Konno procedure at 2.5 months of age developed right ventricular obstruction secondary to severe muscular obstruction at the midbody of the right ventricle 2.5 months later. A large amount of muscle was resected from the midbody of the right ventricle. At 8 years’ follow-up he had no gradient across the left ventricular outflow tract. Another patient who had undergone the Fontan procedure for DORV and surgery to relieve SAS, subsequently underwent orthotopic heart transplantation.

None of the patients developed significant postoperative aortic regurgitation and none developed complete heart block. Two patients developed 2:1 heart block interposed with periods of sinus rhythm and required insertion of a pacemaker. In one patient heart block occurred following reconstruction of the intraventricular baffle and resection of the infundibular septum to relieve SAS and in another following conal septum resection when a concomitant bidirectional Glenn was also performed.

4. Discussion

Ideally, relief of left ventricular obstruction should allow preservation of aortic valve function, allow for growth of the aortic valve and lessen the possibility of a subsequent operation. In this report we have described a cohort of patients with tunnel-like SAS. It may be that in all these patients the hemodynamic factors resulting in SAS are the same, but resulting from different conditions. It is believed that even minimal stenosis in the left ventricular outflow tract can cause turbulent flow causing deposition of fibrous material. Shear stress has been shown to produce significant cellular proliferation [10,11]. Davies et al. [11] have reported that all cells exposed to blood flow experience some degree of shear stress and to adjust to this shear stress, some morphologic changes follow. Cape and colleagues have proposed and supported a four-stage theory for the development of SAS [12]. They propose that associated morphologic abnormalities result in altered shear stress, which triggers a genetic predisposition, leading to proliferation of cells in the outflow tract.

In considering patients for biventricular repair of DORV, the relationship of the aortic annulus to the VSD, the length of the conal septum and its relation to the pulmonary and aortic valves as well as presence of straddling atrioventricular valves must be assessed. The size and type of the VSD and hence the placement of the intraventricular baffle may
predispose the patient to subsequent SAS. None of the patients in the DORV group had SAS prior to their original operation although two required enlargement of the VSD at the time of repair. Serraf et al. [13] reported 30 patients who underwent repair of DORV associated with SAS. The SAS was a result of restrictive VSD in 29 patients and of straddling atrioventricular valve in one patient. Two of the 12 patients in the current series who underwent biventricular repair had straddling atrioventricular valves. These were divided and reimplanted. In a pathological series of DORV, abnormalities of the atrioventricular valves have been demonstrated in up to 30% of the patients [14]. Others have also reported SAS after biventricular repair of DORV [15,16]. Luber et al. [15] reported SAS in two out of 57 patients who underwent repair of DORV. Kirklan and colleagues [16] reported three patients with SAS following repair of DORV. One had subaortic VSD and two of their ten patients with doubly-committed VSD developed SAS. The SAS was a discrete type and not tunnel-like in the two patients with doubly-committed VSD. In the current series, only one patient had a doubly-committed VSD. It may be that due to the lack of the infundibular septum, during repair, sutures must be placed too near the fibromuscular tissue of the aortic valve and this encourages growth of sub aortic fibrous tissue and limits annular growth. Yacoub and Radley-Smith [17] have reported SAS in patients with Taussig–Bing anomaly and side-by-side great arteries. In patients with side-by-side great arteries the conal septum is very well developed and can produce varying degrees of SAS. Rocchini and colleagues [18] have proposed that the development of SAS is related to the decrease in the effective size of the VSD caused either by the baffle or proliferation of fibrous tissue along the baffle, or kinking and shrinkage of the baffle. In this series one patient had removal of fibrous tissue from the baffle and six had refashioning of the baffle.

Previous pulmonary artery banding has been shown to be a risk factor for the development of SAS in patients who are being considered for the Fontan procedure. There is evidence suggesting that pulmonary artery banding may accelerate VSD closure by stimulating muscle hypertrophy [19] or by reducing the ventricular volume causing remodeling of the intraventricular anatomy. In this series, the low incidence of this problem reflects an aggressive prophylactic approach, i.e. early treatment in neonates.

One patient with DORV who had undergone biventricular repair had valvular pulmonary stenosis. It is reported that VSD is larger in patients with single ventricle, transposition of the great arteries, DORV and pulmonary outflow tract obstruction than in those patients with unobstructed pulmonary blood flow. Furthermore, if an obstructive anomaly of the aortic arch is also present, the VSD tends to be smaller than the aortic root [20]. Therefore, one might expect a decrease in the size of the VSD in those with unobstructed pulmonary blood flow causing some degree of SAS. It may be that after the Fontan operation, the SAS becomes unmasked because the preload is reduced and as a result the ventricular size may be diminished. Razzouk et al. [21] have reported SAS in 12 patients following the Fontan operation, with the median interval between the Fontan operation and the diagnosis of SAS of 2.5 years. We believe that once the SAS is diagnosed in patients who have undergone the Fontan procedure, immediate surgical intervention is needed. These patients are at risk of ventricular hypertrophy resulting in altered ventricular compliance which can jeopardize the Fontan physiology. The surgical options for SAS complicating single ventricle have previously been reported [22]. The two patients in this study had resection of the conal septum performed.

Subaortic stenosis is an uncommon problem but known complication associated with common atrioventricular defects. The ‘goose-neck’ type deformity of the left ventricular outflow tract in common atrioventricular canal defect, accessory left atrioventricular valve tissue and chordae, abnormal papillary muscles and aneurysm of the left-sided atrioventricular valves can all contribute to SAS in patients with common atrioventricular canal defects. Subaortic stenosis is more common in patients with septum primum and Rastelli type A defects [23]. Van Arsdell and colleagues [5] have reported 19 cases of SAS associated with common atrioventricular canal defects. Eighteen of 19 patients had fibrous resection and myectomy for relief of obstruction. Seven of these underwent reoperation for recurrent SAS.

There may be an increased risk of subaortic obstruction when coarctation of the aorta is present. According to the flow-related theory, during development, a small discrete subaortic obstruction can lead to underdevelopment of the aorta which later can become unmasked. In this series, a total of four patients had coarctation present and three had associated valvular stenosis.

Other authors have reported that the modified Konno procedure is effective for the relief of diffuse SAS and can safely be performed in infants and children [24]. Vouhe and colleagues have reported 11 patients who underwent the modified Konno procedure [25]. There was one early death and two patients required reoperation, one for iatrogenic aortic regurgitation and one for residual VSD and mitral regurgitation.

In conclusion, we believe the results of this analysis support our preference for the modified Konno procedure and resection of the conal septum in patients with tunnel-like SAS but normal-sized aortic annulus and normal aortic valve. Excellent results can be achieved with these procedures with minimal risk to the aortic valve and the conducting tissue.

Acknowledgements

We would like to thank S. Yen Ho, PhD, FRCPath; for providing the figures of the modified Konno operation.
References


Appendix A. Conference discussion

Dr F. Lacour-Gayet (Le Plessis Robinson, France): I saw that you had 15 double-outlet right ventricle associated in your patients. We have the same experience in Paris. I would like to ask you what factors and what type of double-outlet right ventricle were associated with the risk of subaortic obstruction; namely, was it DORV with remote VSD or Taussig–Bing, or another type?

Dr Jahangiri: As you and your colleagues have pointed out, the risk factor is when the VSD has been restrictive or there has been an existing subaortic stenosis present. In the 15 patients here, two required enlargement of the VSD prior to the repair of a double-outlet right ventricle, and another two patients had straddling AV valve. Also, one patient had a doubly-committed VSD, and we believe, like yourself and your colleagues, that a doubly-committed VSD can also contribute to a future subaortic stenosis.

Dr Lacour-Gayet: I have another question on the association with interrupted aortic arch, that included eight cases in your series. We addressed this particular subject yesterday at the postgraduate course and we really regret that we were not present to discuss your elegant study. Can you again identify the interrupted aortic arch at risk, probably type B, or do you have any comments?

Dr Jahangiri: It’s interesting. They were all type B interrupted arch.

Dr Lacour-Gayet: Thank you. Well, this is also our experience.

Dr R. Neirottti (Grand Rapids, MI, USA): I am a bit confused with your material, because I am not sure if one should regard a patient with double-outlet right ventricle that has an operation and then develops subaortic stenosis as someone having a tunnel-like subaortic stenosis, because the mechanism of subaortic obstruction is either a patch that has not the right geometry or a VSD that is restrictive. So my question is, do you think or are you sure that a patient with a double-outlet right ventricle that has a previous operation and develops subaortic stenosis should be regarded as someone having tunnel subaortic stenosis?

Dr Jahangiri: I hope that I have made this clear, but it is difficult to define tunnel-like subaortic stenosis. In this cohort of patients, we identified any patient who has had, let’s say, a muscular type of obstruction anywhere extending to the LV cavity as having tunnel-like subaortic stenosis. The double-outlet RV group – and it will be clarified in the published paper – is a different subgroup. Two patients did have a restrictive VSD which required enlargement.