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ORIGINAL ARTICLE

Single-Stage Surgical Management of Atrioventricular Septal Defects with Coarctation of the Aorta

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Abstract

Surgical options for coarctation of aorta (CoA) with atrioventricular septal defect (AVSD) include single-stage repair vs. staged approach with neonatal CoA repair and delayed AVSD repair. The durability of left atrioventricular valve (LAVV) function after neonatal repair is questioned, and the optimal approach remains controversial. Eighteen CoA-AVSD patients who underwent single-stage repair 2005–2015 by a single surgeon were retrospectively analyzed. Fifteen patients had complete and three had partial AVSD. Birth weight was 3.19 kg (2.17–4.08). Age at surgery was 16 days (6–127). One- and ten-year survival were 80% and 69%. Freedom from reintervention was 60% and 40% at one and ten-year respectively. Reinterventions included relief of left ventricular outflow tract obstruction (LVOTO) ($n=4$), repair of cleft LAVV ($n=3$), and LAVV and aortic valve replacement ($n=1$). Freedom from LAVV reintervention was 85.6% and 66% at 1 and 10 years respectively. There were four deaths: two post-operative and two following hospital discharge. Mortality was due to sepsis in three patients, and heart failure related to LVOTO and LAVV insufficiency in one. At 68-month (0.6–144) follow-up the majority had mild or less LAVV regurgitation, and all had normal LV dimension and systolic function. There was no recurrent arch obstruction. Single-stage surgical repair of CoA-AVSD is feasible and reasonable. Survival and freedom from reintervention in our cohort approximate those outcomes of two-stage repair with durable left AV valve function and no recurrent arch obstruction. These patients are frequently syndromic and demonstrate mortality risk from non-cardiac causes. Consideration of a single-staged approach is warranted for appropriate patients with CoA-AVSD.

Keywords Atrioventricular septal defect · Coarctation of aorta · Single-stage repair · Trisomy 21 · Congenital heart disease

Introduction

Coarctation of aorta (CoA) complicates the surgical management of atrioventricular septal defects (AVSD) in 5 to 20% of patients [1–5]. One strategy is single-stage repair of the AVSD and CoA in early infancy. Another approach is two-staged repair with neonatal arch repair with or without placement of a pulmonary artery band, followed by complete repair of the AVSD at 4–6 months of age. Repair of AVSD in early infancy is challenging due to the fragility of the AV valve tissue and closing the cleft in the neonatal period would lead to left AV valve stenosis. The durability of left atrioventricular valve (AVV) function after early repair, and the associated morbidity and mortality has been questioned [6, 7]. Importantly however, patients with either complete or partial atrioventricular septal defect and aortic arch obstruction frequently have a single, or nearly single left ventricular papillary muscle. This has the effect of significantly

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reducing the surgeon's ability and requirement to close the left-sided cleft for fear of creating left AVV stenosis. Furthermore, there is the question of single vs double patch repair vs modified single patch techniques. The two-stage approach is enticing as it allows time for the AV valve tissue to mature before repair, however, managing the heart failure in an unrepaired AVSD after CoA repair can be challenging. If an MPA band is utilized, there is potential for distortion of the pulmonary arteries and excessive ventricular hypertrophy. The AVSD repair therefore becomes more complicated through a redo sternotomy with these additional challenges. The optimal surgical approach for the management of CoA-AVSD remains controversial. We present our institutional experience and outcomes of the single-stage repair of CoA-AVSD, including echocardiographic data, post-operative outcomes, overall survival, freedom from reintervention, and cardiac findings at the last follow-up.

Patients and Methods

The surgical database from a single center was retrospectively analyzed to identify patients who underwent CoA-AVSD repair between 2005 and 2015 after approval by the Institutional Review Board. A total of 18 patients with CoA-AVSD underwent a single-stage repair during this time period by a single surgeon (JF). Both complete and partial AVSD patients were included in order to report our full experience with arch repair and AV septal defects. Only one surgeon attempted or treated patients with CoA-AVSD during this time period at our institution. The decision to undergo single-stage repair was made by the surgeon assisted by a cardiologist specializing in noninvasive imaging. All patients had aortic arch hypoplasia. Factors that influenced this decision to proceed with single-stage repair included degree of malalignment of the AV junction, number of papillary muscles, degree of AV valve insufficiency, ventricular function, and presence of chromosomal abnormality. A staged biventricular repair approach was not utilized during this time period due to institutional bias. Unbalanced AVSD requiring single ventricle palliation and AVSD with other associated congenital heart diseases such as Tetralogy of Fallot or heterotaxy syndrome were excluded from this study. Patients with moderate to severe or more AV valve insufficiency were not deemed candidates for this single-stage approach. Demographic characteristics, cardiac anatomic details, chromosomal and other anomalies, comorbidities, surgical data, and complications were obtained from the medical record.

Initial (pre-operative), pre-discharge, and most recent follow-up echocardiograms were reviewed. The morphology of AVSD was defined as either complete or partial (incomplete), implying there was no ventricular component to the AVSD. Other associated cardiac findings such as qualitative assessment of left AV valve insufficiency, left AV valve stenosis, left ventricular outflow tract obstruction (LVOTO), number and spacing of the papillary muscles, and presence of bilateral superior vena cava were noted. Left AV valve stenosis was defined by mean gradients, 5 mmHg or more considered mild, 10 mmHg or more considered moderate, and 15 mmHg or more severe. Two methods of determining ventricular balance were utilized. The atrioventricular valve index (AVVI) was obtained in a subcostal left anterior oblique view by drawing a line from the crest of the interventricular septum to the tip of the infundibular septum at end-diastole and creating a ratio of the left and right AV valve area as described by Cohen et al. [8] Unbalanced AVSD was defined as the ratio of left/right AVV area < 0.67. The modified AVVI, proposed by the Congenital Heart Surgeons' Society was obtained using the same view and technique, but created a ratio of the left AV valve area over the total AV valve area [9]. A ratio of ≤ 0.4 was defined as unbalanced right ventricular dominant AVSD, 0.4 to 0.6 is balanced, and ≥ 0.6 is left ventricular dominant AVSD.

Surgical techniques, number and duration of cardiopulmonary bypass runs, and performance of delayed sternal closures were reviewed. Early and late post-operative outcomes such as duration of stay in the intensive care unit (ICU), duration of total hospitalization, duration of mechanical ventilation, chest tube drainage and complications were noted. Reintervention was defined as any procedure performed for repair of atrioventricular valves (AVV), relief of LVOTO, or other thoracic procedures including pacemaker placement required in the post-operative period. Overall survival, reintervention-free survival, and echocardiographic findings at latest cardiology follow-up were analyzed.

Statistical Analysis

Medians and ranges are reported for continuous variables; frequencies and percentages are provided for categorical variables. The Kruskal–Wallis test and Fisher's exact test are used to compare the distribution of variables by group. Kaplan–Meier plots are created for overall survival and reintervention-free survival. All statistical analysis was performed in R v 3.6.1.

Results

Study Population

Our cohort included eighteen patients of which 50% ($n = 9$) were males. The median birth weight was 3.19 kg (2.17–4.08). Three patients were considered low birth weight (< 2.5 kg). The median gestational age at birth was

38 weeks (35–41). A total of 11 patients (61%) had Trisomy 21. Twelve patients received prostaglandin infusion before surgery. One patient required repair of duodenal atresia at day of life 2 followed by complete cardiac repair at day of life 16. One patient was diagnosed with biliary atresia after single-stage cardiac repair and subsequently underwent Kasai procedure.

Pre-operative Echocardiographic Features

The cardiac morphology was consistent with complete AVSD in 15 patients (83%) and partial AVSD in three patients (17%). Of the 15 patients with complete AVSD, RV dominant unbalanced AVSD was present in five as defined by an AVVI < 0.67 and in eight as defined by the modified AVVI ≤ 0.4 (Table 1). Mild left AV valve stenosis was present in one patient with partial AVSD based on the left AV valve z score < − 2.5 (patient 14, Table 1). Left AV valve regurgitation was mild or less in 13, mild to moderate in 4, and moderate in 1 patient. The degree of right AV valve regurgitation was mild or less in 16 patients and mild to moderate in 2 patients. Six patients had parachute left AV valve (single papillary muscle) and 2 had a forme fruste (pseudo/incomplete) parachute left AV valve, defined as two closely spaced papillary muscles or one functional papillary muscle. Associated LVOTO was present in one patient (patient 16, Table 1), bilateral superior vena cava were present in one patient, and left aortic arch with aberrant right subclavian artery was present in one patient. One patient had discrete CoA and the rest had long segment aortic arch hypoplasia. Due to presence of ventricular septal defect, patent ductus arteriosus, and diffuse arch hypoplasia in majority of patients; the velocity across the hypoplastic arches was not always elevated (median: 1.7 m/s, 0.8 to 3.2 m/s). The one patient with isolated discrete coarctation of aorta had an elevated velocity (2.5 m/s) across the isthmus. Left ventricular systolic function was normal preoperatively in all patients.

Cardiac Surgery

The median age at surgery was 16 days (6–127) and the median weight at operation was 3.59 kg (2.4–5 kg). A single surgeon performed all of the operations via median sternotomy. The cardiopulmonary bypass strategy generally utilized moderate hypothermia (24–26 °C) and selective antegrade cerebral perfusion at 40–50 ml/kg/min. Deep hypothermic circulatory arrest was not routinely utilized. Typically, the arch was repaired first, followed by the AVSD. After ligation and resection of the isthmus, an end-to-side anastomosis of the descending to the distal ascending aorta was performed which typically required 12 to 15 min of selective cerebral perfusion. With this technique, there was no recurrent arch obstruction during the follow-up period. Patients were then warmed slowly while performing the canal repair. A two-patch technique or a modified single patch technique was used for the repair of the complete AVSD based on several technical factors including depth of the ventricular septal defect (VSD) component and the density of the atrioventricular valve attachments to the crest of the interventricular septum. If the common atrioventricular valve was only 2 to 3 mm above the crest of the interventricular septum, the modified single patch repair was utilized. If the VSD had more depth to it, the two-patch technique was utilized. Minimal distortion of the atrioventricular valve was the goal. Either a Dacron patch or autologous pericardial patch was used for VSD closure, and autologous pericardial patch was used for atrial septal defect (ASD) closure. A glutaraldehyde fixed autologous pericardial patch in the ventricular component of the AVSD tended to be less rigid than the Dacron patch leading to less perturbation of the valve tissue position and function. This was therefore favored in younger and smaller patients. When closing the atrial component, placing the pericardial patch such that the coronary sinus entered the left atrium, avoiding sutures in the area of the AV node. Again, we felt that this was an important technical maneuver in these neonates and infants who were smaller than typical AV septal defect patients. When testing the left AV valve at time of repair, irrigation with cardioplegia was utilized every other insufflation, keeping cardioplegia in the coronaries rather than washing it out with saline. After removal of the cross-clamp, utilization of a left atrial line was critical to understanding the adequacy of the left ventricle. As postoperative blood products were administered, if the left atrial pressure was greater than 15 mmHg, volume was removed from atrial or arterial line to maintain a lower, more appropriate left atrial pressure during the first several hours post-bypass. Two-patch technique was used in ten patients with complete AVSD (55.6%) and modified single patch technique was performed in five (27.7%) complete AVSD patients. During the primary repair, the left AVV cleft was left open in six patients, and partial closure of the cleft was performed in four patients. The remaining patients ($n = 8$) were able to have complete, or nearly complete cleft closure. The most common concomitant procedures performed were PDA ligation, closure of secundum ASD, or closure of patent foramen ovale. Median cardiopulmonary bypass time was 130.5 min (84–241) and cross-clamp time was 100 min (45–170). None of the patients required a second run of cardiopulmonary bypass for the repair of CoA-AVSD.

Post-operative Course

Early post-operative course and complications are reported in Table 2. Twelve patients (66.7%) underwent delayed sternal closure. The chest was typically left open not so much for

Table 1 Pre-operative and operative characteristics of patients with CoA and AVSD

Pt	Age at cardiac surgery (days)	Wt at surgery (kg)	T21	AVSD type	VSD size	Balanced/ unbalanced AVSD		Papillary muscles	Surgical repair	Cleft closure	Left AVVR			Cardiac Surgical Reintervention (time since first surgery)
						AVVI	mAVVI				Pre-op	Pre-DC	Last f/u	
Survivors														
1	15	4	No	Complete	Large	0.94	0.45	Parachute	1	Partial	Mild	Mild	Mild	LVOT (1 year) LVOT + MV (2.8yrs) LVOT + MV + partial CC (6 years)
2	14	2.9	Yes	Complete	Large	0.63#	0.22^	Pseudo parachute	2	Closed	Trace	Mild	Mild	
3	10	2.4	Yes	Complete	Large	0.81	0.38^	Parachute	2	No	Trace	Mild	Trace	
4	13	3.27	No	Complete	Mod	0.79	0.35^	Normal	1	No	Mild	Mod	Mild-mod	LVOT + CC (10.8 year) LVOT (9 years)
5	8	3.49	Yes	Complete	Large	0.98	0.47	Normal	2	Partial	Trace	Trace	Mild	
6	13	3.95	Yes	Complete	Large	0.89	0.42	Normal	2	No	Mild	Mod	Mod	
7	71	3.8	Yes	Complete	Large	0.93	0.43	Normal	2	Closed	Mild	Mod	Mild-mod	
8	17	3.64	Yes	Complete	Large	0.64#	0.22^	Pseudo parachute	2	Partial	Mild	Mod	Mild	
9	36	2.77	No	Complete	Large	0.69	0.41	Normal	2	Closed	Mild-mod	Mild-mod	Trace	
10	108	4.75	Yes	Complete	Large	0.99	0.50	Normal	2	Closed	Mild-mod	Mild	•	
11	20	2.6	Yes	Complete	Large	0.47#	0.32^	Parachute	1	Partial	Mild	Mild	•	
12	50	5	Yes	Complete	Large	0.96	0.40^	Parachute	1 (Au)	Closed	Mild	Mild	Mild	LVOT (14 mo)
13	8	3.79	No	Partial	–	–	–	Parachute	1	No	Mild	None	Mild	
14	77	3.32	No	Partial	–	–	–	Parachute	1	No	Mild-mod	Mild-mod	Mild-mod	CC (23 days)
Non-survivors														
15	10	3.54	No	Partial	–	–	–	Normal	1	Closed	Mod	Mild	•	LVOT (5.8 years) LVOT + MVR + AVR (6.6 years)
16	6	2.8	Yes	Complete	Large	0.32#	0.25^	Parachute	2	No	Mild	Mild*		
17	127	4	No	Complete	large	0.66#	0.25^	Normal	1 (Au)	Closed	mild-mod	mild-mod*		
18	120	4.13	Yes	Complete	Large	0.96	0.50	Normal	1	Closed	Mild	Mild	Mild*	

Pt patient, *Wt* weight, *kg* kilogram, *T21* trisomy 21, *AVSD* atrioventricular septal defect, *AVVI* atrioventricular valve index, *mAVVi* modified atrioventricular valve index

#AVVI ≤ 0.67 = unbalanced

^ ≤ 0.4 = RV dominant, unbalanced. Surgical repair: 1-single patch technique, 2-two patch technique, Au-Australian technique

*Degree of left atrioventricular valve regurgitation prior to death

•Follow-up external institution, op-operative, DC-discharge, f/u-follow-up, LVOT- relief of LVOT obstruction, MV-mitral valvuloplasty, MVR- mitral valve replacement, AVR-aortic valve replacement CC cleft closure

Table 2 Early postoperative course and complications after single-stage repair of CoA-AVSD

Outcome	N (%)
Mortality	4 (22%)
Operative mortality	2 (11.1%)
Post-Kasai procedure (biliary atresia)	1
Sepsis	1
Post-discharge mortality	2 (11.1%)
RSV bronchiolitis	1
Cardiac	1
ECMO support	0
Delayed sternal closure	12 (66.7%)
Arrhythmia	
Ectopic atrial tachycardia	2 (11.1%)
Atrial flutter	1 (5.5%)
Junctional ectopic tachycardia	1 (5.5%)
Ventricular tachycardia	1 (5.5%)
Reoperation prior to hospital discharge	1
Tracheostomy	1
G tube placement	1
Vocal cord paralysis	4
Chylothorax	4
Infection	6
Respiratory	3
UTI	1
Sepsis	2
Pulmonary Hemorrhage (AP collaterals)	1
Other thoracic procedures	3
Pleurodesis	1
Aortopexy for left main bronchus	1
Embolization of aorto-pulmonary collateral	1
Diagnostic cardiac catheterization prior to hospital discharge	3
Duration of ventilation, median (range) days	4 (2–27)[a]
Duration of ICU stay, median (range) days	16 (4–60)
Duration of hospital stay, median (range) days	28 (12–68)
Duration of chest tube drainage, median (range) days	4 (2–29)

[a]Excluding patients who died/tracheostomy

bleeding, but to allow the right and left ventricles to have the most compliant environment possible while the ventricles recovered their diastolic function post-cross-clamp. Early postoperative control of preload and afterload was critical for the patients with open clefts; both volume overload and/or increased systemic venous resistance would lead to increased left AV valve insufficiency. The average duration for delayed sternal closure was 1.3 ± 0.9 days. The median duration of mechanical ventilation was 4 days (2–27) excluding the patients who died (2) or had tracheostomy (1). One patient had prolonged intubation for 24 days related to Influenza B infection. Median duration of chest tube drainage was

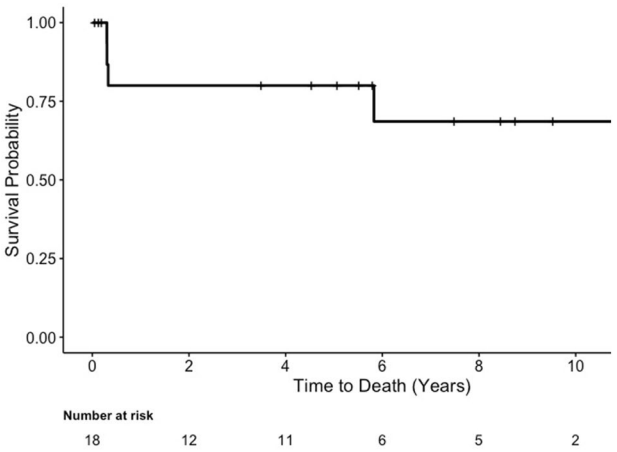

Fig. 1 Kaplan–Meier survival

4 days (2–29). The median length of ICU stay was 16 days (4–60) and post-operative hospital stay was 28 days (12–68). No patients required ECMO support. Other thoracic procedures required in three patients included pleurodesis, aortopexy for left mainstem bronchomalacia, and embolization of aorto-pulmonary collateral in the cardiac catheterization laboratory during the initial hospitalization (Table 2).

Overall Outcomes

Survival at 1 and 10 years was 80% and 69%, respectively (Fig. 1). There were a total of four deaths (22%) at the time of analysis, three of which were related to sepsis, and one cardiac death. There were two (11%) deaths in the immediate post-operative hospitalization period (110 and 120 days after surgery). Patient 17 had biliary atresia and multiple comorbidities, including microphthalmia of right eye and anophthalmia of the left eye (Table 1). After the Kasai procedure, the patient developed wound dehiscence, pseudomonas infection, and chronic liver failure, and was not deemed a candidate for liver transplantation. There was mild to moderate left AV valve insufficiency on the echocardiogram prior to death. This patient's course was further complicated by renal failure, hypertension, hypothyroidism, hypoparathyroidism, stroke, tachyarrhythmias, and subsequent death. The diagnosis of biliary atresia was made after cardiac surgery. This approach would not have been utilized had this diagnosis been known preoperatively. The post-operative course of patient 16 was complicated by sepsis, respiratory failure, multi-organ system failure, and death. There was only mild left AV valve insufficiency on the latest echocardiogram. Two additional deaths occurred during the follow-up period. Patient 18 with Trisomy 21 died three months after hospital discharge related to RSV bronchiolitis with the most recent echocardiogram demonstrating no significant left ventricular outflow tract obstruction and mild

left AV valve insufficiency. Patient 15 died 6 years after hospital discharge related to LVOTO, mitral valve insufficiency, and heart failure. This was the only patient with moderate left AV valve insufficiency preoperatively. The initial surgery was performed at our institution; however, subsequent follow-up and two additional surgeries were performed at another institution.

Among the fourteen patients who survived, the median duration of follow-up was 68 months (0.6–144). Cardiac reintervention was performed in five patients (Table 1). One- and ten-year freedom from any cardiac reintervention was 66% and 40%, respectively (Fig. 2A). Freedom from LAVV reintervention was 85.6% at 1 year and 66% at 10 years from the primary repair (Fig. 2B). Closure or partial closure of an unrepaired cleft was performed in three patients at 23 days (patient 14), 10 months (patient 4), and 6 years (patient 1) after the primary operation. The left AVV reintervention was considered a secondary indication in the latter two patients (Patients 1 and 4), as left ventricular outflow obstruction was the primary indication for reintervention. Of note, three of the six survivors who had either partial or no cleft closure at the time of complete repair did not require additional left atrioventricular valve surgery during the follow-up period. A total of eight surgeries for recurrent left ventricular outflow tract obstruction were required in four patients during the follow-up period. No patients required any form of reintervention for arch obstruction. We did not have enough patients to demonstrate a correlation between AVV index (or modified AVV index) or number of papillary muscles with patient outcomes or need for cardiac reintervention.

Outpatient follow-up echocardiographic images were available for review in 13 patients (including the infant who died of RSV). The degree of left AVV regurgitation was mild or less in nine (69%), mild to moderate in three (23%), and moderate in one patient (8%). The degree of left AVV stenosis was mild or less in all patients. None of the patients had significant right AVV regurgitation nor stenosis. There were no residual ASDs. Two patients had a small, hemodynamically insignificant residual VSD. The LV internal diastolic dimension (z score -0.04, -1.86 to $+1.75$) and systolic function (shortening fraction: 37%, 33–46) were normal in all patients. There was no evidence of recurrent arch obstruction on follow-up echocardiography. None of the patients in this cohort required pacemakers for heart block during the follow-up period.

Discussion

Aortic arch obstruction poses a unique surgical challenge in patients with AVSD. We present our institutional experience of single-staged repair of CoA-AVSD over a 10-year period. Our report is novel in that it is the largest single center, single surgeon experience with this rare and difficult cohort. Furthermore, it contains 10-year mortality and freedom from reintervention data that is useful for counseling and guiding post-surgical follow-up. There has been long standing controversy regarding the ideal timing and surgical approach for CoA-AVSD. In 2010, Hraska et al. recommended a two-stage approach for patients less than two months of age with competent AV valves. They reserved the one-stage surgical approach for patients (< 2 months of age) with significant AV valve insufficiency in whom medical management with or without relief of arch obstruction by a trans-catheter approach was not feasible or did not relieve symptoms [6]. Two months of age was arbitrarily chosen as a distinguishing time point highlighting the challenge of repairing friable young AV valve tissue. This differs from our cohort in which complete repair was successfully preformed in 12 of 13 patients less than 60 days of age, 11 of whom were less than 30 days at the time of repair. With this approach, the majority (69%) of patients had mild or less left AV valve

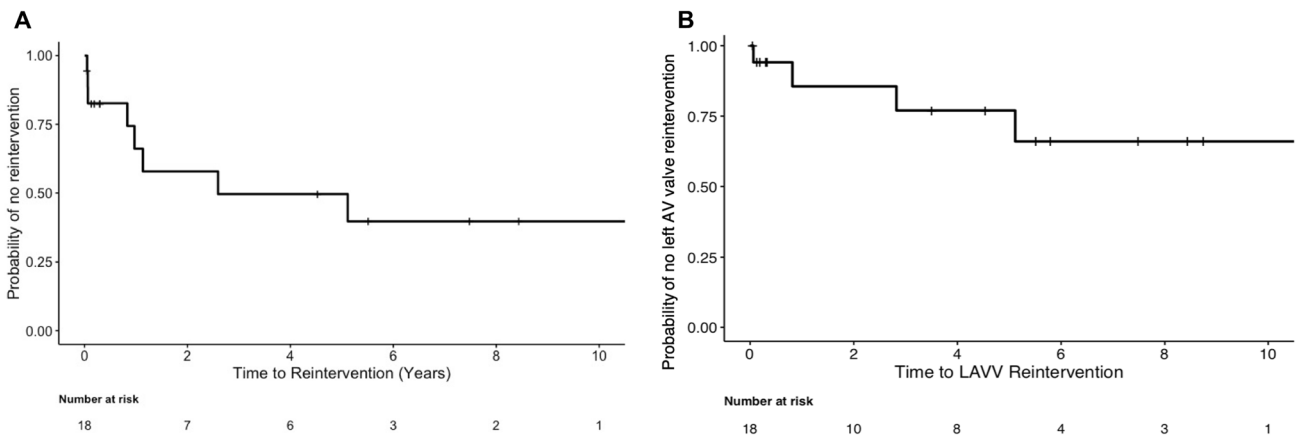

Fig. 2 Kaplan–Meier reintervention-free survival (**A**) and left atrioventricular valve reintervention-free survival (**B**)

regurgitation, no patients had more than mild left AVV stenosis, and only 22% required left AV valve re-intervention at 68-month follow-up demonstrating the durability of early repair. Our experience suggests that single-stage repair need not be reserved for those with significant AV valve insufficiency as suggested by Hraska et al., instead perhaps selecting patients with good function, less AV valve insufficiency is the key to successful single-stage repair, and the majority of patients did not require subsequent cleft closure during a median follow-up of more than 5 years.

Previous studies recommended single ventricle approach for complete AVSD patients with a large VSD and AVVI < 0.67 [8]. In our cohort, there were five syndromic patients with "smallish" left ventricles (AVVI < 0.67) who were not considered desirable candidates for single ventricle palliation and thus single-stage biventricular repair was attempted. Three were successful. There were two deaths related to sepsis, rather than failed biventricular approach suggesting that this index may not apply to AVSD with CoA. The syndromic right ventricular dominant AVSD patients with CoA and a single left-sided papillary muscle pushed to a biventricular repair made us realize that by portioning the AVSD early the left side grew significantly in the early postoperative period and the AV valve worked reasonably well despite not closing the cleft. Several patients benefited from one pericardial pledgeted stitch (7-0 GORE-TEX) in the left AV cleft right at the crux of the heart to impart the appropriate "roll" of the superior and inferior bridging leaflets so the left AV valve coaptation was optimized which essentially left the cleft open but stabilized the valve. Early in our experience prior to 2005, isolated arch repair in this patient population led to long postoperative courses and unsatisfactory results. Thus, the question became which is worse, a pulmonary artery band or AVSD repair with leaving the cleft open? In the majority of the patients who underwent reintervention on the left AV valve, this was performed at the time of left ventricular outflow tract resection with the outflow tract obstruction being the driver for reoperation rather than mitral insufficiency. Only one patient, a partial AVSD, required isolated cleft closure, thus we felt it important to include patients with partial AVSD in the cohort in order to fully disclose our experience with early arch repair leaving a left AV cleft open, as well as, to highlight the challenges of this subset of AVSD-CoA patients.

In 2013, Shuhaiber et al. retrospectively compared multicenter outcomes of single-stage versus two-stage repair of AVSD–CoA suggesting the two-staged approach is superior. Our cohort had similar characteristics such as age at surgery (16 vs.11 days), birth weight (3.2 vs. 3.1 kg), weight at surgery (3.6 vs. 3.0 kg), and percentage of patients with right dominant AVSD (53% vs 56%) in comparison to their single-stage neonatal CoA-AVSD repair population.[7] Our number of post-operative deaths (11.1%, 2 of 18 patients) was lower than the post-operative mortality rate of single-stage repair (25.8%, 8 of 31 patients) and comparable to the two-staged repair of AVSD (9%, 3 of 33 patients) reported in their study [7]. None of our patients required ECMO support in contrast to the published single-staged CoA-AVSD repair cohort 9.67% (3 of 31 patients) [7]. In our cohort, however, more patients underwent delayed sternal closure (66.7%, 12 of 18 patients) as compared to that multicenter study (32.2%, 10 of 31 patients). Tracheostomy support was required in one patient in both our cohort and the previously reported single-stage repair cohort [7]. Our median duration of ventilation was 4 days (2–27) excluding one patient who required tracheostomy. This was similar to that of both single- and two-stage repair cohort [8 days (4–23) vs 9 days (4–20), respectively] reported previously. Our median lengths of stay [ICU16 days (4–60) and hospitalization 28 days (12–68)] were similar to the previously published single-stage CoA-AVSD repair [ICU 17 days (10–27) and hospitalization 28 days (12–40), respectively] [7].

Our study contrasts with Shuhaiber et al.'s findings with lower early (within 30 days) cardiac reintervention rate with only one patient requiring closure of cleft in the LAVV at 23 days after primary repair. On pre-discharge echocardiogram, 25% had moderate left AVV insufficiency compared to 42% of the multicenter single-stage repair cohort [7]. Our freedom from left AVV re-intervention was better at 86% at 1 year and comparable at 6 years (66%) to the published literature (about 70% at 1 year and 65% at 6 years) [7]. None of our patients had recurrent aortic arch obstruction, in contrast to the multicenter experience which reports an unplanned Norwood operation 16 days after a neonatal coarctation repair in a transitional AVSD patient [7]. None of our patients' heart block requiring pacemaker placement. Our 6-year freedom from re-intervention (40%) was comparable to that of single-stage group in Shuhaiber's cohort (45% at 6 years), both of which are significantly lower than the long-term freedom from re-intervention reported in isolated CAVSD to be 88% at 10 years again highlighting morbidity added to the diagnosis of AVSD when arch obstruction is present [10, 11]. The most common re-interventions performed in our group include relief of LVOTO [four patients (22%) requiring a total of eight surgeries] followed by re-intervention of the left AVV [four patients (22%) requiring a total of five surgeries].

In our experience of single-stage CoA-AVSD repair, the 1-year survival was 80%. This contrasts with the multicenter experience describing 60% 1-year survival in the single-stage and 80% survival in two-stage repair of CoA-AVSD [7]. Our overall 6-year survival for single-stage repair CoA-AVSD (69%) was comparable to that reported by Shuhaiber et al. (57% for single-stage CoA-AVSD repair and 78% for patients whose first cardiac operation occurred between 2002 and 2009) [7]. The mid-term survival in both

reports is much lower than that of isolated AVSD repair, whose 8-year survival is reported to be 92.2% (surgical era 2000 to 2011) emphasizing the significant increased risk of mortality associated with a diagnosis of arch obstruction with AVSD [10]. In select patients, it is possible to avoid the deleterious effects of a pulmonary artery band or unrepaired atrioventricular septal defect after arch repair, or conversely the deleterious effects of even moderate aortic arch obstruction after atrioventricular septal defect repair with single-stage CoA-AVSD repair.

Limitations

Despite a reasonable sample size of patients with single-stage repair CoA and AVSD operated at a single institution by a single surgeon, our study is limited by its retrospective design and lack of comparison to the two-stage approach. Patients were identified from a surgical database, thus patients with moderate or severe AV valve insufficiency or poor ventricular function were not candidates for surgery and thus were not included. As a referral center, it was challenging to evaluate long-term outcomes in patients followed by cardiologists outside our center.

Conclusion

Our experience highlights the feasibility and relative safety of the single-stage repair of CoA and AVSD in selected patients. Survival and freedom from reintervention in our cohort approximate those outcomes seen with two-stage repair. These patients are frequently syndromic and demonstrate mortality risk from non-cardiac causes. Left ventricular outflow tract obstruction appears to be the most common recurrent lesion in this population. Left ventricular systolic function and dimensions were normal at latest follow-up, with durable left AV valve function, no recurrent arch obstruction, and no significant heart block. Ultimately the choice of approach depends on multiple factors including other anomalies or syndromes, availability of transcatheter intervention, and surgical expertise and/or preference. Consideration of a single-staged approach is warranted for patients with CoA-AVSD.

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Declarations

Conflict of interest The authors have no competing interests to declare that are relevant to the content of this article.

Ethical Approval All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript. The authors have no financial or proprietary interests in any material discussed in this article.

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