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1. Introduction
Congenital heart disease; Aortic stenosis

Keywords: Congenital Aortic stenosis

Abstract
We reviewed the surgical outcomes in adults and children with subaortic stenosis and intact ventricular septum in the current era. The case notes of 50 patients were reviewed for retrospective evaluation of preoperative, intraoperative and postoperative data. Data of primary operations during the period 2000–2005 were compared with data from patients who had re-do surgery during the same period. Thirty-five patients had primary operation and 15 patients had re-do surgery. The median age at primary operation was eight years (range 3 to 44), at second operation was 14 years (range 9 to 26) and at third operation was 15 (range 9 to 47). The entire group had been followed up postoperatively for a median of 2.5 years (range 0 to 5). Pre-operatively, aortic regurgitation was moderate in 13 and severe in three patients. Moderate to severe aortic regurgitation was present in 7 (20%) patients with primary operations and 9 (60%) patients with re-do surgery (P=0.01). Reviewing the first operations of all the re-dos (15 patients) in our series, one patient had myectomy and the rest (14 patients) had isolated resection. Aortic valve regurgitation is more prevalent in patients with recurrent subaortic stenosis. Addition of myectomy is better than shelf resection only.

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1. Introduction
There is unresolved speculation and controversy on why and how left ventricular outflow obstruction develops and when is the optimal time for surgical intervention to prevent recurrence of obstruction or aortic valve damage [1]. Most clinical series are on adults only or are mixed series including patients with ventricular septal defect or major intracardiac anomalies [1–11,13,15]. We herein reviewed a group of patients – children and adults – with intact ventricular septum undergoing surgical treatment of subaortic stenosis. By excluding patients with ventricular septal defects and additional complex anomalies, we aim to provide analysis in a group that has relatively comparable morphology. Our aim was to compare data for primary operations and re-do surgery.

2. Patients and methods
We retrieved case notes of all patients who underwent surgery for relief of discrete subaortic stenosis during the period 2000 to 2005 at the Royal Brompton Hospital, London, for retrospective analysis (Table 1). These patients had subaortic stenosis and intact ventricular septum with-

2.1. Statistical analysis
Data are expressed as mean values. Risk factors associated with re-do surgery were assessed by univariate analysis. Non-parametric Mann–Whitney test was used to compare data for primary operations and re-do surgery, and whether patients had myectomy or not.

3. Results
There were 26 male and 24 female patients with discrete subaortic stenosis. Thirty-five patients were operated on
for the first time, seven for the second time and eight for the third time. The 15 patients who were re-dos had previous resection from an earlier era between 1964 to 2000. For all patients, the follow up since the last operation for subaortic stenosis, achieved a median of 2.5 years (range 0–5 years). For patients having re-do surgery, the follow up since their first operation for subaortic stenosis achieved a median of 13 years (range 4–41 years).

The median age at first operation was 8 years (range 3 to 44), at second operation was 14 years (range 9 to 26) and at third operation was 15 years (range 9 to 47). There were 32 patients in New York Heart Association functional class I, 17 in class II, and one in class III at first and repeat surgery. Twenty-seven patients were asymptomatic.

Mean Doppler echocardiographic gradient across the LVOT was 70 ± 21 mmHg. Aortic valve regurgitation was present in 38 patients. It was mild in 22, moderate in 13, and severe in three patients. Aortic valve stenosis was present in six patients. Mitral stenosis was found in one patient and two patients had mild mitral valve regurgitation.

### 3.1. Surgical techniques

All surgical procedures were performed with cardiopulmonary bypass. The mean operative temperature for patients having first time surgery was 31.7 ± 2.4 °C and for patients having redo surgery it was 28.2 ± 3 °C. Myocardial protection was given with crystalloid or blood cardioplegia. The mean cardiopulmonary bypass time was 41 ± 14.6 min and the mean crossclamp time was 27 ± 11.4 min for first-time surgery. This was 109 ± 60 min and 76 ± 42 min for redo surgery. The obstructive lesion was approached through an aortotomy.

Shelf resection alone was carried out in 22 patients at primary operation and in two re-do patients, whereas shelf resection with myectomy was performed in ten patients and two patients at primary and re-do procedures, respectively (Table 2). Fibrous trigone mobilization in addition to shelf resection was carried out in three patients. The remaining patients had aortic valve replacement with mechanical valve, or root replacement with homograft, or Ross procedure. Two of the Ross procedures were carried out at primary operation, one at second and three at third operation.

There were no hospital deaths over the five-year period. Complications were: five required permanent pacemakers (10% risk) and two had sternal infection re-explored, nine had other complications (hypertension, infection and arrhythmias) which resolved. The four patients needing re-operations within five years were for residual gradient (≥30 mmHg) (1 first-timer), drainage of pericardial effusion (1 first-timer), emergency exploration for cardiac arrest (1 third-timer) and ventricular fibrillation and right heart failure needing a coronary bypass graft (1 third-timer).

### 3.2. Echocardiographic gradient

The preoperative LVOT echocardiographic gradient was reduced from 65 ± 18.7 mmHg to 19 ± 10.5 mmHg in first-timers and from 82 ± 21.8 mmHg to 26.9 ± 9.1 mmHg in re-dos after surgery. At the last available follow up the postoperative LVOT echocardiographic gradient was 30 ± 24.6 mmHg and 31.3 ± 13.7 mmHg for patients having surgery for the first time and re-do surgery, respectively. Amongst the primary operations three patients had gradient ≥30 mmHg in the immediate postoperative period (they had subaortic stenosis resection only) and six patients at the last echocardiographic data available. Amongst the re-dos, four patients had gradient ≥30 mmHg at immediate postoperative period and two patients at last echocardiography.

### 3.3. Aortic valve function

Sixteen patients had moderate to severe aortic regurgitation before operation. These were seven patients at primary operation and nine patients at re-do. The nine patients in the re-do group had aortic valve procedures (four Ross procedures, two aortic valve replacements and three root replacements) and the rest had subaortic stenosis resection. Aortic valve repair was not performed in any patient.

### 3.4. Recurrence

In total, fifteen patients (30%) were having re-do surgery. Reviewing the first time operations of all the re-dos (15 patients) in our series, only one patient had myectomy and the rest (14 patients) had isolated resection.
4. Discussion

There were no deaths in our series but in a combined series of 314 patients compiled from the literature with subaortic surgery in an earlier era, mortality was around 5% [7]. Those patients with previous moderate to severe aortic regurgitation are more likely to have re-do surgery for subaortic stenosis.

4.1. Aortic valve

Multifactorial aetiologies have been put forward for development of aortic regurgitation in association with subaortic stenosis: involvement of aortic valve leaflets by the shelf lesion, leaflet distortion by turbulent blood flow, infective endocarditis, cusp thickening from poststenotic turbulence, etc. [1–3].

Some reports have suggested that aortic regurgitation may progress after surgery for subaortic stenosis while others have suggested that aortic regurgitation remains trivial or mild unless endocarditis occurs [4,7].

Preoperatively, in the first-timers there were seven patients (20%) and in re-dos nine patients (60%) with moderate to severe aortic regurgitation (Table 1). Overall, 32% of patients had moderate to severe aortic regurgitation preoperatively compared to none postoperatively. We agree with Serraf and colleagues that relief of subvalvar aortic stenosis reduces the degree of aortic regurgitation in these patients [12]. Early surgery seems to prevent aortic valve disease in our cohort, in concordance with Brauner and colleagues [13].

Since our study is a five-year retrospective study, the follow-up might not be long enough to include late development of aortic regurgitation postoperatively.

A major proportion of patients who had re-do surgery for subaortic stenosis with intact ventricular septum also required valve replacements (Table 2).

We agree with Shem-Tov et al. [5] that, because of the progressive nature of left ventricular outflow tract obstruction, it is unreasonable to use the same echocardiographic pressure gradient (50 mmHg) criteria for surgery as those used for valvar aortic stenosis. In view of high incidence of aortic regurgitation with subaortic stenosis and low mortality [7,11–13] associated with surgery, we suggest the use of a milder echocardiographic gradient (40 mmHg and above in asymptomatic and 30 mmHg and above in symptomatic patients) as an indication for surgery.

4.2. Recurrence

Seven patients were presenting for the second time and eight patients were presenting for the third time. Lupinetti and colleagues [8] (40 patients with 12 years follow up) and Rayburn and colleagues [9] (23 patients with 14 years follow up) recommended the addition of septal myectomy to shelf resection in order to reduce the frequency of reoperation. In those without myectomy, the LVOT gradient increased postoperatively at a greater rate than those with myectomy.

Furthermore, in our series there was a significant difference for preoperative LVOT gradient between first-timers and re-dos. There were more patients in the re-do group with immediate postoperative gradient in LVOT 30 mmHg than first-timers. It is important to pay attention to immediate postoperative gradient as recurrence, reoperation, and mortality can be influenced by residual postoperative left ventricular outflow gradient [7,12]. Prior coarctation repairs were 3 (9%) at primary operation and 4 (27%) at re-do surgery with a P-value of 0.21. A large prospective study is needed to analyse the relative risk of recurrence in the setting of associated coarctation. We have excluded patients with ventricular septal defects and major cardiac anomalies as they can contribute to turbulent fluid shear stress and proliferation. The effect of coarctation as a contributing factor into turbulence in the left ventricular outflow tract needs to be investigated in further studies.

Stewart and colleagues, with a ‘mixed’ series, reported 50% of their patients including those with ventricular septal defect required reoperation, some more than 15 years after the initial surgery, while de Vries and colleagues found recurrence of stenosis and progression of aortic regurgitation in as high as 55% of cases. Brauner and colleagues found 20% recurrence over 6.7 years and, despite relief of obstruction, progressive aortic regurgitation was noted in 40% of those with echocardiographic gradient >40 mmHg compared with only 12.5% of those with gradient ≤40 mmHg, suggesting that early intervention or more extensive surgery might prevent recurrence, reoperation and aortic valve damage [13].

Subaortic stenosis surgery has a very low mortality in specialized units [10–14]. Early surgery and at a lower gradient (30 mmHg) have been recommended to prevent the development of aortic regurgitation and improve results [10,13]. Some surgeons even advocate surgery for subvalvar stenosis at the time of diagnosis [15], although this does not prevent the recurrence of subaortic stenosis in the long term. Close long-term follow up of 10–30 years is needed since recurrence can occur nearly thirty years after initial surgery.

4.3. Limitations

The retrospective nature of this study may be considered an important limitation, and the data should be confirmed by prospective studies. This study is based on the experience of one tertiary unit limiting patient numbers and patient selection to local practice.

A further limitation is the relatively short period of follow up (five years with median of 2.5 years). Many previous studies mixed isolated discrete subaortic stenosis with cases that had additional complex cardiac lesions [1,6–11,13,15].

Extending this type of study to multicentres will allow comparisons of working practices such as indications for surgery and type of surgery that will allow consistent and coherent guidelines to be constructed to treat this group of patients.

5. Conclusions

A major proportion of patients who have reoperation for subaortic stenosis also require valve replacement because of moderate to severe aortic regurgitation. We emphasize the need for earlier repair and need for concomitant myectomy.
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References


