Surgical repair of subaortic stenosis resection: 10 years of single-center experience in 65 patients

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Abstract

Background

Subaortic stenosis (SAS) was a rare congenital heart disease of left ventricular outflow tract (LVOT), ranging from "isolated" lesions to "tunnel" or "diffuse" lesions. We conducted a retrospective study to describe the characteristics of patients with different lesions and analyze the risk factors for reoperation.

Methods

In this study, we examined a single-center retrospective cohort of SAS patients undergoing resection from 2010 to 2019. Patients were classified as simple lesion group (n = 37) or complex lesion group (n = 28). Demographics, perioperative findings, and clinical data were analyzed.

Results

The surgical effect of the two groups was significantly lower than that before the operation ($p < 0.05$). The median age at operation was 6(3-11.8) years. There was no operative mortality. In complex lesion group, extracorporeal circulation time (CPB time), aortic cross clamp time (ACC time), mechanical ventilation time and ICU stay time were longer. The median follow-up period was 2.8 years (range 1-3.8), with two late death. Six patients (9.2%) required reoperation due to restenosis or severe aortic insufficiency. The freedom from reoperation rates at 5 years was 66.7% for simple lesion but only 52.3% for complex lesion ($p = 0.036$).

Conclusions

Although the lesions include many forms, subaortic stenosis resection was still satisfactory. However, the reoperation after initial surgical treatment was not infrequent, especially in patients with complex lesion.

Introduction

Subaortic stenosis is a relatively rare congenital heart disease, accounting for 1-2% of all congenital abnormalities, accounting for 8-30% of patients with congenital left ventricular outflow tract obstruction[1-3]. There are many reasons for subaortic stenosis. Some studies have shown that structural findings are the cause, such as steeper aorto-septal angle, associated defects or geometry of left ventricular outflow tract (LVOT)[4-6]. With the advancement of surgical techniques in recent years, the surgical results are satisfactory. However, the recurrence is still frequent. The causes of membrane recurrence are poorly understood. In some studies, the intraoperative age is low, high preoperative LVOT gradient, aortic valve disease or other heart malformations are the risk of reoperation after aortic valve stenosis [7-10].

Subaortic stenosis(SAS) may be related to morphological type, complexity of aortic valve stenosis or related defects. Kitchiner[11] classified descriptions (such as "simple form", "discrete" or "membranous") as "isolated" lesions. Complex lesion were described as “tunnel form” or “diffuse” obstruction. However, none of these studies studied the SAS status between simple lesion and complex lesion. Only several studies[7,8,12] reported that higher reoperation or mortality for complex lesion compared with simple lesion of isolated membrane-type lesion. The purpose of this research based on a decade-long retrospective study was to analyze the differences between simple lesion and complex lesion to find out the characteristics, surgical results, and reoperation rate after surgical resection.

1.Methods

1.1 Study Population

This retrospective study included SAS patients who underwent cardiac surgery at our center between April 2010 and November 2019. The inclusion criteria were a fixed sub-aortic obstruction confirmed by echocardiography and surgical records. The main indications for surgery were left ventricular outflow tract obstruction and LVOT gradient greater than 40mmHg. When the LVOT gradient was lower than 40mmHg, there were symptoms (fatigue, chest pain, syncope) and surgery was necessary. The criteria for exclusion were severe aortic stenosis, single ventricle anatomy, and hypertrophic cardiomyopathy. According to their cardiovascular pathology, we divided the patients into two groups (simple and complex). The simple lesion group consisted of all patients described as "short, discrete, or membranous" subaortic lesions. The complex lesion group consisted of all patients described as "tunnel form", "diffuse", shone’s syndrome or other lesions.

1.2 Data collection

Data were extracted from the electronic medical record system. The baseline data collected for these patients included demographics, anatomical characteristics, and SAS subtypes. Surgical parameters included surgical technique, cardiopulmonary bypass time, aortic clamping time, mechanical ventilation time and ICU stay time. Echocardiographic parameters included LVOT gradient and left ventricular ejection fraction (LVEF). Clinical and echocardiographic follow-up data for all patients were obtained.

1.4 Ethical approval

Given the retrospective design of the study, which is based on data collected for routine clinical care, administrative, and audit purposes, individual informed consent was not required. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. This study has approval by the institutional research committee at Beijing Anzhen hospital, who permitted the collection of data for audit and research purposes.

1.3 Surgical data

Surgery was carried out using conventional cardiopulmonary bypass at 28-32 °C. The aortic valve and the sub-valvular area were exposed and inspected through aortic incision. Fibroelastic tissue was excised completely. In the cases of fibromuscular ridge or tunnel-like, the scope of resection can be expanded by removing fibrous as well as muscular tissue.

1.3.1 membranous and fibromuscular ridge resection

Seventeen patients (26.2%) underwent simple membranous resection. Forty-five patients (69.2%) underwent fibromuscular ridge resection. The size of the wedge of muscle tissue removed depends on the age of the patient and the thickness of the interventricular muscular septum. Special attention was made to avoid injury to the mitral valve apparatus and conduction system.

1.3.2 Reconstruction of the intraventricular baffle

Two patients underwent prolonged septoplasty. One patient accepted the removal of the Dacron patch and the reconstruction of the intraventricular baffle. The patient was a 12-year-old boy who has undergone double outlet right ventricle (DORV) biventricular repair for 11 years. The LVOT gradients before and after the operation were 115 and 27 mmHg, respectively. Another patient underwent accessory mitral valve tissue resection, and the ventricular septal defect (VSD) was enlarged repair. He was a six-month-old boy who had his first surgery

1.3.3 Nicks procedure

One patient underwent Nicks procedure. She was a 30-year -old women who had undergone first operation. The patient underwent a prosthetic valve implantation and used the Dacron patch to enlarge the left ventricular outflow tract. The gradient of LVOT before and after the procedure was 63 and 23 mmHg, respectively.

1.4 Statistical analysis

All data were analyzed using SPSS 24.0 software. Quantitative data are given as mean standard deviation or median (range). Qualitative values were expressed as percentages. Patients were divided into two groups with simple lesion and complex lesion. In the comparison of normal variables between the two groups, we used the independent sample t test; in the comparison of categorical variables between groups, the Chi-square test was used. Logistic regression was used to determine the associations between post-operative peak LVOT gradients and patients’ ages and each of the measured outcomes. Kaplan–Meier survival curves were assessed and compared with the log-rank test according to the clinical subgroups. One-, 5-, and 10-year survival was estimated for each of the groups. All tests were mutual, and $p < 0.05$ was considered statistically significant.

2. Results

2.1 Preoperative Characteristics

During the 10 years in our center, 65 patients underwent SAS heart surgery. There were 36 males with a median age of 6 (3-11.8) years of surgery. The baseline characteristics are described in Table 1. Only ten patients (15.4%) had symptoms of chest tightness, and the rest were found by examination. There were no in-hospital deaths, and two patients needed pacemakers. Forty-seven patients (72.3%) had other malformations. There were more patients had other malformations in complex lesion group. ($p < 0.05$). The most commonly associated other cardiac defects were ventricular septal defect (VSD) (27.7%, n = 18), patent ductus arteriosus (PDA) (20%, n=13), atrioventricular septal defect (AVSD) (10.8%, n=7), left superior vena cava (LSVC) (10.8%, n=7), shone's syndrome (9.2%, n=6), coarctation (COA) (6.2%, n=4), double outlet right ventricle (DORV) (3%, n=2). In the complex lesion group, 5 patients (17.9%) were classified as "other" lesions, of which 4 were mainly caused by the accessory mitral valve tissue, and 1 was caused by papillary fibrocytoma. Twenty-seven patients (41.5%) with secondary subaortic stenosis after the initial heart surgery included 18 patients with simple lesion and 9 patients with complex lesion. The intermediary interval between the initial heart surgery and the resection of aortic stenosis is 6 years (range 3-10 years).

It is statistically significant that the CBP time, ACC time, mechanical ventilation time and ICU stay time of the complex lesion group were longer than those of the simple group. The average LVOT gradient after operation in the two groups was significantly lower than that before operation ($p < 0.05$). The average pressure gradient of the left ventricular outflow tract in the simple group decreased from 75±24.5 mmHg to 8.7 (4.6-15.3) mmHg (Figure 1). The average pressure gradient of the complex lesion group decreased from 86.7±32.1 mmHg to 18.3 (7.5-28.5) mmHg (Figure 2). It is statistically significant that the postoperative LVOT gradient of the simple lesion group was lower than that of the complex lesion group ($p < 0.05$). Thirty-four cases (51.5%) had aortic regurgitation. The simple lesion group had more patients combined with aortic regurgitation ($p<0.05$).

In our study, 8 patients had pathological reports of about 4 subtypes of resected tissue. 2 cases of membranous patients developed hyaluronic degeneration and mucoid degeneration. In 3 patients with fibromyopathy, cardiomyocyte hypertrophy, local endocardial fiber thickening and interstitial fibrosis. A tunnel-like patient developed fibrous tissue hyperplasia. One patient developed papillary fibroelastic tumor, and another patient developed hyaluronic degeneration and mucoid degeneration of the accessory mitral valve.

2.2 Follow-Up

The main postoperative complications included mortality, aortic regurgitation and mitral regurgitation. Two died late. One SAS patient was a 2.5-year-old boy with LSVC who underwent the initial correction for 2 years. The LVOT gradients before and after surgery were 69 and 10 mmHg, respectively. However, he had moderate mitral regurgitation after surgery. Maybe mitral regurgitation was the cause of death. The other SAS patient was a 7.6-year-old boy with VSD and LSVC who underwent reoperation for 5.6 years. His LVOT gradients before and after the operation were 78 and 35 mmHg, respectively.

The average follow-up time was 2.8 years (1-3.8 years). In all patients, the average LVOT gradient at the last follow-up was 10.8 (4.8-27.9) mmHg. The conditions that require repeated operation of this study included 5 patients with recurrence of stenosis and 1 patient with severe aortic regurgitation (Table 2). The probability of reoperation was 9.2%. The actuarial freedom from reoperation for 5 years in the simple lesion group was 66.7%, while that of the complex lesion group was only 52.3% (p = 0.036). (Figure 3)

3. Discussion

This single-center retrospective analysis of 65 patients underwent subaortic stenosis resection showed that the recurrence rate required for reoperation was 9.2%, which is consistent with previous studies (recurrence rate: 8%-31% [1, 7, 12]). Our research suggested that, compared with simple lesions complex lesion were accompanied by more heart defects, and surgery was more difficult. We found that both groups had satisfactory surgical outcomes, and the postoperative LVOT gradient of the complex lesion group was higher than that of the simple lesion group. During follow-up, the rate of reoperation in the complex group was higher than that in the simple group.

SAS is isolate or combined with other malformations. Some studies[2,7,8,12] indicated that the pathophysiological substrate of complex malformation provided valuable information for the long-term prognosis of SAS after cardiac surgery. In this study, 47 patients had other heart abnormalities. Compared with the simple lesion group, the complex lesion group had more heart defects.

In the presence of related cardiovascular abnormalities, SAS recurrence is common. SAS has been reported after surgical repair of several congenital heart defects. RON BRAUNER[13] et al found that the shape and size of VSD may affect the detection and resection of subaortic obstructive lesions, so it is difficult to accurately appreciate it clinically and on echocardiography. Some studies[14,15]prove that subaortic stenosis after surgical repair of atrioventricular septal defect was associated with “goose neck” feature. Hirata et al[16,17] found that the patients who underwent concomitant myectomy patients suffering from aortic coarctation or interrupted aortic arch (IAA) were at additional risk of reoperation. Twenty-seven patients (41.5%) in the study experienced a secondary subaortic stenosis after heart surgery. In this study, the most common secondary subaortic stenosis after correction were VSD (n = 9), AVSD (n = 6) and COA / IAA (n = 2), but there were no statistics between the two groups.

Both the simple lesion group and the complex lesion group had satisfactory surgical results. The postoperative LVOT gradient of the two groups was significantly lower than that before surgery. The most commonly used cardiac technique was the fibromuscular ridge resection (69.2%, n=45). It depended on the type of stenosis and the surgeon’s choice. Although, many studies[4-6] have shown that the geometry of the LVOT in hearts with subaortic stenosis and ventricular septum is abnormal. There were changes in LVOT with aorto-septal angle. But for the surgeon, the fibromuscular ridge resection can minimize the incidence of restenosis and ensured that the LVTO is sufficiently unobstructed. Mark Ruzmetov[2] suggested that the routine addition of a generous myectomy to fibrous subaortic resection for relief of tunnel LVOT obstruction may reduce the incidence of recurrent obstruction. In addition, when abnormal fibrous tissue extends or covered the anterior mitral valve leaflet, complete removal of abnormal fibrous tissue from the LVOT is important to prevent the recurrence of subaortic stenosis. But pay special attention to avoid injury to the mitral valve apparatus and conduction system. Two patients had complete heart block after fibromuscular ridge resection and required pacemakers (one DORV and one TOF). Therefore, in this study, for the complex lesion group, 20 cases (71.4%) underwent fibromuscular ridge resection. The CPB time, ACC time, mechanical ventilation time and ICU stay time of the complex lesion group were longer than those of the simple group.

Our study showed that two patients underwent extended septoplasty (one DORV and one accessory mitral valve tissue). For the DORV, the decrease in the effective VSD size, turbulent flow, the sinuous shape of the tunnelization, caused subaortic septal or conal muscle hypertrophy or fibrous tissue deposits[19]. Extended septoplasty was a safe and effective method for the treatment of subaortic stenosis as Fibro-Muscular Resection[18]. Aortic regurgitation was the most dreaded sequelae of SAS, appear to be at increased risk, also was the main risk of reoperation[8.20]. This study showed that 61(93.8%) patients had tri-leaflet valves, and 34(51.5%) patients had aortic regurgitation before surgery. Compared with the complex lesion group, the incidence of aortic insufficiency was higher in the simple group ($p < 0.05$). Ezon DS[21] showed that a membrane closer to the aortic valve is more likely to cause progression of stenosis, while a membrane farther from the aortic valve is more likely to cause aortic and mitral regurgitation. In our study, although many patients had mild aortic regurgitation before surgery, it was difficult to predict which patients would progress to moderate or severe. Some study[9] suggested aortic valve disease progression occurred independently of surgery and SAS recurrence. Moderate aortic regurgitation may progress to severe. However, our study found that 11 (16.9%) patients had moderate aortic regurgitation in preoperative, only one patient occurred severe and only one patient had severe regurgitation after one year of correction. In addition, in this study, there were fewer patients with SAS and AS, and only one patient underwent Nicks surgery and the results of the surgery were satisfactory.

It is still controversial whether complex lesions are related to reoperation. K. Valeske[7] showed that recurrence of subaortic stenosis within several years after initial surgical treatment was frequent in patients with complex lesion. Our research confirmed this finding. Kaplan–Meier analysis showed that there was significantly difference in probability of reoperation between two groups. At 5 years, the proportion of reoperation in the complex lesion group was higher than that in the simple lesion group. This result may be related to different pathology and morphology. Although the operation time, mechanical ventilation time and ICU-stay time were longer in complex lesion group, the postoperative LVOT gradient of the complex group was higher than that of the simple group. For complex lesion, regular follow-up is particularly important.

4.Limitations

This study was a retrospective review from a single institution, and there was no standardized echocardiographic review and detailed imaging data. The decision of surgical intervention and re-intervention was made by individual practitioners, and there was no standardized standard. These limited the analysis of SAS risk factors. Further research is needed to validate these echocardiographic predictors in a larger prospective study. Similarly, the median follow-up time was relatively short, and stronger inferences can be drawn from a longer follow-up time.

5.Conclusion

Subaortic stenosis is a rare congenital heart disease with different deformity characteristics and surgical results. Our study showed the average LVOT gradient after surgery in both groups was significantly lower than before operation, but the surgical procedures and outcome of complex lesions were not as good as those of patients with simple lesions. These patients with complex lesions had more malformations, longer operation time and ICU stay time, and higher post-operative LVOT gradient. During follow-up, the rate of reoperation in the complex lesion group was higher than that in the simple lesion group. Patients should continue to be closely followed up after surgery to find and promptly treat the recurrence of subaortic valve stenosis or early signs of aortic valve disease progression.

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