Long-term follow-up and outcomes of discrete subaortic stenosis resection in children

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ABSTRACT

Background : **Studies of long-term outcomes of discrete subaortic stenosis (DSS) are rare. Therefore, we reviewed the long-term outcomes of subaortic membrane resection in children with isolated DSS over 16 years from a single institution.**

Materials and Methods : **We retrospectively reviewed the records of patients (n = 27) who underwent resection of DSS between 2000 and 2017. Patients with major concomitant intracardiac anomalies were excluded. Indications for surgery were mean left ventricular outflow tract (LVOT), Doppler gradient >30 mmHg, and/or progressive aortic insufficiency.**

Results : **The mean age at diagnosis was 3.77 ± 3.49 years (range, 0.25–13 years) and the mean age at surgery was 6.36 ± 3.69 years (range, 1–13 years). All patients underwent resection of subaortic membrane. The mean LVOT Doppler gradient decreased from 40.52 ± 11.41 mmHg preoperatively to 8.48 ± 5.06 mmHg postoperatively (P < 0.001). The peak instantaneous LVOT Doppler gradient decreased from 75.41 ± 15.22 mmHg preoperatively to 18.11 ± 11.44 mmHg postoperatively (P < 0.001). At the latest follow-up, the peak gradient was 17.63 ± 8.93 mmHg. The mean follow-up was 7.47 ± 3.53 years (median 6.33 years; range 2.67–16 years). There was no operative mortality or late mortality. Recurrence of subaortic membrane occurred in 7 (25.92%, 7/27) patients who underwent primary DSS operation. Four (14.81%, 4/27) patients required reoperation for DSS recurrence at a median time of 4.8 years (3.1–9.1 years) after the initial repair. Risk factors for reoperation were age <6 years at initial repair. Eighteen (66.66%, 18/27) patients had AI preoperatively and progression of AI occurred in 70.37% (19/27). This included 4 (22.22%, 4/18) patients who had worsening of their preoperative AI. Short valve-to-membrane distance was found to be prognostically unfavorable. One (3.7%, 1/27) patient had an iatrogenic ventricular septal defect, and 2 (7.4%, 2/27) patients had complete AV block following membrane resection.**

Conclusions : **Resection of subaortic membrane in children is associated with low mortality. Higher LVOT gradient, younger age at initial repair, and shorter valve-to-membrane distance were found to be associated with adverse outcome. Recurrence and reoperation rates are high, and progression of aortic insufficiency following subaortic membrane resection is common. Therefore, these patients warrant close follow-up into adult life.**

Keywords : Discrete subaortic stenosis, follow-up, outcomes, subaortic membrane, surgical resection

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INTRODUCTION

Discrete subaortic stenosis (DSS) is a well-described cause of isolated left ventricular outflow tract (LVOT) obstruction in children.[1] The lesion is of obvious hemodynamic significance, but in addition, it is recognized to be the result of a dynamic process that continues and has consequences into adulthood.

DSS is an often progressive disease due to membranous or fibromuscular obstruction in the LVOT. Discrete subaortic membrane accounts for 8%–10% of all cases of LVOT obstruction in children.[2] If untreated, severe DSS has a high morbidity and mortality.[3] Aortic valve regurgitation (AR) is the most common and important complication of DSS.[4]

To prevent progressive valvular damage and ventricular hypertrophy, early surgery is proposed by some groups, claiming that younger patients and patients with low LVOT gradients have the best surgical outcomes.[5] However, other investigators believe that prophylactic intervention has no benefits and is therefore not necessary.[6]

DSS is commonly found in conjunction with other cardiac abnormalities, such as ventricular septal defects (VSDs) and aortic arch abnormalities. The majority of studies on surgical outcomes of subaortic stenosis (SAS) are mixed series, including patients with complex cardiac abnormalities. Studies of discrete SAS are rare. We, therefore, sought to review the long-term outcomes of surgical resection of discrete SAS over the last 16 years in a cohort with similar LVOT morphology and an intact ventricular septum.

MATERIALS AND METHODS

We retrospectively reviewed the records of patients who are followed at the Pediatric Cardiology Clinic of Chest Diseases Hospital, Kuwait, between 2000 and 2017. Twenty-seven patients underwent fibromuscular resection of discrete SAS. We included all patients treated for membranous SAS without other associated intracardiac lesions that could have additional effects on the LVOT, including VSDs and hypertrophic obstructive cardiomyopathies. Twenty-seven patients met our criteria.

Patients with major concomitant intracardiac anomalies were excluded from analysis. We included only minor associated anomalies that would not alter the primary diagnosis of "discrete SAS." Patients with atrial septal defect, pulmonary valve stenosis, patent ductus arteriosus (PDA), or coarctation of aorta (CoA) were included in our study. Patients with narrow LVOT obstruction (tunnel type of SAS) were excluded.

Indications for surgery were mean LVOT Doppler gradient >30 mmHg and/or progressive AI. Medical records were retrospectively reviewed until the last cardiology follow-up. This included inpatient notes, surgical reports, and outpatient notes.

The following baseline variables were analyzed: mean age at diagnosis; mean age at surgery and last follow-up; gender; SAS type (discrete); and the presence of concomitant cardiac lesions such as bicuspid aortic valve (AV), PDA, atrial septal defect, pulmonary valve stenosis, and CoA.

Pre- and postoperative assessment was done using echocardiography in all patients. Pressure gradients were measured across the LVOT. The presence and degree of AR, aortic valve morphology, its distance to the AV, and left ventricular (LV) systolic function were obtained from the echocardiographic reports. Type of surgery, cardiopulmonary bypass time, aortic cross-clamp time, and other intraoperative data were obtained from the operation notes.

Recurrence of SAS was defined as peak instantaneous LVOT Doppler gradient >40 mmHg at any time after the first postoperative month.[6] Progression of AI was defined as worsening of preoperative AI, new AI developing postoperatively, or aortic valve repair or replacement occurring postoperatively.

Statistical analysis

The data management and analysis were carried out using the computer software "Statistical Package for Social Sciences," SPSS version 25.0' (IBM Corp., Armonk, NY, USA). The descriptive statistics has been presented as frequencies and percentages for categorical variables. The quantitative or continuous variables were ascertained for normal distribution assumption, applying the Kolmogorov–Smirnov test, and presented as; means ± standard deviations and range as well as median, range, interquartile[7] for most of the variable data were skewed. The mean values, before and after, were compared using nonparametric Wilcoxon signed-rank test. The two-tailed $P < 0.05$ was considered statistically significant.

RESULTS

During the period from January 2000 to December 2017, a total of 27 patients were treated for membranous SAS. All the patients had a discrete subaortic membrane. Of the 27 patients, 17 (62.97%) were male and 10 (37.03%) were female. Of the 27 patients, 3 (11.11%) underwent membrane resection plus aggressive septal myectomy and 24 (88.89%) underwent membrane resection alone. The mean age at diagnosis was 3.77 ± 3.49 years (range, 0.25–13 years) and the mean age at surgery was 6.36 ± 3.69 years

(range, 1–13 years). The baseline demographic data for those patients who underwent surgical resection are shown in Table 1.

Concomitant cardiovascular anomalies are described in Table 2. Four patients underwent cardiac surgery before developing SAS. These were CoA repair ($n = 3$) and PDA ligation ($n = 1$). Two patients underwent cardiac catheter interventions before developing SAS. These were CoA balloon angioplasty ($n = 1$) and balloon dilatation of the pulmonary valve ($n = 1$). All patients underwent resection of subaortic membrane.

Table 1: General characteristics of the patients

Characteristic	Value
Male:female (n)	17:10
Male:female (ratio)	1.7:1.0
Age (years)	
Mean±SD	11.27±4.46
Median (range) IQ	11.25 (4.33-19) 7.08-15.41
Age at diagnosis (years)	
Mean±SD	3.77±3.49
Median (range) IQ	2.50 (0.25-13) 0.83-7.0
Distance from aortic valve (mm)	
Mean±SD	07.3±01.54
Median (range) IQ	07.0 (05-10.0) 06.0-08.25
Age at surgery (years)	
Mean±SD	6.36±3.69
Median (range) IQ	5.58 (1-13) 3.75-10
Recurrence (%)	7 (26)
Redo surgery (%)	4 (14.8)
Age at redo surgery (years)	
Mean±SD	7.65±2.44
Median (range) IQ	0.88 (4-9) 5.15-7.75
Postoperative complications (%)	
Complete heart block	2 (7.4)
Iatrogenic VSD	1 (3.7)
Follow-up period (years)	
Mean±SD	7.47±3.53
Median (range) IQ	6.33 (2.67-16) 5-9.75

IQ: Inter quartile, VSD: Ventricular septal defect, SD: Standard deviation

Table 2: Concomitant cardiovascular anomalies

Variables	Frequency, *n* (%)
ASD	3 (11.1)
CoA	3 (11.1)
PDA	2 (7.4)
PVS	1 (3.7)
Bileaflet aortic valve	5 (18.5)
Aortic valve stenosis	1 (3.7)
Down syndrome	1 (3.7)

CoA: Coarctation of aorta, PDA: Patent ductus arteriosus, ASD: Atrial septal defect, PVS: Pulmonary valvular stenosis

The mean follow-up was 7.47 ± 3.53 years (median 6.33 years; range 2.67–16 years). There was 100% follow-up till date. There was no operative mortality or late mortality.

The mean LVOT Doppler gradient decreased from 40.52 ± 11.41 mmHg preoperatively to 8.48 ± 5.06 mmHg postoperatively ($P < 0.001$). The peak instantaneous LVOT Doppler gradient decreased from 75.41 ± 15.22 mmHg preoperatively to 18.11 ± 11.44 mmHg postoperatively ($P < 0.001$) [Table 3]. At the latest follow-up, the peak gradient was 17.63 ± 8.93 mmHg.

Recurrence of SAS occurred in seven patients who underwent primary DSS operation (25.92%, 7/27). Four (14.81%, 4/27) patients required reoperation for DSS recurrence at a median time of 4.8 years (3.1–9.1 years) after the initial repair.

Of the seven patients with evidence of regrowth of the subaortic membrane, four of them eventually underwent repeat surgery for a recurrence of subaortic obstruction. The mean LVOT gradient for these four patients at the time of second surgery was 43.1 ± 12.4 mmHg. The intraoperative finding in all these patients was recurrence (growth) of discrete subaortic membrane. For these redo cases, the median follow-up period from their first surgery to the last surgery was 7 years (3–10 years). At the second surgery, all patients underwent membrane resection plus aggressive septal myectomy. The recurrence rate was noted to be high in patients who underwent first surgery at the early age (<6 years) and in patients with shorter valve-to-membrane distance (<6 mm).

Eighteen (66.66%, 18/27) patients had AI preoperatively with trivial AI in nine patients, mild AI in eight patients, and moderate AI in one patient. Postoperatively, 16 (59.25%, 16/27) patients had AI with trivial AI in eight patients, mild AI in seven patients, and moderate AI in one patient. There was no severe AI. At the latest follow-up, 19 (70.37%, 19/27) patients had AI with trivial AI in six, mild AI in eleven patients, and moderate AI in two patients. This included four patients who had worsening of their preoperative AI (22.22%, 4/18). One patient had no AI preoperatively had developed mild AI at the latest follow-up.

One (3.7%, 1/27) patient had an iatrogenic VSD following DSS repair that required device closure later. In addition,

Table 3: Left ventricular outflow tract gradients and aortic regurgitation before and after surgery

Variables	Preoperative	Postoperative	Last follow-up
Peak instantaneous LVOT gradient (mmHg)	75.41±15.22	18.11±11.44 (*P*<001)*	17.63±8.93 (*P*<001)**
Mean LVOT gradient (mmHg)	40.52±11.41	8.48±5.06 (*P*<001)*	8.93±5.64 (*P*<001)**
Aortic regurgitation			
No	9	11	8
Trivial	9	8	6
Mild	8	7	11
Moderate	1	1	2

*Preoperative versus early postoperative, **Preoperative versus last follow-up. LVOT: Left ventricular outflow tract

2 (7.4%, 2/27) patients had complete AV block following membrane resection; both patients required insertion of a permanent pacemaker.

DISCUSSION

SAS encompasses a variety of anatomic lesions that can occur either alone or in combination. The following discrete entities have been described in literature:[8] thin, crescent-shaped membrane just below the aortic valve (discrete SAS) – this represents 75%–85% of SAS cases; thick fibromuscular ridge; and tunnel or tubular: long, narrow, fibromuscular channel along the LVOT.

DSS in its discrete form, without a tubular obstruction of the LVOT, is essentially a circular rim of tissue, with a fibrous inner ring of varying width. The location will vary from just beneath the aortic valve, where occasionally it will be fused with the dependent portion of a cusp, to a position lower down the LVOT with attachments to the anterior leaflet of the mitral valve. This location means that not only will it place a load on the LV, but the resulting turbulence will also affect the aortic valve. The mechanism for DSS formation has been the subject of debate since its original description. Despite being generally classified as a congenital heart defect and on rare occasion, appearing in infancy, the general opinion is that DSS is an acquired lesion.[1] There have only been rare reports, suggesting a familial occurrence, and overall, there is little evidence that it is a primarily genetic disorder.[9] Although a developmental origin has the most support, it does not appear to completely explain these lesions, and questions remain. The mechanism of DSS formation has not been settled completely; nevertheless, the information available has led to what appears to be a mechanism for its development with implications for longer-term treatment.

Factors associated with the rate of progression of LVOT obstruction are not completely clear. It is thought that abnormal fluid dynamic forces at the LVOT level can cause septal shear stress, causing cellular growth factors to engineer regional cellular proliferation contributing to the worsening of LVOT obstruction.[10] Why the rate of progression is different in children compared with adults is not completely understood at this time. Perhaps, the earlier in life the septal shear stress is increased above a threshold, the more intense the response and the more rapid the progression of the LVOT obstruction. The action of the shear forces on the endothelial cell layer of the LVOT could stimulate proliferation of these cells and start the process.

The progressive nature of LVOT obstruction caused by DSS in children has been well documented in literature.[1] DSS may progress rapidly in some patients,[11] while it follows a slower course in others. The exact etiology and factors contributing to the rate and severity of progression remain unknown. However, discrete SAS progresses slowly in adulthood.

Echocardiography is the test of choice to diagnose SAS. It is used to characterize the anatomy of the subaortic lesion and to assess LVOT involvement and dimensions and function of the LV, as well as the integrity of the aortic and mitral valves. However, often, it is difficult to assess the degree of obstruction of outflow in SAS on a 2-dimensional echocardiogram, and thus, Doppler examination is indicated.

Doppler examination may be inconclusive, transesophageal echocardiography is more reliable for the accurate diagnosis of a subaortic membrane that is masked by the hypertrophied and prominent ventricular septum.

Cardiac catheterization is sometimes performed to further clarify the mechanism and extent of subaortic obstruction. This provides hemodynamic data such as the gradient across the valve, measurement of cardiac output, and estimates of the degree of AR. However, cardiac catheterization is not typically indicated in the diagnosis of SAS, but it can be utilized for preoperative hemodynamic evaluation and for preoperative workup before surgical repair to rule out significant coronary artery disease.

Definitive therapy for DSS consists of surgical correction of the obstruction, which may involve simple membrane removal or extensive ring resection with or without myectomy. Currently, there are no established medical therapies to reverse or stop the progression of DSS, including balloon dilation.

The surgical resection of a DSS is well understood, as are its pitfalls. Although surgical repair of DSS has excellent short-term outcomes, it is associated with up to 8% chance of an iatrogenic VSD,[12] up to 14% chance of complete atrioventricular block,[13] and a recurrence rate of 5%–27%.[14] Resection of the fibromuscular tissue typically begins below the right coronary ostium and can be carried posteriorly and leftward along the ventricular septum without significantly incurring the risk of heart block. More posteriorly along the septum, the chance of creating a VSD will be greater with a deeper resection; however, and not to minimize this complication, it should be apparent and can be repaired at the time.

Surgical resection of DSS has generally excellent short-term outcomes, with low mortality. There was no operative mortality in our cohort, consistent with other studies of discrete SAS reporting an operative mortality rate close to 0%. Rohlicek *et al.* studied 42 patients who underwent SAS resection between 1985 and 1998 with no operative mortality and 2.4% (1/42) late mortality.[12] Serraf *et al.*, however, in their study of 160 patients

between 1980 and 1997 reported operative mortality of 3% (5/160).[14]

Extension of the subaortic membrane onto the aortic or mitral valve has also been associated with poor outcomes. We found that extension of the membrane onto the aortic valve was a significant risk factor for SAS recurrence and reoperation. Geva *et al.* similarly found that the need for intraoperative peeling of the SAS membrane from the aortic or mitral valves is associated with a shorter time to reoperation.[15]

Despite successful resection of subaortic membrane, recurrence and reoperation rates remain high. In our study, recurrence occurred in seven patients (26.0%, 7/27), over a mean follow-up period of 7.47 years, with four patients (14.8%, 4/27) requiring reoperation. A further three patients (11.1%, 3/27) had recurrence, but did not require reoperation during the study. This rate of reoperation is 2% per year, which is similar to other studies. Drolet *et al.*[16] conducted a study of 92 patients diagnosed with SAS between 1985 and 1998. Forty-nine patients required surgery at a mean of 3.3 years after diagnosis and 10 (20%, 10/49) patients required reoperation during a mean follow-up period of 6.2 years. This is equivalent to a reoperation rate of 3.3% per year. Babaoglu *et al.*[17] conducted a similar study, reporting a reoperation rate of 8.3% (2/24) over a follow-up period of 4.8 years, equivalent to 1.7% per year. Similarly, in a mixed study, Ruzmetov *et al.* also reported a high reoperation rate in their study of 190 patients.[18] Over the follow-up period of 9.6 years, 26.3% (50/190) of patients required reoperation for recurrent SAS, equivalent to 2.7% per year.

A few risk factors for SAS recurrence and reoperation have been reported in the literature.[14] Of particular significance is the peak instantaneous LVOT Doppler gradient, which appears to be an indicator for poor outcomes when >50 mmHg, whether at diagnosis, preoperatively, or postoperatively. Rohlicek *et al.* found reoperation to be associated with a higher peak instantaneous LVOT Doppler gradient at diagnosis, with those requiring reoperation having a mean peak gradient of 66 mmHg compared to those who did not require reoperation, with a mean peak gradient of 34 mmHg.[12] Geva *et al.*, who reported a reoperation rate of 1.7% per year in their study of 111 children, found that preoperative peak instantaneous LVOT Doppler gradient >60 mmHg was an independent predictor of earlier time to reoperation.[15] Hirata *et al.*, in their mixed study of 106 children, also found the preoperative peak instantaneous LVOT Doppler gradient significant in predicting SAS recurrence.[19] Serraf *et al.* found postoperative peak instantaneous LVOT Doppler gradient to be a predictor of recurrence and reoperation.[14] In our cohort, we found an association between preoperative peak instantaneous LVOT Doppler gradient and risk of recurrence or reoperation.

Other reported risk factors for recurrence and reoperation include concomitant cardiovascular defects such as CoA[14] and younger age at operation.[15] Serraf *et al.* and Hirata *et al.* in their studies found CoA to be a risk factor for recurrence and reoperation, suggesting that children with CoA be regularly assessed for SAS.[14] In our study, we did not find any association between minor concomitant cardiovascular anomalies and SAS recurrence or reoperation. In our cohort, patients with SAS and concomitant CoA had good outcomes. We did, however, find younger age to increase the risk of SAS reoperation. Geva *et al.* reported that younger age at initial surgery predicted earlier reoperations and suggested that children diagnosed earlier have more aggressive underlying pathology.[15] Our study similarly found age <6 years at initial repair to be an independent predictor of DSS reoperation. Of 7 (26%, 7/27) patients, who had recurrence, 6 (85.71%, 6/7) patients had initial surgery at the age of 6 years or less. All the four patients, who had reoperation, had their initial surgery at the age of <6 years.

More common and usually of much greater long-term significance is damage to the aortic leaflets from the jet effect and turbulence generated by the DSS. Although turbulence is a universal feature of DSS, the amount of AR that results has been variable even over the medium term. Regurgitation results from cusp thickening and retraction, the result of a general response that appears to resemble the formation of a DSS. Certainly, a turbulent jet hitting the undersurface of the aortic valve at the onset of systole might damage the surface layer of the cusp, inciting a fibroblast response and, with more injury and inflammation, produces thickened, contracted cusps with impaired cooptation.[20] Once damage has begun, it will probably continue, especially with the likely residual and increasing LVOT obstruction producing turbulence.

Our study showed that although AI was improved immediately postoperatively, at long-term follow-up, the severity of AI was variable, with improved or unchanged AI in 23 patients. At latest follow-up, 19 (70.37%, 19/27) patients had AI with trivial AI in six, mild AI in eleven, and moderate AI in two patients. Progression of AI occurred in 66.66% (19/27) of patients. This included four patients who had worsening of their preoperative AI (22.22%, 4/18).

Rohlicek *et al.* found AI to progress in both patients who underwent surgery during the study period and patients who did not.[12] They concluded that surgery had little beneficial effect on severity of postoperative AI or the development of new AI postoperatively. This is in contrast to other studies recommending early "prophylactic" SAS resection to prevent the development

of moderate–severe AI.[21] Our results suggest that AI may progress despite resection of discrete SAS.

When the initial gradient was ≥80 mmHg, the results indicated that the DSS would recur more rapidly and the number of patients with AR would increase. There would likely be widespread agreement that an operation should be performed for a peak gradient of 60 mmHg or a mean gradient of 40 mmHg.[22] For many, the presence of aortic valve dysfunction would lead to a recommendation for resection at an even lower gradient.

Recent studies[16,21] confirm that a higher LVOT gradient at diagnosis is an independent predictor of various adverse outcomes such as AR, faster AR progression, faster progression of LVOT obstruction, recurrence, and surgical intervention. One of the other studies[17] found that the peak LVOT gradient was significantly higher in patients with progressive AR than in those whose AR showed no signs of progression, but did not perform multivariable analyses on their data. The observation that LVOT outflow tract obstruction severity is correlated with AR progression provides important information for prognostication and clinical decision-making.[23]

The risk of reoperation may be due to inadequate resection at the first operation, yet recurrent obstruction may appear despite the adequacy of surgical excision.[24]

Myectomy is another intervention that can be done to help alleviate LVOT obstruction in DSS. However, even after undergoing myectomy, there is still a high chance of recurrence, with reoperation rates between 10% and 20% within 10 years. In addition, myectomy is associated with an increased risk of complete heart block. Therefore, given the combination of no long-term benefit and the risk of heart block, myectomy should not be performed routinely, and it only should be performed if marked LVH is present.[25]

Surgical complications of DSS resection have found a high number of patients developing complete AV block and iatrogenic VSDs. Drolet *et al.* found the rate of AV block to be similarly high, at 6% (3/49), with all requiring a permanent pacemaker. These patients subsequently went on to have a permanent pacemaker inserted. In our study, iatrogenic VSD occurred in only 1 (3.7%, 1/27) patient, who required device closure later. This rate is also consistent with other studies.[12,13] In addition, 2 (7.4%, 2/27) patients had complete AV block following membrane resection; both patients required insertion of a permanent pacemaker.

Although the subvalvular obstruction may be a complex 3D structure that does not necessarily encircle the LVOT, a level can often be identified to allow measurement of its distance to the AV. Interestingly, two studies[25,26] found a longer distance of the subvalvular obstruction from the base of the AV to be associated with less progressive LVOT obstruction and potentially predictive of being a low-risk patient.

Geva *et al.*[15] found a shorter valve-to-membrane distance to be prognostically unfavorable, which would suggest earlier surgical intervention be considered in patients with a shorter valve-to-membrane distance. In our study, of 7 (26%, 7/27) patients, who had recurrence, 6 (85.71%, 6/7) patients had valve-to-membrane distance of 6 mm or less. Of 4 (14.8%, 4/27) patients, who had reoperation, 3 (75%, 3/4) had valve-to-membrane distance of 6 mm and 1 (25%, 1/4) had valve-to-membrane distance of 3 mm.

One recent study[16] found that the presence of AI preoperatively was predictive of surgical intervention. Thus, AR is a major sequela in SAS patients with significant prognostic implications and should, therefore, play an integral role in the surgical decision-making process.

AR was found in >50% of patients with SAS, but only 20% are considered to be hemodynamically significant.[27] If present, the degree of AI can progress in patients who did not have any repair procedure for SAS. Studies have shown that there is a direct relationship between the severity of SAS and AI.[28] An LVOT gradient ≥80 mmHg was found to be a significant risk factor for developing AR postoperatively.

Many studies have reported a higher preoperative LVOT gradient to be a risk factor for development and progression of AI.[29,30] Babaoglu *et al.* found that patients with progressive AI postoperatively had a significantly higher preoperative LVOT gradient than patients with nonprogressive AI.[17] Our study did find this. Risk factors for progression of AI were age <6 years at operation and the need for reoperation, which suggests a more aggressive underlying pathology. These children require close monitoring of their aortic valve function.

Study limitations

This study is subject to the usual limitations of a retrospective study. Statistical analyses were limited due to the relatively small number of patients and outcomes.

CONCLUSIONS

This study underlines the importance of LVOT gradient in surgical decision-making in pediatric DSS patients and found a higher LVOT gradient to be associated with adverse outcome. The presence of AR should also be taken into consideration as prognostic factor. Younger age at initial repair and shorter valve-to-membrane distance were found to be associated with adverse outcome. Resection of discrete subaortic membrane provides safe and effective relief of LVOT obstruction in children, with low mortality. Survival is excellent after

surgery for DSS; however, recurrence and reoperation rates remain high, and these patients warrant close long-term follow-up.

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Conflicts of interest

There are no conflicts of interest.

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