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Surgical outcome of atrioventricular septal defect: A 10 years review.

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Abstract

Background: Surgical management of patients with Atrioventricular Septal Defect (AVSD) has advanced over the last decades. The aims of this study were to evaluate the early and late outcomes in patients undergoing operation at a single heart center.
Methods: Between April 2006 and April 2016, 337 consecutive patients underwent Atrioventricular Septal Defect (AVSD) repair at the Shaheed Rajaie Heart Center. The patients were subtyped as 73 (21.7%) partial, 67 (19.9%) transitional and 197 (58.5%) complete.
Results: At the time of preoperative evaluation, there were 39 cases (11.7%) with moderate to severe or severe Left Atrioventricular Valve Regurgitation (LAVVR). Previous Pulmonary Artery banding (PA banding) had been performed in 144 (42.7%) patients and associated malformations were found in 115 (34.1%). Type of surgical repair was single patch 193 (57.3%), double patch 144 (42.7%), and single atrial septal defect patch with primary ventricular septal defect closure in TAVSD 64 (95.5%). 141 patients (71.6%) with CAVSD underwent definitive early repair by using a two patch technique and complete cleft closure. Annuloplasty and/or commissuroplasty were performed for 60 (17.8%) patients and debanding was also implemented in all patients. Mean hours for ventilation were determined as 26.8 ± 2.8. Mean days for intensive care unit was defined to be 4.4 ± 0.2, and total hospitalization time was as 26.8 ± 0.5. A 1 month, 21.9% (72 of 328) had ejection fraction less than 55%; 33 (10.9%) had more than moderate LAVVR; 73 (24.2%) had residual Ventricular Septal Defect (VSD), followed by 14 cases (4.6%) with residual Atrial Septal Defect (ASD) and 2 cases (0.6%) with Left Ventricular Outlet Tract Obstruction (LVOTO).
Conclusions: Definitive early repair for AVSD can be performed with acceptable results. Despite a complete cleft closure, LAVVR remains the most common residual defect that more frequently required reoperation in the future.

Keywords: Atrioventricular, Septal defect, Cardiac surgery, Outcomes.

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Introduction

Atrioventricular Septal Defects (AVSD) is a complex congenital disease that includes a range of *Ventricular* Septal *Defect* (VSD) and *Atrial* Septal *Defect* (ASD). The disease comprises 17-17% of congenital heart disease and includes the subgroups inclosing partial, transitional and completes [1]. This defect is present in 50-50% of cases in patients with Down syndrome (Trisomy 21) [2]. AVSD is characterized by atrioventricular canal defects or endocardial cushion defects [3]. First successful repair of AVSD was performed by Lillehi et al. using the Cross Circulation method, and by sewing the atrial septum to the lateral wall of the ventricular septum [4]. The initial experience of AVSD treatment was associated with high mortality and morbidity such as heart block, ventricular valve failure and stenosis under the aortic valve, but a good understanding of lesion anatomy and improved surgical technique resulted in improved results [5].

Significant progress was made in 1958 when Manrice described the location of the branch of his in AVSD [6]. Rastelli et al. at Myo clinic had introduced the classification system in 1966 and emphasized on the cardiovascular spectrum consequently advised to use different technique for various anatomical findings [7]. In 1962, Malonogo et al. described the Single-Patch method [8]. Moreover, the Two-Patch method has been first suggested by Trusler in 1975 [9]. Recently, Nunn reported from the Sydney Children's Hospital that there was no need for a patch to close the ventricular wall [10]. In spite of its initial success in the two decades after the successful treatment of this anomaly, surgery has often been associated with a high mortality rate (25%).

Furthermore, the amount of residual defects such as mitral valve, shunt, and heart block failure were high [11]. However, improvements in precise preoperative diagnosis, better understanding of surgical anatomy, progression in surgical technique, protection and post-operative care (including treatment of pulmonary pressure events) contribute to increased survival rates and decreasing re-surgical intervention [12-18].

However, in the past decade, mortality has not been significantly improved and varies 21.7 to 8.7%, and the rate of re-operation varied from 6.4 to 16.6% [18-23]. Primary repair reduces the risk of premature death and obstructive pulmonary vascular disease, but there are patients who, due to lack of response to medication for heart failure, should undergo surgery in the first year of life. Because the risk of side effects of medication preservation is considerable over the long term [18]. It is worth noting that mostly one-stage operation is preferred but sometimes palliative therapy (the PA-banding) and two-stage.

Some surgeons believe that palliative action still plays an important role in the treatment of children or low weight children because of the fact that there is concern about the fracture of the valve tissue in these, there is also a technical problem in their surgery [15-17,24]. Despite a definitive surgical repair, in North America and Europe, between 9% and 17% of patients with AVSD have a significant LAVVR lesion at discharge from the hospital. Although, TEE is done for them during surgery and this failure often develop and develop up to three years after surgery [19,25,26].

This impairment not only increases the risk of "reoperation", but also can increase the mortality rate and even the cost of hospitalization, by hemodynamic impairment [14,27-29]. In addition, AVSD reveals a wide range of heart and non-cardiac diseases, so it requires careful preoperative examination and multiple surgical procedures with different treatment options. On the other hand, due to multiple heart defects these patients have specific problems before surgery and the possibility of high pulmonary resistance is premature. Therefore, they often undergo surgery early in the first year and due to the effect of heart failure on other organs and the lack of development of other organs, the risk of operation in these patients is relatively high compared with other heart diseases. This issue causes them to be examined early in the event of heart failure symptoms and undergo surgery in the best condition. To make the right decisions, there is a need for close cooperation between the heart, children, anesthesia, perfusion, surgery and post-operative care teams.

Therefore, this issue and its necessity require that the issue be assessed in an important referral center in a ten-year period and patient information be collected based upon medical records, pre-operative clinical examinations, surgical procedures and post-operative care. Subsequent studies can be then carried out on the basis of specific goals including age, sex, type of operation, operation results, residual atrio-ventricular valve failure, and residual *defects* in the atrial and ventricular *walls*.

Moreover, early and late mortality rates can be determined and effective factors in mortality and needs to re-operation will be identified, and then the results can be compared with advanced countries. Subsequently, the results of the working group should be evaluated scientifically in order to identify the deficiencies in the course of treatment compared with the results of other countries for eliminating these disadvantages. By reducing mortality and surgical complications, this will be taken a step towards improving the quality of treatment, where eliminating possible bugs will be the centre's scientific advancement.

Material and Methods

Ethical considerations

Information is collected on the basis of records in the records which is only used for scientific medical examination and maintained in compliance with ethical and medical principles.

Patients, study design, and definition

Patients undergoing CAVSD surgical repair at Tehran's Shahid Rajaee Heart Center were selected for a retrospective study (Retrospective Cohort). After the ethical approval of the Research Ethics Committee, they entered the study between April 2006 and April 2016.

Patients with partial AVSD were those who had two orifices with a cleft in Left Atrioventricular Valve (LAVV). Patients with transitional AVSD were belonged to those with a small VSD defect and two whole openings with left atrioventricular valves. Patients with complete AVSD were those with a large ventricular hole with common atrioventricular valves. Over a period of 10 y, a total of 177 women, 160 men with atrioventricular septal defect underwent surgery, of which 73 (21.7%) had PAVSD, followed by TAVSD (67 patients 19. 9%), and CAVSD (197 patients, 58.5%). A few patients (7.3%) had a history of PA banding, which underwent surgery in two stages, and 193 patients (57.3%) were undergoing one stage surgery. The age of the patients varied from 2 months to 60 y (mean 5.2 ± 63.5). Down syndrome was found in 118 cases (35%) and coronary anomalies in 115 patients (34.1%) (Table 1).

Data were collected from patients' records of the hospital archives including findings of echocardiography, cardiac catheterization, surgery, and follow-up (in all patients). Preoperative data was available for severity of defects in the right and left atrioventricular valve in 334 patients (99.1%). The evaluation of the right and left atrioventricular valve defect was recorded based on the criteria (Grade 0 to 6), and moderate to severe and severe defect was considered as significant. The average pulmonary arterial pressure above 25 mmHg was considered to be a high pulmonary artery pressure in 117 patients out of 237 cases (49.4% %) while 46 cases (13.6%) had shown median pulmonary artery pressure above 40 mmHg. Reconstructive surgery was performed based upon use of incision, median sternotomy and both bicaval venous cannulation using CPB with moderate hypothermia.

Table 1. *Patient demographics.*

Variables	PAVSD (73; 21.7%)	TAVSD (67; 19.9%)	CAVSD (197; 58.5%)	P value
Age	0			
0-6 months	2 (2.7)	1 (1.5)	9 (4.6)	
6-12 months	3 (4.1)	7 (10.6)	37 (18.9)	
1-5 y	33 (45.2)	38 (57.6)	126 (64.3)	
>5 y	35 (47.9)	20 (30.3)	24 (12.2)	
Weight at operation (kg; mean ± SE)	24.3 ± 2.4	19.5 ± 2.4	11.8 ± 0.6	0
Male to female ratio	34/39	26/41	100/97	0.235
CAVSD Rastelli type				
Type A	-	-	107 (54.3)	
Type B	-	-	72 (36.5)	
Type C	-	-	18 (9.1)	
Associated anomalies	0.11			
Down syndrome	9 (12.3)	17 (25.4)	92 (46.7)	0
PDA	6 (8.2)	13 (19.4)	41 (20.8)	0.052
LSVC	2 (2.7)	6 (9.0)	14 (7.1)	0.291
Unroofed coronary sinus	1 (1.4)	2 (3.0)	5 (2.5)	0.799
Common atrium	2 (2.7)	6 (9.0)	5 (2.5)	0.053
Double orifice mitral valve	3 (4.1)	1 (1.5)	3 (1.5)	0.388
Parachute mitral valve	4 (5.5)	3 (4.5)	3 (1.5)	0.169
Right aortic arch	0 (0.0)	0 (0.0)	2 (1.0)	0.489
TOF	0 (0.0)	0 (0.0)	12 (6.1)	0.012
TOF, Right aortic arch	0 (0.0)	0 (0.0)	1 (0.5)	0.7
DORV	0 (0.0)	0 (0.0)	1 (0.5)	0.7
Coarctation of aorta	0 (0.0)	0 (0.0)	1 (0.5)	0.7
Interrupted IVC	1 (1.4)	0 (0.0)	1 (0.5)	0.557
PS	2 (2.7)	0 (0.0)	9 (4.6)	0.184
Ebstein anomaly	1 (1.4)	0 (0.0)	0 (0.0)	0.163
Subvalvular AS	1 (1.4)	0 (0.0)	0 (0.0)	0.163
Dextrocardia	1 (1.4)	0 (0.0)	2 (1.0)	0.661
PAPVC	2 (2.7)	0 (0.0)	0 (0.0)	0.026
Single ostium coronary artery	1 (1.4)	0 (0.0)	0 (0.0)	0.163
Absent RSVC	0 (0.0)	1 (1.5)	0 (0.0)	0.133
Severe AI	0 (0.0)	0 (0.0)	1 (0.5)	0.7
VSD	1 (1.4)	0 (0.0)	3 (1.5)	0.602
Cor-triatriatum	1 (1.4)	0 (0.0)	1 (0.5)	0.557

Situs inversus	0 (0.0)	0 (0.0)	1 (0.5)	0.7
Previous PA banding	1 (1.4)	4 (6.0)	64 (32.5)	0
Previous PDA closure and PA banding	7 (9.6)	11 (16.4)	57 (28.9)	0
Previous systemic to pulmonary shunt	0 (0.0)	0 (0.0)	2 (1.0)	0.489

PAVSD=Partial atrioventricular septal defect; TAVSD=Transitional atrioventricular septal defect; CAVSD=Complete atrioventricular septal defect; PDA=Patent ductus arteriosus; LSVC=Left superior vena cava; TOF=Tetralogy of fllot; DORV=Double outlet right ventricle; IVC=Inferior vena cava; PS=Pulmonary stenosis; AS=Aortic stenosis; PAPVC=Partial anomalous pulmonary venous connection; RSVC= Right superior vena cava; AI=Aortic insufficiency; VSD=Ventricular septal defect; PA=Pulmonary artery; SE=Standard error.

Heart protection was conducted with the help of a cold cardioplegia solution (40°C) or cold blooded cardioplegia and topical hypothermia using ice slush. The cleft of the left atrioventricular valve was completely repaired in most patients and was partially closed in a small number due to the possibility of stenosis. It was performed in 20 patients (5.9%). Annuloplasty with proline 6-0 was performed in 40 patients (11.9%). One patient needed replacement of the mitral valve with a biological valve [21]. In 64 patients with TAVSD (98.5%), a small interstitial ventricular septum was seamlessly sewn with 6-0 proline. After left atrioventricular valves repair, Cold Saline was injected into the left ventricle for testing. 141 patients with CAVSD (71.6%) had double-patched surgical repair, and 56 cases (28.4%) were repaired by Single Patch method. AVSD was restored in all patients with autologous pericardial patches and coronary sinuses were directed to the left atrium in 188 patients (55.8%) to avoid atrioventricular node damage. Conductive disorder occurred in 11 cases (8.5%), and Complete Heart Block (CHB) was present in 6 cases. After surgery, in 5 cases epicardial pacemaker was implanted and in the unroofed coronary sinus, the LSVC hole was directed to the right atrium with a patch.

Early death was defined as death in the first thirty days after the operation and death of the hospital. Early reoperation was considered as a surgical procedure in the first thirty days after surgery. Perioperative infection was those that were involved in the prognosis and included septicemia, septic shock, and endocarditis and mediastinitis and topical infection with sepsis. Patient follow-up information was obtained from medical records after discharge. The mean follow-up time in patients was 1.8 ± 24.9 months and varied from one month to 124 months, while 49 patients (27.8%) were not followed up after surgery. In 33 patients (9.8%), there was a significant (moderate to severe) failure in the left atrioventricular valve at the time of discharge which essential actions were performed for 7 patients (21.2%). There was VSD in a patient who was undergoing surgery again. Late deaths were seen in only three cases due to the inability to complete follow up of patients.

Data analysis

Statistical analysis was performed using SPSS version 17 (SPSS Inc, Chicago USA). All results were expressed as Mean ± SE. Statistical analyzes were performed using X^2 test, Fisher exact test and Independent sample T-test and One-way ANOVA. The value of $P<0.05$ was considered as significant level.

Results

This study was performed on 337 patients who included AVSD subgroups, 73 cases were belonged to PAVSD (21.7%), followed by 67 cases of TAVSD (19.9%) and 197 cases of CAVSD (58.5%). From CAVSD patients, 107 patients had Rastelli type A (54.3%), followed by 72 individuals with Rastelli type B patients (36.5%) and 18 Patient with Rastelli type C (9.1%), Table 1. The mean age of patients was 5.2 ± 63.5 months (ranged from 2 months to 60 y). Average age for patients with PAVSD was recorded to be 8 ± 124.7 ± 17.8 months (ranged from 2 months to 60 y). Moreover, average age for patients with TAVSD and CAVSD were 73.6 ± 11.9 y (ranged from 2 months to 40 y) and 73.3 ± 3.1 months (ranged from 4 months to 36 y), respectively. The mean overall weight of patients was 16 ± 0.8 kg (range 2.9 to 86 kg), mean weight for PAVSD was recorded as 24.3 ± 2.4 kg (range 2.9 to 85 kg), For patients with TAVSD and CAVSD, it was set at 2.4 ± 19.5 (range 4.7 to 86 kg) and 11.8 ± 0.6 kg (range 4.4-72 kg), respectively. Of the total number of patients, 177 cases (52.5%) were females and 160 cases were (47.5%) were male where the male to female ratio was 9.0. Down syndrome was reported in 118 patients (46.7%) and high rates (92 cases; 47.5%) were prevalent in CAVSD. Other cardiac anomalies included 60 cases of PDA (17.8%), 22 cases of LSVC (6.5%), 8 cases of unroofed coronary sinus (2.4%), 13 common atrium (3.9%), 7 cases of double orifice mitral valve (2.1%) and 10 cases of parachute mitral valve (3%), (Table 2). The left *atrioventricular valve* defect was observed in 327 patients (97%) and right *atrioventricular valve* defect was seen in 326 cases (96.7%). The total score for left *atrioventricular valve* defect was as follows: mild (74 cases; 22.2%), mild to moderate (24 cases; 7.2%), moderate (170 cases; 50.9%), moderate to severe (21 cases; 6.3%) and severe (38 cases; 11.4%). The total score for right *atrioventricular valve* defect was as follows: mild state (86 cases; 25.7%), mild to moderate (23; 6.9%), moderate (178 cases; 53.3%), moderate to severe (13; 3.9%) and severe (26 cases; 7.8%).A total of 13 patients (6.7%) suffering from CAVSD had Tetralogy of Fallot (TOF) that narrowing Right Ventricular Outflow Tract (RVOT) was improved, where there was no early mortality.

Table 2. *Pre-operative clinical data.*

Variables	PAVSD (73; 21.7%)	TAVSD (67; 19.9%)	CAVSD (197; 58.5%)	P value
G. LAVVR	0.075			
None	2 (2.7)	0 (0.0)	1 (0.5)	
Trivial	1 (1.4)	0 (0.0)	3 (1.5)	
Mild	12 (16.4)	14 (20.9)	48 (24.7)	
Mild to moderate	7 (9.6)	4 (6.0)	13 (6.7)	
Moderate	32 (43.8)	31 (46.3)	107 (55.2)	
Moderate to severe	7 (9.6)	7 (10.4)	7 (3.6)	
Severe	12 (16.4)	11 (16.4)	15 (7.7)	
G. RAVVR	0.389			
None	1 (1.4)	0 (0.0)	1 (0.5)	
Trivial	3 (4.1)	1 (1.5)	2 (1.0)	
Mild	23 (31.5)	10 (14.9)	53 (27.3)	
Mild to moderate	4 (5.5)	6 (9.0)	13 (6.7)	
Moderate	34 (46.6)	39 (58.2)	105 (54.1)	
Moderate to severe	2 (2.7)	5 (7.5)	6 (3.1)	
Severe	6 (8.2)	6 (9.0)	14 (7.2)	
EF (mean ± SE)	56.0 ± 0.7	57.3 ± 0.8	59.8 ± 0.6	0.001
PA pressure (mean ± SE)				
With PA banding	22.0 ± 4.9	28.8 ± 4.2	31.6 ± 1.7	0.207
Without PA banding	23.6 ± 1.1	26.1 ± 1.4	41.0 ± 3.1	0
SVo_2	87.9 ± 0.9	87.4 ± 0.9	86.3 ± 0.7	0.409

G=Grade; LAVVR=Left atrioventricular valve regurgitation; RAVVR=Right atrioventricular valve regurgitation; EF=Ejection fraction; PA=Pulmonary artery; SVo_2=Mixed venous oxygen saturation.

Intraoperative and postoperative information

The mean CPB time was 2.4 min (118 ± 62.2 min; ranged from 53 to 302 min) and the mean aortic clamp time was defined as 79.5 ± 1.7 (ranges from 25 to 183 min). In comparison, PAVSD had the shortest pump time (CPB) and clamping aorta compared to CAVSD.In all patients with PAVSD, the Single Patch of autologous pericardial patch *was used.* In 64 patients with VSD (95.5%), the small VSD was separately sewn and the ASD was sewn with a pericardial patch. For patients with CAVSD, the Double Patch method was used in 141 cases (71.6%) and the Single Patch method was applied in 56 cases (28.4%) (Table 3). The cleft repair of right *atrioventricular valve* was complete in 331 patients) (98.2%) and was closed relatively in 5 patients (1.5%), where the cleft repair of left *atrioventricular valve* was completely closed in 11 cases (3.3%) and 110 cases (32.6%) were relatively closed. Annuloplasty in left *atrioventricular valve* was performed in 40 (11.8 %) and*Chemisphere* Palesty *in* left *atrioventricular valve* was implemented in 20 patients (5.9%). Neochordae was used in 5 patients (1.4%), and the splitting of *papillary muscle formitral valve* was performed in 4 patients (1.1%) using Parachute Mitral Valve. In the post-operative echoscopy, left and light *atrioventricular valve* defects were seen in 305 patients (90.5%) and 227 patients (82.1%), respectively. The left *atrioventricular valve* defects was classified as mild (137 cases; 41.8%) mild to moderate, (49 cases; 14.9%), moderate (96 cases; 28.3%), moderate to severe (17 cases; 5%) and severe (6 cases; 1.8%). Its rate was higher in CAVSD as compared to PAVSD. Moreover, the right *atrioventricular valve* defects in mild state was found in 173 cases (52.7%), followed by mild to moderate (34 cases; 10.4%), moderate 54 cases; 16.5%), moderate to severe (10 cases; 3%) and severe (6 cases; 1.8%). The rate of *atrioventricular valve* defects above the moderate level was belonged to the six cases after surgery

(12.8%) at the age of one year, as well as 27 cases (9.3%) at older age.

Table 3. *Intraoperative clinical data.*

Variables	PAVSD (73; 21.7%)	TAVSD (67; 19.9%)	CAVSD (197; 58.5%)	P value
Bypass data				
CPB time (min)	92.8 ± 3.4	102.8 ± 3.4	133.8 ± 3.0	0
ACC time (min)	58.9 ± 2.7	66.2 ± 2.7	91.9 ± 2.2	0
Surgical technique	0			
Single patch (no, %)	73 (100)	64 (95.5)	56 (28.4)	
Double patch (no, %)	0 (0.0)	3 (4.5)	141 (71.6)	
Closure of LAVVC (no, %)	0.315			
Complete	70 (95.9)	66 (98.5)	195 (99.0)	
Partial	2 (2.7)	1 (1.5)	2 (1.0)	
None	1 (1.4)	0 (0.0)	0 (0.0)	
Closure of RAVVC (no, %)	0.015			
Complete	1 (1.4)	5 (7.5)	5 (2.5)	
Partial	15 (20.5)	21 (31.3)	74 (37.6)	
None	57 (78.1)	41 (61.2)	118 (59.9)	
Left AVV repair (no, %)	0.131			
Annuloplasty	11 (15.1)	9 (13.4)	20 (10.2)	
Commissuroplasty	2 (2.7)	2 (3.0)	16 (8.1)	
Additional procedure (no, %)				
Synthetic chordae				
Tricuspid valve	0 (0.0)	3 (4.5)	1 (0.5)	
Mitral valve	1 (1.4)	0 (0.0)	0 (0.0)	
Shortening chordae	0 (0.0)	0 (0.0)	1 (0.5)	
Papillary muscle splitting MV	0 (0.0)	3 (4.5)	1 (0.5)	
LSVC directed to LA	41 (56.2)	43 (64.2)	104 (52.8)	
Debanding	0 (0.0)	14 (20.8)	113 (57.3)	
Debanding plus pericardial patch	0 (0.0)	1 (1.5)	13 (6.6)	
PDA closure	5 (6.8)	11 (16.4)	25 (12.7)	
RVOTO relief/TAP/debanding	1 (1.4)	0 (0.0)	16 (8.1)	
PVR	0 (0.0)	0 (0.0)	1 (0.5)	
AVR	0 (0.0)	0 (0.0)	1 (0.5)	
Cor-triatriatum	1 (1.4)	0 (0.0)	0 (0.0)	
Muscular VSD closure	0 (0.0)	0 (0.0)	2 (1.0)	
LSVC/Shunt ligation	0 (0.0)	0 (0.0)	3 (1.5)	
Pulmonary valve commissurotomy	2 (1.0)	0 (0.0)	0 (0.0)	
Subvalvular AS	1 (1.4)	0 (0.0)	0 (0.0)	

Open PFO	2 (2.7)	0 (0.0)	3 (1.5)
Fenestrated ASD	1 (1.4)	1 (1.5)	5 (2.5)

CPB=Cardiopulmonary bypass; ACC=Aortic cross clamp; LAVVC=Left atrioventricular valve cleft; RAVVC=Right atrioventricular valve cleft; AVV=Atrioventricular valve; LSVC=Left superior vena cava; MV=Mitral valve; LA=Left atrium; PDA=Patent ductus arteriosus; RVOTO=Right ventricular outflow tract obstraction; TAP=Transannular patch; PVR=Pulmonary valve replacement; AVR=Aortic valve replacement; VSD=Ventricular septal defect; AS=Aortic stenosis; PFO=Patent foramen ovale; ASD=Atrial septal defect.

Hospital outcomes and patient follow up

The *length of stay* on the ventilator for postoperative patients was 26.8 ± 2.8 h (1 to 580 h), the average length of hospitalization in the intensive care unit was 4.4 ± 0.2 days (1 to 40 d). The hospitalization time was 26.8 ± 0.5 d (from 1 to 62 d). CHB was detected in 6 patients (1.7%) from all patients undergoing surgery, and epicardial permanent pacemaker was embedded in 5 cases during admission, while 70 patients (20.7%) had *complications after surgery.* Four patients (1.1%) received Extracorporeal Membrane Oxygenation (*ECMO*), while one patient (0.3%) received LVAD. Furthermore, RVAD was used in one patient (0.3%) and all patients using the device in the hospital causes of heart failure. However, all patients who used the device died at a hospital due to heart failure. Early mortality was observed in 36 patients) (10.7%), of which 31 (86.1%) occurred in patients with CAVSD. The cause of death was determined to be heart failure in 5 patients (13.8%), while 10 patients (27.7%) died due to heart failure with Multiple Organ Failure (MOF) etc. (Table 4). The mean age of patients with early mortality was 33.4 ± 5.2 month, which was lower than that of other patients (67 ± 5.8 month). In patients who died early, 6 cases of them (4.9%) had moderate to severe or sever atrioventricular septal defect, which it had a significant effect on early mortality as compared to other patients. Seventy patients underwent surgery again due to blockage of the heart and atrioventricular septal defect at the same hospitalization. The mean pulmonary arterial pressure in patients with early-onset mortality was 37.2 ± 6.6 mm Hg, which its effect on early-onset mortality was not significant as compared to patients who did not have PA Band. In patients with CAVSD, early mortality was seen in 9 cases (29%) with the one stage operation, while it was occurred in 22 cases (71%) with two-stage operation. However, there was no significant difference in the effect of type of operation in early mortality (Table 5).

Table 4. *Postoperative clinical data.*

Variables	PAVSD (73; 21.7%)	TAVSD (67; 19.9%)	CAVSD (197; 58.5%)	P value
G. LAVVR (no, %)	0.275			
None	3 (4.1)	2 (3.1)	11 (5.8)	
Trivial	1 (1.4)	0 (0.0)	6 (3.2)	
Mild	29 (39.7)	26 (40.0)	82 (43.2)	
Mild to moderate	10 (13.7)	10 (15.4)	29 (15.3)	
Moderate	22 (30.1)	21 (32.3)	53 (27.9)	
Moderate to severe	8 (11.0)	3 (4.6)	6 (3.2)	
Severe	0 (0.0)	3 (4.6)	3 (1.6)	
G. RAVVR (no, %)	0.456			
None	6 (8.2)	2 (3.1)	14 (7.4)	
Trivial	9 (12.3)	7 (10.8)	13 (6.8)	
Mild	39 (53.4)	40 (61.5)	94 (49.5)	
Mild to moderate	6 (8.2)	8 (12.3)	20 (10.5)	
Moderate	9 (12.3)	7 (10.8)	38 (20.0)	
Moderate to severe	3 (4.1)	0 (0.0)	7 (3.7)	
Severe	1 (1.4)	1 (1.5)	4 (2.1)	0
Residual VSD (no, %)	5 (6.8)	10 (15.2)	58 (30.5)	0.017
Residual ASD (no, %)	7 (9.6)	0 (0.0)	7 (3.7)	0.571

EF (mean ± SE)	55.8 ± 1.1	56.4 ± 1.3	57.2 ± 0.7	0.616
EF<55% (mean ± SE)	41.7 ± 2.4	40.3 ± 3.0	43.3 ± 1.5	
Postoperative echo finding	0.003			
Severe LVOTO	1 (1.4)	0 (0.0)	0 (0.0)	
Pericardial effusion	0 (0.0)	1 (1.5)	0 (0.0)	
Pulmonary stenosis				
Mild	0 (0.0)	0 (0.0)	11 (5.8)	
Moderate	0 (0.0)	0 (0.0)	9 (4.7)	
Pulmonary insufficiency				
Mild	1 (1.4)	0 (0.0)	3 (1.6)	
Moderate	0 (0.0)	1 (1.5)	4 (2.1)	
Severe	0 (0.0)	0 (0.0)	3 (1.6)	
Mild aortic stenosis	1 (1.4)	0 (0.0)	0 (0.0)	
Moderate aortic insufficiency	2 (2.7)	0 (0.0)	0 (0.0)	
Bicuspid aortic valve and mild PS	1 (1.4)	0 (0.0)	0 (0.0)	

G=Grade; LAVVR=Left atrioventricular valve regurgitation; RAVVR=Right atrioventricular valve regurgitation; EF=Ejection fraction; VSD=Ventricular septal defect; ASD=Atrial septal defect; LVOTO=Left ventricular outflow tract obstruction; PS=Pulmonary stenosis; SE=Standard error.

Table 5. *In hospital results.*

Variables	PAVSD (73; 21.7%)	TAVSD (67; 19.9%)	CAVSD (197; 58.5%)	P value
Mean time on ventilator (h)	17.5 ± 2.0	30.8 ± 9.0	29.0 ± 3.7	0.207
Mean ICU stay (d)	3.9 ± 0.4	4.4 ± 0.6	4.9 ± 0.4	0.341
Mean hospital stay (d)	17.7 ± 0.7	20.3 ± 1.2	21.7 ± 0.7	0.008
Early mortality (No, %)	1 (1.4)	4 (6.0)	31 (15.7)	0.001
Cause of death	0.953			
Heart failure	0 (0.0)	0 (0.0)	5 (16.1)	
MOF	0 (0.0)	1 (25.0)	2 (6.5)	
Cardiac arrest	1 (100)	1 (25.0)	6 (19.4)	
HF/MOF	0 (0.0)	2 (50.0)	8 (25.8)	
HF/Cardiac arrest	0 (0.0)	0 (0.0)	4 (12.9)	
MOF/Cardiac arrest	0 (0.0)	0 (0.0)	2 (6.5)	
VAP	0 (0.0)	0 (0.0)	2 (6.5)	
VAP/HF	0 (0.0)	0 (0.0)	1 (3.2)	
Sepsis/DIC	0 (0.0)	0 (0.0)	1 (3.2)	
Complications (N=70, %)				
Bleeding	0 (0.0)	0 (0.0)	4 (7.5)	
Cardiac complication				
CHB	1 (10.0)	0 (0.0)	5 (9.4)	
RBBB	0 (0.0)	1 (14.3)	1 (1.9)	

SSS	1 (10.0)	0 (0.0)	0 (0.0)	
Subvalvar PS	0 (0.0)	0 (0.0)	1 (1.9)	
Pericardial effusion	1 (10.0)	0 (0.0)	0 (0.0)	
Heart failure	1 (10.0)	0 (0.0)	10 (18.9)	
Pulmonary complications				
Prolonged intubation (>48 h)	2 (20.0)	1 (14.3)	10 (18.6)	
Pleural effusion	2 (20.0)	3 (42.9)	6 (11.3)	
Diaphragmatic paralysis	0 (0.0)	0 (0.0)	1 (1.9)	
Pneumonia and CHF	1 (10.0)	0 (0.0)	0 (0.0)	
Sepsis/DIC	0 (0.0)	0 (0.0)	1 (1.9)	
Sternal dehiscence	0 (0.0)	0 (0.0)	3 (5.7)	
Fever	0 (0.0)	0 (0.0)	1 (1.9)	
Wound infection	1 (10.0)	0 (0.0)	5 (9.4)	
Stroke/deep coma	0 (0.0)	1 (14.3)	4 (7.5)	
Abdominal complications				
Obstruction (internal hernia)	0 (0.0)	0 (0.0)	1 (1.9)	
GIB and respiratory distress	0 (0.0)	1 (14.3)	0 (0.0)	
Early reoperation (No, %)	1 (1.4)	1 (1.5)	5 (2.5)	0.78
LAVV repair/replacement	0 (0.0)	1 (100)	1 (20)	
Pace maker implantation	1 (100)	0 (0.0)	4 (80)	
Cardiac support devices (No, %)				
ECMO	0 (0.0)	1 (100)	3 (60.0)	
LVAD/RVAD	0 (0.0)	0 (0.0)	2 (40.0)	

ICU=Intensive care unit; MOF=Multi-organ failure; HF=Heart failure; VAP=Ventilator associated pneumonia; DIC=Disseminated intravascular coagulation; CHB=Complete heart block; RBBB=Right bandle branch block; SSS=Sick sinus syndrome; PS=Pulmonary stenosis; GIB=Gastrointestinal bleeding; LAVV=Left atrioventricular valve; ECMO=Extracorporeal membrane oxygenation; LVAD=Left ventricular assist device; RVAD=Right ventricular assist device.

Out of 337 patients, 36 patients were not followed up because of their early death, and 94 patients (31.2%) were not followed up due to lack of referral after the discharge. The duration of patients follow up varied from 1 to 120 months (average: 24.8 ± 1.8 months) and 35 (11.6%) were followed up incompletely. The failure of the left atrioventricular septal defect was more than moderate level *follow-up of* 33 patients (10.9%). The mitral valve was restored in two patients, which the mitral valve were replaced with a mechanical valve (St-Jude) and Carbomedix 27, the second time interval of the first operation was 9.9 ± 7.9 months, the interval between the first operation and next operation was 29.7± 10.9 months on average. Two patients underwent LVOTO surgery after 29 months and 33 months, the average interval between this operation and the first operation was determined as 29.2 ± 5.9 months (range: 10-60 months). Late deaths occurred in 3 cases (0.9%), but 129 patients (42.8%) were not fully or relatively followed up. Therefore, the accuracy or inaccuracy of it could not be exactly judged (Table 6).

Table 6. *Follow-up outcomes.*

Variables	PAVSD (73; 21.7%)	TAVSD (67; 19.9%)	CAVSD (197; 58.5%)	P value
Mean follow-up period (months)	21.5 ± 3.0	23.3 ± 3.1	27.3 ± 2.8	0.374
Lost to follow-up (no, %)	8 (10.9)	18 (26.8)	68 (34.5)	
More than moderate LAVVR (no, %)	11 (15.1)	6 (9.0)	16 (8.1)	0.226

Late reoperation (no=7, 2.1%)	5 (6.8)	0 (0.0)	2 (1.0)	0.005
Residual VSD	0 (0.0)	0 (0.0)	1 (50.0)	
LAVVR	2 (40.0)	0 (0.0)	1 (50.0)	
Both LAVVR and RAVVR	1 (20.0)	0 (0.0)	0 (0.0)	
LVOTO relief	2 (40.0)	0 (0.0)	0 (0.0)	
Late mortality (No=3/301)	0	1 (0.03)	2 (0.6)	

LAVVR=Left atrioventricular valve regurgitation; RAVVR=Right atrioventricular valve regurgitation; VSD=Ventricular septal defect; LVOTO=Left ventricular outflow tract obstruction.

Discussion

The mortality rate after AVSD varies considerably and is often dependent on the experience of the therapeutic system in which surgery is performed and consequently varies between 3-21.7% [15,16,19].

In the present study, early death was observed in 36 patients (10.7%), of which 31 patients were categorized as CAVSD (15.7%), followed by TAVSD (4 patients; 6%) and PAVSD (1 patient; 1.4%), which is consistent with the above studies [15]. There was a significant difference in the mean age of patients with premature death (P=0.006). Unlike other studies, the premature mortality rate has increased with age. Out of 36 premature deaths in 26 patients, an echo was performed after surgery, of which 6 (21.4%) had LAVVR (more than moderate level). Consequently, its effect on early mortality was statistically significant compared to other live patients (P=0.008). In recent decades, an understanding of the anatomical form of the disease and progress in surgical techniques has gradually reduced postoperative mortality [15-17,19]. Moreover, progress in preoperative diagnosis and postoperative care has led to improvement of operation outcome. Although the age of children undergoing CAVSD surgery has generally declined significantly in recent decades, the age for restoration is still controversial, and some believe that patients should be treated until one year old [24,30]. One of the major goals of the initial restoration is the prevention of irreversible pulmonary vascular disease, which can also occur early in 6 *month-old* [31]. In this study, the mean age of CAVSD patients who operated as one stage was 33.4 ± 6 months. In 24 cases (32%), an operation was performed at one year of age which the difference in early mortality was not significant compared with those older than one year (P=0.309).

The complete initial repair in AVSD may have the benefits, because degenerative changes or dilation of the annulus of *common atrioventricular valve* or both may be increased with the patient's age. Some studies have shown that annulus dilatation is effective in causing LAVV defect and an increased risk of re-operation in older patients [16,19]. However, in this study, the incidence of moderate atrioventricular septal defect in patients less than 1 y old was determined as 6 patient (12.8%), where the operation was reported in 27 patients (9.3%), (P>0.05). In one study, the early mortality rates for PAVSD and TAVSD were reported to be 3.1% and 2.9% respectively [32]. In the present study, mortality and morbidity were found to be consistent with mentioned study. Patients with AVSD and Down syndrome historically had a better prognosis than the patients without Down syndrome, and had less reoperation [33-36]. Most recent reports indicate that Down syndrome has no effect on survival after surgery [37].

In this study, early survival was reported in 8 patients (22.2%) in Down syndrome patients, while 28 patients without syndrome Don (77.8%) was registered that, there was no significant difference (P=0.061). Despite improvement in patient mortality, surgery has had a limited effect on the LAVVR after surgery in the last few years.*Despite complete cleft closure,* the prevalence of LVAVR is almost constant [13,34,35]. In North America and Europe, 9%-17% of patients with AVSD have significant left atrioventricular septal defect despite surgical repair [14,26].

In this study, 33 patients (9.8%) were found to be more than average a moderate atrioventricular septal defect after the operation that could increase the mortality rate and reoperation after surgery. Among patients with premature death, 6 cases (21.4%) showed LVVR more than moderate level. In addition, 14 patients (4.1%) underwent early and late operations for valvular heart disease and residual defects. The prevalence of Left Ventricular Outflow Tract Obstruction (LVOTO) after the AVSD repairvaries in-between 2.7-5.6 [20]. In this study, the prevalence of reoperation for LVOTO was 0.6% (2 patients). Five patients received (1.4%) postoperative epicardial pacemakers due to CHB. Over the past few decades, the simultaneous occurrence of CAVSD with the TOF has been relatively low, and observed in 5-10% of children, while patients with TOF have shown 1.7% CAVSD [38,39]. In these patients, after surgery, Pulmonary Valve Failure (PVR) can increase Pulmonary Arterial Pressure (PAP) and exacerbate the Right Atrioventricular Valve (*RAVV*). On the other hand, the severe Left Atrioventricular Valve (LAVV) will increase the Pulmonary Arterial Pressure (PAP). This will also exacerbate the pulmonary valve defect (PVR). Due to this hemodynamic disorder, the failure of the *tricuspid* andpulmonary valves can be increased and the risk of heart failure will increase significantly [38,40]. In this study, both CAVSD and TOF were seen in 13 (7.6%) patients, despite the hemodynamic impairment, there was no early mortality in these patients. Complete follow-up in 172 patients (57.2%) was feasible and the incidence of atrioventricular septal defect in follow-up of

33 patients (10.9%) was moderate, of which 4 cases needed late reoperation [4]. Other cases were followed up with drug therapy and in general, late mortality rate was calculated as 0.9% (3 cases). Despite the referrals of patients with high age for surgery, the results of this study were acceptable compared with the results of other countries. However, the age of complete repair in this study was high with average 63.5 ± 5.2 months (Ranged from 2 months to 60 y). 144 patients (42.7%) have been repaired using two stages operation, which can explain the high rate of LVVR after surgery. The most important cause of early mortality can be remarkable LVVR persistence and cardiac failure. Nowadays, it is recommended that patients with atrioventricular septal defect undergo an early and one-time operation. However, in this study, 42.7% of patients received a two-stage operation at an advanced age. The next limitation was follow-up of patients after surgery, in which 57.2% of patients had complete follow-up. Therefore, follow-up, prognosis and late-mortality were unacceptable.

Conclusion

To address these issues, patients should be referred to centres that have more experience in performing congenital complicated operations for implementing an early surgery in one-step. The next issue is patient follow up which is better, like advanced countries, to train nurses (practitioners) for following up patients after discharge. This will make it easy for patients and their relatives to access them where they can deal with their problems at the right time. Consequently, they can be informed of their possible deaths and their details can be recorded in the hospital file. In the case of patients who may die due to possible post-operative complications in distant areas, communication between the hospital and the registry office should be facilitated to provide them the necessary information in the absence of referrals.

Finally, it is suggested that the electronic records be returned to patients in the near future and if funds are available like some advanced countries. Therefore, doctors can access it throughout the country and record possible complication. These facilities can provide the necessary conditions for follow up, recording patient information, analyzing and improvement of treatment strategies.

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None.

Conflicts of Interest

None.

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