Outcomes of Partial Atrioventricular Septal Defect Repair in Children

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ABSTRACT

Introduction: Repair of the partial atrioventricular septal defect (pAVSD) has an excellent survival but re-operation is still an issue. Left atrioventricular valve regurgitation (LAVVR) is the most common reason for reoperation. This study aimed to retrospectively analyze the results of patients undergoing pAVSD repair to determine the morbidity and mortality rates.

Patients and Methods: Follow-up data of a total of 25 patients younger than 18 years of age, who underwent pAVSD at our centre within the period from 2011 to 2019, were obtained from hospital records. The incidence of Down syndrome, death, complications and reoperation rates were analyzed.

Results: Mean age at operation was 4.64 years. Preoperative LAVVR grade was found to be relatively high in patients who underwent surgery at an older age ($p= 0.027$). Two patients (8%) had Down syndrome. Follow-up ranged from 2 months to 96 months (mean: 33.48 months). There was no perioperative mortality. Arrhythmia were observed in 4 patients (16%) after the operation. One patient (4%) had complete AV block requiring permanent pacemaker implantation. The rate of postoperative LAVVR was observed to increase as the time elapsed after the operation increased. The presence of Down syndrome was observed to be protective against postoperative LAVVR ($p< 0.01$). Two patients (8%) had reoperation for LAVVR. The mean time from the initial operation to reoperation was six years. The absence of Down syndrome was found to be a risk factor for reoperation ($p< 0.01$).

Conclusion: Repair of pAVSD is performed with low mortality and favourable outcomes. Most common reason for reoperation following the repair of pAVSD is LAVVR and regurgitation rate increases as the time elapsed following the initial operation increases. Furthermore, preoperative LAVVR rate becomes higher as the operation age of the patient increases. Down syndrome is protective in terms of postoperative LAVVR and its absence is a risk factor for reoperation. High reoperation rates suggest the requirement of close follow-up.

Key Words: Left atrioventricular valve; regurgitation; reoperation.

Çocuklarda Parsiyel Atriyoventriküler Septal Defekt Tamir Sonuçları

ÖZ

Giriş: Parsiyel atriyoventriküler septal defektin (pAVSD) onarımı sonrası sağkalım mükemmel olsa da reoperasyon halen sorun oluşturmaktadır. Sol atriyoventriküler kapak yetersizliği (SAVKY), reoperasyonun en sık nedenidir. Bu çalışmada, pAVSD onarımı yapılan hastaların verilerini retrospektif olarak analiz ederek morbidite ve mortalite oranlarını belirlemek amaçlanmıştır.

Hastalar ve Yöntem: Merkezimizde 2011-2019 yılları arasında, pAVSD nedeniyle tamir yapılan 18 yaşından küçük toplam 25 hastanın takip verileri hastane kayıtlarından elde edildi. Down sendromu insidansı, ölüm, komplikasyonlar ve reoperasyon oranları analiz edildi.

Bulgular: Ameliyat sırasında ortalama yaş 4.64 yıl idi. Daha geç yaşta ameliyat edilen hastalarda ameliyat öncesi SAVKY derecesi göreceli olarak yüksek bulunmuştur ($p< 0.05$). İki hastada (%8) Down sendromu vardı. Takip süresi 2-96 ay arasındaydı (ortalama: 33.48 ay). Perioperatif mortalite yoktu. Dört hastada (%16) ameliyat sonrası aritmi görülmüştür. Bir hastada (%4) kalıcı kalp pili implantasyonu gerektiren tam atriyoventriküler blok gelişmiştir. Operasyon sonrası geçen süre arttıkça postoperatif SAVKY oranının arttığı gözlenmiştir. Down sendromunun varlığının postoperatif SAVKY'ye karşı koruyucu olduğu görülmüştür ($p< 0.05$). İki hasta (%8) SAVKY nedeniyle reoperasyon geçirmiştir. İlk ameliyattan reoperasyona kadar geçen süre ortalama altı yıldır. Down sendromu, reoperasyon için risk faktörü olarak bulunmuştur ($p< 0.05$).

Sonuç: PAVSD'nin onarımının mortalitesi düşük ve sonuçları iyidir. PAVSD tamirini takiben reoperasyon için en sık neden SAVKY'dir ve ilk operasyonun ardından geçen süre arttıkça yetersizlik oranı artar. Ayrıca hastanın ameliyat yaşı arttıkça ameliyat öncesi SAVKY oranı da yükselmektedir. Down sendromu postoperatif SAVKY açısından koruyucu iken reoperasyon için risk faktörüdür. Reoperasyon oranının yüksek olması yakın takip gerekliliğini ortaya koymaktadır.

Anahtar Kelimeler: Reoperasyon; sol atriyoventriküler kapak; yetersizlik.

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INTRODUCTION

The prevalence of atrioventricular septal defect (AVSD) is five per 10.000 children[1]. Partial AVSD (pAVSD) constitutes 25% of all AVSDs and is characterized by primum atrial septal defect (ASD) and cleft in the left atrioventricular (AV) valve[2]. There is a common AV junction in pAVSD; however, the right and left AV orifices are separate. The left AV valve in pAVSD is a trileaflet valve and differs from the normal mitral valve in terms of leaflet, chordae and papillary muscle morphology[3]. Surgical repair of pAVSD is usually performed in pre-school age.

Although the early-term mortality rate is low and the long-term survival rate is high, the reoperation rate at 30-year follow-up is 25%[4]. Long-term survival significantly decreases in patients requiring reoperation[5]. The main reason for reoperation is left AV valve regurgitation (LAVVR) with a rate of 12.1%[4]. Lower body weight, absence of Down syndrome and absence of mitral cleft repair are reported to be risk factors for reoperation[6].

This retrospective study aimed to present demographic characteristics, grade of preoperative and postoperative LAVVR, length of stay in the hospital and intensive care unit (ICU) in the postoperative period, complications, and reoperation rates due to LAVVR among patients undergoing pAVSD repair, and to analyze the correlation of Down syndrome with LAVVR.

PATIENTS and METHODS

A total of 25 patients undergoing surgical repair due to pAVSD at our center within the period from September 2011 to December 2019 were retrospectively analyzed. The data were obtained by examining patient records. Approval for this study was obtained from the Clinical Research Ethics Committee of Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital (Decision no: 2021/18, Date: 23.02.2021).

The diagnosis of pAVSD was established by transthoracic echocardiography (TTE). Diagnosis for the repair of pAVSD was considered sufficient for a surgical indication and the timing of surgery was after infancy. In the preoperative and postoperative period, the left AV valve was evaluated via TTE. Severity of mitral regurgitation was graded according to the classification of Sellers et al.[7]: I + (mild), 2 + (moderate), 3 + (moderate to severe) and 4 + (severe). The indication for reoperation in terms of intervention to the left AV valve was the presence of grade 4 regurgitation.

Median sternotomy was performed in all patients. During cardiopulmonary bypass, the patient was cooled to 34°C and cardiac arrest was achieved with antegrade cardioplegia. The cleft in the left AV valve was closed completely from the annulus to the point of chordal insertion with the intermittent suture technique. Primum ASD was closed with an autologous pericardial patch leaving the coronary sinus in the right atrium.

Statistical Analysis

Continuous variables are summarized as a median with corresponding 25th (Q1) and 75th (Q3) percentiles, and comparisons between patient groups were based on the Chi-Square test. If Asymptotic Significance (p-value) for the Pearson Chi-Square statistic is less than 0.05, there is a relationship between the variables. Incident outcomes of primary interest consisted of all cause deaths, first reoperation, left atrioventricular valve regurgitation and arrhythmias.

RESULTS

Of the 25 patients, 10 (40%) were female and 15 (60%) were male. The median age at operation was 4 years (Q1, 2 years; Q3, 7.5 years). Two patients (8%) had Down syndrome. The length of stay in the ICU and hospital ranged from 1 day to 16 days (median, 1 day; Q1, 1 day; Q3, 2 days) and 3 to 21 days (median, 7 days; Q1, 5 days; Q3, 10.5 days), respectively.

Postoperative complications were observed in 6 patients (24%). Arrhythmia developed in four patients, two of which were supraventricular tachycardia while the other two were AV block. One of the patients who developed AV block had first-degree AV block and returned to sinus rhythm on the postoperative 15th day. The other patient with AV block had complete AV block and permanent pacemaker implantation was performed on the postoperative 16th day. Staphylococcus aureus growth was identified in the blood culture of one patient and was treated with intravenous antibiotic therapy. Pneumothorax developed in one patient and was treated with tube thoracostomy.

The median follow-up duration was 21 months (Q1, 5 months; Q3, 63 months). The median grade of preoperative LAVVR was 2 (Q1, 2; Q3, 4) and it was also 2 (Q1, 1; Q3, 3) at the last postoperative follow-up. The mean follow-up duration was 48.37 months in patients with grade ≥ 3 LAVVR whereas it was 26.47 months in patients with grade < 3 LAVVR. There was no perioperative mortality.

The mean preoperative LAVVR grade was 2 in patients with Down syndrome and 2.52 in those without. The mean postoperative LAVVR grade was 1.5 in patients with Down syndrome and 2 in those without.

In the postoperative period, 2 (8%) patients underwent reoperation due to grade 4 LAVVR. These patients did not have Down syndrome. The mean interval between the primary repair and the subsequent reoperation was six years.

The general characteristics of the patients are shown in Table 1, and the comparison of variable data is shown in Table 2.

Table 1. General characteristics of patients

Patients	Gender	Age (years)	Down syndrome	ICU/Hospital stay (days)	Complication	PREOP LAVVR grade	POSTOP LAVVR grade	Followup period (months)
1	Male	6		1/6		4	1	22
2	Male	3	+	4/5		1	1	8
3	Male	1		2/7		2	1	17
4	Male	4		1/5		2	1	6
5	Male	9		1/3		1	1	11
*6	Female	11		1/19	SVT	2	4	45
7	Male	1		1/7		1	1	2
8	Female	2		2/9	SVT	4	2	2
9	Male	5		1/4		4	1	2
10	Female	10		1/5		2	1	28
11	Male	7		1/6		3	1	4
12	Male	2		1/6		2	3	63
13	Male	8		1/19	Infection	2	2	93
14	Female	4		1/7		3	1	96
15	Female	9		1/21	First-degree AV block	4	3	77
*16	Female	2		5/12	PNX	1	4	93
17	Male	11		2/7		2	3	2
18	Female	1		2/7		4	3	32
19	Male	7		1/6		1	3	12
20	Male	2		2/6		2	3	63
21	Female	1	+	9/17		3	2	27
22	Male	2		2/5		4	2	21
23	Female	2		2/2		3	2	14
24	Female	2		16/18	Complete AV block	4	2	2
25	Male	4		2/7		4	1	95

AV: Atrioventricular, ICU: Intensive care unit, LAVVR: Left atrioventricular valve regurgitation, PNX: Pneumothorax, POSTOP: Postoperative, PREOP: Preoperative, SVT: Supraventricular tachycardia.

Table 2. Comparison of variable data

Mean age at operation in patients with preoperative LAVVR grade of ≥ 3: 5.63 years[1]	Mean age at operation in patients with preoperative LAVVR grade of < 3: 4.18 years	p= 0.027
Mean follow-up duration in patients with postoperative LAVVR grade of ≥ 3 at the last follow-up: 48.37 months[2]	Mean follow-up time in patients with postoperative LAVVR grade of 3 at the last follow-up: 26.47 months	p= 0.884
Number of reoperated patients with Down syndrome: 0[3]	Number of reoperated patients without down syndrome: 2	p< 0.01
Mean postoperative LAVVR rate at the last follow-up in patients with Down syndrome: 1.5[4]	Mean postoperative LAVVR rate at the last follow-up in patients without Down syndrome: 2	p< 0.01

LAVVR: Left atrioventricular valve regurgitation.

[1] Preoperative LAVVR grade was found to be relatively high in patients undergoing surgery at an older age (p< 0.05).

[2] Postoperative LAVVR was observed to increase as the time elapsed after the operation increased.

[3] Absence of Down syndrome was found to be a risk factor for reoperation (p< 0.05).

[4] Presence of Down syndrome was observed to be protective against postoperative LAVVR (p< 0.05)

DISCUSSION

The early mortality rate after pAVSD repair is 1.2%, and the survival rate at 30-year follow-up has been reported to be 94%[4]. Performing pAVSD repair in infancy has been found to increase reoperation rate and it was determined that early mortality was higher in patients with lower age[8,9]. All patients included in the present study were operated after infancy, and the mean age of operation was 4.64 years. The mean operation age of patients with preoperative LAVVR grade of ≥ 3 was 5.63 years, while it was 4.18 years in those with grade < 3 LAVVR. So preoperative LAVVR grade was found to be relatively high in patients undergoing surgery at an older age (p= 0.027). According to a study; children with preoperative weight z-scores of -1.0 or less repaired before 18 months had the most catch-up growth, without increasing their risk of significant LAVVR or other morbidities and without prolonging ventilation, intensive care, or hospital days[10]. Another study found that; results of partial AVSD repair at a median age of 18 months are excellent and operating at this age is not associated with increased mortality, reoperation, or LAVVR[11]. We recommend operation by the age of 18 months, even the patient is asymptomatic, to prevent progression of LAVVR.

The rate of grade ≥ 3 LAVVR was reported to be 41% and 51% one year and five years after pAVSD repair, respectively[12]. In the present study, this rate was 28% after one year and 32% after five years. In the last follow-up, the mean follow-up duration was 48.37 months in patients with grade ≥ 3 LAVVR whereas it was 26.47 months in patients with grade < 3 LAVVR. Postoperative LAVVR was observed to increase as the time elapsed after the operation increased (p= 0.884). Furthermore, the median values of the preoperative and postoperative LAVVR grades of the patients at the last follow-up were found to be the same. These findings show that postoperative close follow-up of the patients is important.

Surgical repair techniques have largely depended on primary closure of the residual cleft in the anterior leaflet. Better survival rates were reported with the closure of the cleft in the left AV valve[12]. Moreover, incomplete closure of the left AV valve cleft was found to be a predictive factor for postoperative LAVVR[13]. The cleft has been closed in all of our patients undergoing pAVSD repair. However simple cleft closure may be ineffective and LAVV patch augmentation can be used as an additional technique. Analysis of factors impacting durability of valve repair determined that cleft closure alone at the time of repair posed a higher risk for subsequent valve replacement than if leaflet patch augmentation was performed concomitantly (40% versus 0%)[14]. Due to another study; freedom from reoperation at 10 years was 59.4% in cleft closure group whereas, in the cleft patch augmentation group, it was 92.3%[15]. We recently include LAVV patch augmentation technique in the repair of all patients independently of aortomitral angle, in order to enhance mobility and durability of LAVV. We did not determine systolic anterior motion in these patients afterwards.

LAVVR is the most common reason for reintervention, the reoperation rate ranges from 9% to 12%[6,4,12]. In the present study, 2 (8%) patients underwent reoperation due to grade 4 LAVVR in the postoperative period. Presence of grade ≥ 3 LAVVR before pAVSD repair has been reported as a risk factor for reoperation[13]. In the present study, none of the patients with reoperation had grade ≥ 3 LAVVR before the first pAVSD repair operation. Other common causes of reoperation are left ventricular outflow obstruction (4.8%) and residual ASD (2%)[4]. In the present study, none of the patients underwent reoperation due to these two reasons. In a study involving patients undergoing reoperation for both left ventricular outflow tract stenosis and LAVVR, the mean time between pAVSD repair and reoperation was found to be 10 years[5]. In the present study, the mean time between the pAVSD repair and reoperation LAVVR was six years. During the reoperation for LAVVR, valve repair should be preferred instead of valve replacement, if possible, to avoid the risk of morbidity due to anticoagulation, the need for repeat valve replacement and cardiomyopathy[14]. In the present study, valve repair was performed in both of the patients undergoing reoperation. We use mitral valvulotomy and/or mitral annuloplasty techniques depending of the mitral lesion during reoperation.

Among individuals with Down syndrome, 42% have congenital heart disease, 6% of which are pAVSD[16]. In the present study, Down syndrome was present in two patients (8%). The absence of Down syndrome is a risk factor for reoperation[8]. In this study, none of the patients undergoing reoperation had Down syndrome, and the absence of Down syndrome was observed to be a risk factor for reoperation (p< 0.01). There was no difference in terms of surgical technique between Down patients and non-Down patients. Furthermore, the postoperative LAVVR grade at the last follow-up was higher in patients without Down syndrome than in those with Down syndrome (p< 0.01). Presence of Down syndrome was observed to be protective in terms of LAVVR.

Arrhythmia may develop after pAVSD repair. The rate of permanent pacemaker implantation due to complete AV block has been reported to be 3.2%[4]. In the present study, postoperative pacemaker implantation was performed only in 1 patient (4%) for the same reason. In one study, supraventricular arrhythmias were observed in 16% after the operation and supraventricular arrhythmias were found to increase with increasing age at primary operation[12]. We found that supraventricular arrhythmia, which did not affect hemodynamics, developed in 2 patients (8%), and the age of the patients was eleven and two years.

In a study involving patients diagnosed with congenital heart disease, the rates of patients who were not followed after the age of 6, 13 and 18 were found to be 28%, 47% and 61%, respectively[17]. In the present study, the mean follow-up period was 33.48 months.

The major limitation of this study is its retrospective design. Low complication and mortality rates may have reduced the accuracy of statistical analysis. Long term follow-up data were not available in all patients.

CONCLUSION

Surgical management of partial AV canal defects should be electively repaired in early childhood. When left AV valve regurgitation is severe, repair should be performed earlier to prevent further impairment of valve function. Down syndrome appears to be a protective factor for LAVVR and reoperation. Despite low mortality, reoperation and long-term morbidity remain an issue. Longitudinal follow-up that mainly focuses on LAVVR is required in all patients. Survival is excellent; however, we believe that follow-up will reduce long-term morbidity due to reoperation risk.

Ethics Committee Approval: Approval for this study was obtained from the Clinical Research Ethics Committee of Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital (Decision no: 2021/18, Date: 23.02.2021).

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