Early and mid-term results of surgical treatment of atrioventricular septal defect; analysis of risk factors for adverse outcomes: left atrioventricular valve regurgitation

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Received: 11 September 2023
Revised: 16 September 2023
Accepted: 17 September 2023
Published: 7 December 2023

The Egyptian Journal of Surgery 2023, 42:1125–1130

Background
Advances in the perioperative management and optimization of the timing of primary repair have gradually improved the early clinical outcome of surgical correction of atrioventricular septal defects (AVSDs). However, the late outcome still remains compromised by the substantial risk of reoperation, of which mainly the development of left ventricular outflow tract obstruction (LVOTO) and left atrioventricular valve (LAVV) dysfunction challenge the long-term survival and morbidity of these children.
In particular, LAVV regurgitation (LAVVR) is responsible for a reoperation rate between 5 and 19%, both for complete AVSD (cAVSD) and partial AVSD (PAVSD) children.
Results of surgical correction of AVSDs) have improved over the last decades; however, the need for reoperation after primary AVSD repair remains a major concern. We conducted this study to analyze risk factors leading to LAVVR in the early and mid-term follow-up postsurgical repair of AVSD.
Patients and methods
All patients who underwent biventricular repair for AVSD at our institute, in the time between Jan 2015 and Jan 2021, meeting the inclusion criteria and none of the exclusion criteria were enrolled in the study. Preoperative, operative and postoperative data were collected from the patients' medical file. Last follow-up Echoes were reviewed.
Results
Patients who had preoperative and early postoperative more than moderate LAVVR are more likely to develop LAVVR in the late follow-up and also have a more risk to need a reoperation in their midterm follow-up.
Conclusions
Patients who had preoperative and early postoperative more than mod LAVVR are more likely to develop LAVVR in the late follow-up and also more likely to need reoperation.

Keywords:
atrioventricular septal defect, left atrioventricular valve regurgitation, risk factors

Egyptian J Surgery 42:1125–1130

1110-1121

Introduction

Atrioventricular septal defects (AVSDs) consist of a spectrum of cardiac abnormalities resulting from a defect in endocardial cushions development leading to a defect in the atrial and ventricular septum to a varying degrees together with abnormalities in the AV valve [1–4].

AVSD affects 4–5.3/10 000 live birth, accounting for 7% of all congenital heart disease [2,5,6].

Each year, ~1 in 2000 babies are born in the United States with AVSD [2,3].

There is a strong association between AVSD and trisomy 21 and ~50% of cases occur in patients with Down's syndrome [1] while almost 50% of Down syndrome babies show AVSD (commonly complete and rarely partial AVSD) [6,7].

Elective repair of PAVSD is typically performed in the preschool years, usually between 2 and 4 years of age, to be repaired when diagnosed and before an operation would interfere with school [1,8,9]. unless signs of heart failure developed earlier [4,10].

On the other hand recently due to marked advancement in the surgical techniques, cardiopulmonary bypass strategies and proper postoperative care most centers have adopted early repair of CAVSD between 3 and 6 month of age [11–17].

Although the whole spectrum of AVSD are surgically repaired with excellent early and late survival, still the LAVV competency is a matter of investigation, representing the most common cause for reoperation ranging from 5 to 20% [8,11,18,19].

CAVSD repair is probably a more demanding procedure than a standard arterial switch and that the surgeon is more important than the technique itself, confirmed by the observation that different surgical techniques are not associated with different outcomes [20]. Also PAVSD may not be as benign as previously thought as it has a risk of reoperation approaching 25% at 25 years [8].

Aim

To analyze risk factors leading to LAVVR in the early and mid-term follow-up post AVSD surgical repair.

Patients and methods

We investigated a total of 111 patients who underwent biventricular repair for AVSD from Jan 2015 to Jan 2021, excluding any unbalanced patients directed to univentricular repair, patients with associated construal abnormalities and syndromes other than Down syndrome. On the other hand patients who had pulmonary artery banding prior to the definitive repair, and patients who underwent simultaneous closure of ASD, PFO or PDA were included.

We used the hospital medical records to collect our data needed in our statistical analysis such as patients' demographic characteristics, preoperative findings, operative details and postoperative charts.

All 54 (48.6%) patient were diagnosed as cAVSD, 57 (51.4%) patient were diagnosed as PAVSD preoperatively. Out of 111, 61 (55.0%) were females and 50 (45.0%) were males, 44 (39.6%) were Down, with a median BSA 0.48 with interquartile range (IQR) (0.38–0.86).

The basic demographic data for our patient population is demonstrated in table below (Table 1).

The surgical technique

Surgery was conducted through a median sternotomy, moderate hypothermia while on cardiopulmonary bypass on an arrested heart in diastole by antegrade cardioplegia.

All PAVSD patients 57 (51.4%) underwent Pericardial patch closure for primum ASD, with only two patients suffered from difficult weaning from bypass till a fenestration was made in the ASD patch.

On the other hand 54 (48.6%) patients who were diagnosed as complete defect preoperatively were repaired by either a DPT (Double Patch Technique) or an MSP (Modified Single Patch) technique according to the surgeon preference relying on the size and extension of the VSD judged by the surgeon's experience intraoperatively.

Among those CAVSD patients 25 (22.5%) were repaired using a 2 patch technique closing the VSD by a Gortex and using a native pericardial patch to close the ASD.

We applied the MSP technique in 29 (26.1%) patients closing the VSD by approximating the crest of IVS from below to the anticipated line dividing the right

Table 1 Basic demographic data

	Total no.=111
Age (months)	
Median (IQR)	23.8 (13–73)
Range	4–468
Sex	
Male	50 (45.0%)
Female	61 (55.0%)
Down Status	
No	67 (60.4%)
Yes	44 (39.6%)
Weight	
Median (IQR)	10 (7–23)
Range	4–93
Height	
Mean±SD	94.10±30.96
Range	50–176
BSA (m^2)	
Median (IQR)	0.48 (0.38–0.86)
Range	0.24–2.13
PAB	
No	105 (94.6%)
Yes	6 (5.4%)
Age of PAB	
Mean±SD	5.48±1.54
Range	3.1–7.67
Complete canal	
No	57 (51.4%)
Yes	54 (48.6%)

BSA, Body surface area; IQR, Interquartile range; LAVV, Left atrioventricular valve regurgitation; PAB, Pulmonary artery banding; SD, Standard deviation

(Rt) and left (Lt) AVV by interrupted proline sutures, the same sutures are then used to fixate the ASD patch from above the common AVV.

ZOA (Zone Of Apposition) between superior and inferior bridging leaflets –some authors call it LAVV cleft- was routinely closed in all patients by interrupted proline sutures to the degree that effectively treats regurgitation but without causing any stenosis. This was feasible for all patients except for one patient whose valve deemed to be stenotic, so the LAVV was kept as a trifoliate valve with a satisfactory postoperative result.

Additional LAVV posterior annuloplasty were performed for only 24 (21.6%) patients of the total population, who had a dilated annulus preoperatively and intraoperatively according to the surgeon preference also.

The competency of AVVs are then tested by saline injection in the LV prior to closure of RA, desiring of left side of the heart through the aortic root, then weaning from bypass, hemostasis and closure in layers.

The gathering of follow-up data was closed in Aug 2023. With a minimum follow-up period of 2 years reaching to 7 years.

Statistical analysis

Data were collected, revised, coded and entered to the Statistical Package for Social Science (IBM SPSS) version 27. The quantitative data were presented as mean, standard deviations and ranges when parametric and median, IQR when data found nonparametric. Also qualitative variables were presented as number and percentages. The comparison between groups regarding qualitative data was done by using χ^2 test and/or Fisher exact test when the expected count in any cell found less than 5. The comparison between two independent groups with quantitative data and nonparametric distribution were done by using Mann–Whitney test. The confidence interval was set to 95% and the margin of error accepted was set to 5%. So, the p-value was considered significant at level of *P*-value less than 0.05. Survival or event- free survival as freedom from reoperation is analyzed by the Kaplan–Meier product-limit estimation method.

Results

Late post op LAVVR

In our study, late postoperative LAVVR was more than moderate in 28 (26.7%), there was no statistically significant difference in the incidence of late postoperative LAVVR between either different demographic groups or different surgical techniques used (Table 2).

We found that patients with preoperative more than moderate LAVVR were more likely to develop late postoperative LAVVR than others, with the difference reaching statistical significance. *P* 0.002.

Also those who had residual more than moderate LAVVR in their predischarge echo were more likely to develop more than moderate LAVVR in their last follow-up echo, reaching statistically significance. *P* 0.000.

And as expected they needed prolonged ventilation, reaching statistically high significance. *P* 0.014

Interestingly adding LAVV posterior annuloplasty was not protective from developing LAVV incompetence in the late follow-up. *P* value 0.639 (Table 3).

Reoperation

A total of 104 patients completed a minimum follow-up period of 2 years (ranging 2–7 yrs.) with only 10 (9.6%) patients requiring a reoperation.

Table 2 Showing the suspected risk factors for late left atrioventricular valve regurgitation

	LAVVR late postoperative		
Risk factors	No No.=76	Yes No.=28	*P*-value
Age (months)			
Median (IQR)	24.52 (14–81.45)	24.9 (13.85–47)	0.700
Range	4–456	5–353.23	
Down status			
No	47 (61.8%)	17 (60.7%)	0.916
Yes	29 (38.2%)	11 (39.3%)	
BSA (m2)			
Median (IQR)	0.48 (0.39–0.87)	0.51 (0.41–0.73)	0.951
Range	0.24–2.13	0.3–1.66	
Complete canal			
No	43 (56.6%)	12 (42.9%)	0.214
Yes	33 (43.4%)	16 (57.1%)	
Preoperative > moderate LAVVR			
No	53 (69.7%)	10 (35.7%)	0.002
Yes	23 (30.3%)	18 (64.3%)	
Early postoperative > moderate LAVVR			
No	84 (100.0%)	10 (50.0%)	0.000
Yes	0	10 (50.0%)	
Prolonged Ventilation > 48 h			
No	71 (94.7%)	22 (78.6%)	0.014
Yes	4 (5.3%)	6 (21.4%)	

BSA, Body surface area; LAVV, Left atrioventricular valve regurgitation.

Table 3 Effect of posterior annuloplasty on left atrioventricular valve regurgitation at last follow-up

	LAVV annuloplasty		
	No No.=82	Yes No.=22	*P*-value
LAVVR at follow-up			
No	67 (81.7%)	17 (77.3%)	0.639
Yes	15 (18.3%)	5 (22.7%)	

LAVV, Left atrioventricular valve regurgitation.

Regarding the patients requiring reoperation 8 (80%) patients were re-operated on due to severe LAVVR, 2 of them had associated residual septal defects. Another one (10%) patient was re-operated by excising a subaortic membrane that was causing LVOTO. The remaining one (10%) patient required reoperation to closure his residual septal defects that was causing severe shunting at the atrial and ventricular levels.

In our investigation patients with preoperative more than mod LAVVR were more likely to require reoperation, seven (70%) patients of the 10 patients requiring reoperation had more than moderate LAVVR preoperatively. The difference was statistically significant, *P* 0.037. Also those with their early postoperative echo showing more than moderate LAVVR carried a higher risk for reoperation *P* 0.021 Table 4.

We were obliged to replace the LAVV by mechanical prosthesis in two (25%) patients from those who were re-operated for LVVR, One of them the valve was severely dysplastic and the other was double orifice with small left lateral leaflet.

Freedom from reoperation was 97.7, 89, and 83.9% at 3, 5, and 7 yrs. of follow-up, respectively.

Univariate analysis of preoperative, operative and postoperative revealed that only the more than moderate LAVVR in the preoperative and early postoperative echo as the only significant determinant for the need for reoperation.

Additional reoperations

One (10%) for relieving LVOTO was a type B CAVSD repaired by two patch technique at age of 1 y then after 6.5 y developed a subaortic membrane that was surgical removed.

One (10%) for residual septal defects was a case of CAVSD operated at age of 2.5 y. using an MSP technique then developed dehiscence of septal patch 2.5 y. PO. So was re-operated closing ASD by dacrone and directly closing the VSD. This patient may need a second reoperation sooner for still residual septal defects and progressing LVOTO, so needs a close follow-up.

Table 4 The suspected Risk Factors for Reoperation

	Reoperation and cause		
	No No.=94	Yes No.=10	*P*-value
Age (months)			
Median (IQR)	24.13 (13.7–70)	38.28 (15.77–124.37)	0.297
Range	4–456	12.43–353.23	
Down Status			
No	55 (58.5%)	9 (90.0%)	0.052
Yes	39 (41.5%)	1 (10.0%)	
BSA (m2)			
Median (IQR)	0.48 (0.38–0.8)	0.66 (0.42–0.97)	0.209
Range	0.24–2.13	0.3–1.7	
PAB			
No	89 (94.7%)	9 (90.0%)	0.546
Yes	5 (5.3%)	1 (10.0%)	
Complete canal			
No	50 (53.2%)	5 (50.0%)	0.848
Yes	44 (46.8%)	5 (50.0%)	
Preoperative > mod LAVVR			
No	60 (63.8%)	3 (30.0%)	0.037
Yes	34 (36.2%)	7 (70.0%)	
Postoperative > mod LAVVR			
No	87 (92.6%)	7 (70.0%)	0.021
Yes	7 (7.4%)	3 (30.0%)	

BSA, Body surface area; LAVV, Left atrioventricular valve regurgitation; PAB, Pulmonary artery banding.

Discussion

Although the more understood anatomy and physiology of LAVV in AVSD, more than moderate LAVV incompetence postoperatively remains the main predictor for reoperation [11]. Improvement of surgical techniques and bypass strategies had enabled the surgeons to repair cAVSD at a younger age deferring from staged palliation by PAB [21].

Although current practice is to correct PAVSD in the preschool age or earlier if heart failure develops [10].

The age for elective CAVSD repair has been lowered, from 1 year a few decades ago to 3 to 6 month, intending to minimize the risk of premature death or pulmonary vascular obstructive disease [16,21,22].

In our study CAVSD Patients were operated markedly younger than PAVSD, with the youngest patient was operated at age of 4 month, younger than that we used to perform a first stage PAB to protect the pulmonary circulation that can cause permanent pulmonary hypertension [23]. This staged repair had a comparable results with the primary repair with no statistically significant difference regarding mortality or reoperation due to LAVVR.

In recent years, many units have adopted routine cleft closure in the hope of reducing the risk of progressive LAVVR and reoperation [24].

Ginde and colleagues observed that LAVVR was the most common indication for reoperation in 14 patients (7.1%)of their study [25] also other studies by Reynen and colleagues and Hoohenkerk and colleagues have found the LAVVR as the major cause for reoperation in 7 to 20% of patients freedom from reoperation at 10, 15, and 20 years was 82.7, 81.1, and 77%, respectively [10,12,13,19,26].

We demonstrated a similar result with the LAVVR the main cause for reoperation in 8 (80%) of re-operated patients with incidence of 7.6% of AVSD repaired patients. Freedom from reoperation was 97.7, 89, and 83.9% at 3, 5, and 7 yrs. of follow-up, respectively.

In all cases, LAVV repair is preferred to LAVV replacement avoiding the need for mechanical valve with its hazards in pediatric population [10,11].

Buratto and colleagues reported that most cases of LAVV regurgitation were amenable to repair (77%, 24/31), rather than replacement (23%, 7/31) [10].

Similarly we could repair LAVV in six (75%) of patients and only had to replace LAVV by mechanical valve in only two (25%) patients whose valves were severely dysplastic.

The risk factors for left AV valve reoperation included low weight at initial surgery, associated cardiac malformations, persistent left superior vena cava, tetralogy of Fallot, and unbalanced valve/ventricles [16].

Preoperative severe LAVVR and moderate or greater postoperative LAVVR were identified as risk factors for overall cardiac reoperation after cAVSD repair. While a diagnosis of trisomy 21 (P<0.03, HR 0.6, 95% CI: 0.33–0.96), and more recent era of surgery were protective [24].

Fong and colleagues found that patients without Down syndrome and moderate L-AVV regurgitation on postoperative echocardiogram were found to be independent risk factors for reoperation in the Australian series [19].

Others had found associated cardiovascular anomalies, LAVV dysplasia, and nonclosure of the cleft to be Independent risk factors for LAVVR reoperation [12].

But in our study, we could not find neither low weight nor the down status to be risk factor for reoperation but the preoperative more than moderate LAVVR, also the early postoperative more than moderate LAVVR the main predictors for the risk of reoperation.

There is a significant rate of moderate-to- severe postoperative LAVV regurgitation (16% at discharge), probably very disappointing when taking into consideration the correlation between postoperative LAVV regurgitation and the need for reoperations [20].

Although Azzab and colleagues detected the adding post annuloplasty to cleft closure to be a protective factor for LAVVR at 6 month [27]. We found it is not significantly affecting the LAVVR, may be because small number of patients had post annuloplasty in our cohort who were followed up for a longer time 5 y (2.5–7.5 y).

Our current practice is to entirely close the ZOA between the superior and inferior bridging leaflets of the LAVV except for those patients whose valves are prone to stenosis and those with small left mural leaflet. We may add postannuloplasty if the annulus is dilated.

Bové and colleagues were able to repair up to 90% of their regurgitate LAVVR, so as done by other reports ranging from 70 to 85% Interestingly we have nearly the same result with 75% of patients were successfully repaired.

Conclusion

The routine periodic regular follow-up for all patients who had AVSD repaired is mandatory with spotting the light on the LAVV competency as it was found by our study and most of the recent publication to be the most common cause for reoperation second to it was the LVOTO and residual defects.

Because we found that the preoperative as well as the early postoperative more than moderate LAVVR the significant risk factors for development of LAVVR in the mid-term follow-up and hence need for reoperation. So the routine intraoperative TEE is now mandatory for assessment of the AVV function making sure that AVVR is not more than grade 2, and no significant gradient across it during diastole.

Limitations

This study was limited by its small sample size, retrospective nature conducted in a single-centre and the inability to control for surgeon preference or decision-making in choice of repair technique.

Acknowledgements

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest

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