



Outcomes of Total Repair of Atrioventricular Canal Defects: A Two-Decade Single-Centre Experience

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Abstract

Background: Atrioventricular canal defect (AVCD) is considered to be one of the complex congenital heart diseases. It is categorized as a spectrum, from the simple partial AVCD to the most complex, the complete AVCD. Surgical repair of AVCD is the optimal management option for this condition. We carried out this study to analyse our centre's experience regarding the outcomes of the management of this condition. **Patients and Methods:** Medical records of patients who underwent AV canal repair from January 2003 to December 2023 were reviewed retrospectively and the clinical data, including age at operation, weight, type of surgery, associated co-anomalies, palliation, etc., were collected and assessed. The outcomes of surgery, including mortality, length of hospital stay, pacemaker insertion for Atrioventricular block and redo surgery, were analysed. **Results:** The study looked at 110 patients diagnosed with AV canal defect and underwent total corrective repair during the period from January 2003 until December 2023. Fifty-nine of the patients (53.6%) were females. Out of the 110 patients, 45 patients (40.9%) were diagnosed with Down syndrome. 59% of these had CAVCD. Among those with associated malformations 14 had PDAs and 5 had Tetralogy of fallot. 8 (12.9%) of these patients had to undergo palliative procedures before definitive surgery. Median (IQR) age at the time of surgery was 6 (4 - 9) months with a mean (SD) weight of 12.1 (16.6) kg. The choice of operative procedure was the double-patch surgical technique for CAVCD and patch repair of ASD and cleft repair for the partial AVCD. In the postoperative period, we had 4 (3.6%) deaths in hospital, with 3 dying on the

table. Five patients had permanent pacemakers inserted due to AV block. The average length of hospital stay was 15 days, with patients post CAVCD repair spending longer in the 3 categories, with a mean length of stay of 16 days. There were 5 (4.5%) reoperations before discharge, 3 for severe LAVVR, 1 for residual VSD and 1 for both residual VSD and severe RAVVR. 5 patients were readmitted for reoperation, with 3 for severe LAVVR and 1 for PDA closure and another for subaortic membrane resection. **Conclusion:** From our study, Atrio-ventricular septal defects, especially the CAVCD, were safely corrected between 3 and 6 months with less morbidity. The other types were safely operated on later after 1st year of life, especially the partial type. The commonest cause of redo surgery is the regurgitation of the LAVV component, especially in patients with CAVCD. AV block occurs more in patients with CAVCD and subsequent permanent pacemaker insertion. Overall, partial AVCD and intermediate AVCD have less morbidity compared to CAVCD with less mortality, shorter hospital stays and complications of AV blocks.

Subject Areas

Cardiology

Keywords

Atrioventricular Canal Defects, Complete Atrio Ventricular Canal Defect, Complete Repair, Outcomes

1. Introduction

1.1. Definition

Atrio ventricular canal defects (AVCD) also known as endocardial cushion defects or atrio ventricular septal defects are defined by the presence of a common atrio-ventricular valve and variable defects in the atrioventricular (AV) septum [1].

AVCD comprise of 3% - 7% of congenital heart diseases (CHD) [2]-[4]. Atrio-ventricular canal occurs in two out of every 10,000 live births. Both sexes are equally affected [5] [6]. Atrioventricular defects can exist alone or in association with other congenital cardiac anomalies. They are also commonly associated with chromosomal anomalies, especially trisomy 21 (Down syndrome), though they can occur in non-syndromic patients. Endocardial cushion defect is the commonest form of congenital heart disease in children with Down syndrome [7].

AVCDs are categorized into different types according to the anatomical description and Kirklin Barrat-Boyes (pg 1230, 4th edition) describes them according to 3 groups.

The simplest type, Partial AV septal defect or ostium septal defect, in which there is an interatrial communication but no interventricular communication and a connection of variable width between the Left superior leaflet (LSL) and left inferior leaflet (LIL).

The intermediate or transitional AV septal defects, which are varied but usually include the presence of two AV valve orifices and a restrictive inlet ventricular septal defect (VSD), with dense chordal attachments to the ventricular septum.

The most extreme form, Complete Atrio ventricular canal defects (AVCD), with large deficiencies in atrial and ventricular septa, a common AV valve orifice, and large interatrial and interventricular defect.

1.2. Diagnosis of AVCD

Patients can present early in life with features of congestive heart failure.

1.2.1. Echocardiography

Echocardiography is the mainstay of diagnosis of AVCD. It is able to show the features of the anomaly that include primum atrial septal defect, ventricular septal defect, and presence of the common atrioventricular valve. It can be used to identify the type of AVCD that we are dealing with by clearly identifying if there's one common valve as in CAVCD or two separate valves as in the intermediate and partial types. In the CAVCD it can further be used to give details on the anatomy of the leaflets of the single valve and hence determine the Rastelli classification. The echocardiographic examination further shows the degree of dysfunction of the common atrioventricular valve, as well as the presence of associated cardiac malformations. Another important investigation is cardiac catheterization, which is used to give an accurate assessment of the left-to-right shunt as well as the severity of pulmonary hypertension and the reversibility of pulmonary resistance.

AVCD can also be diagnosed prenatally and this is an important aspect in planning for the management of the newborn. With two-dimensional echocardiography, atrioventricular canal malformations can be diagnosed accurately. Color Doppler flow mapping gives additional hemodynamic information that is crucial in the prognosis of the patient [8] [9].

1.2.2. Catheterization

These studies are mostly required only when major cardiac anomalies coexist and when operability is questioned because of evidence of pulmonary vascular disease, especially in patients above 6 months of age. Over time, in patients with CAVCD, pulmonary hypertension becomes irreversible, thus precluding the surgical therapy. This is the reason why cardiac catheterisation is not mandatory in infants (less than 6 months) but is indicated in older patients if irreversible pulmonary hypertension is suspected [5]. Direction and magnitude of shunting; pulmonary and systemic pressures, resistances, and flows; and right and left ventricular pressures can be measured and calculated from data obtained at cardiac catheterization.

1.3. Types of Atrioventricular Canal Defects

1) Partial atrioventricular canal defects

This is the simplest form of the spectrum and as defined before is characterized

by an interatrial communication but no interventricular communication and a connection of variable width between the Left superior leaflet (LSL) and left inferior leaflet (LIL). The atrioventricular valve is actually separated into two entities. Primum ostium defects generally account for between 5% and 10% of all ASDs [10].

It is more common in patients without Down syndrome compared to those with a complete AV canal defect [6].

Repair of partial AV canal defects or primum atrial septal defects can be postponed until 18 to 36 months of age if the patient does not have pulmonary hypertension or growth failure [11].

2) Pathophysiology and presentation

In partial AVCD the shunt is at atrial level and usually large, but it may be small or moderate and, in such cases, a pressure gradient can be demonstrated between the left and right atria. When the shunt is large and left AV valve regurgitation is mild or absent, the hemodynamic state of the patient is identical to that in isolated ASD, right ventricular stroke volume is increased. When important left AV valve regurgitation is present, the left-to-right shunt becomes much larger and the regurgitation jet goes directly from LV to right atrium. Left as well as right ventricular stroke volume is increased, and marked cardiomegaly and heart failure develop early in life.

3) Symptoms

Patients often present in the first decade of life but may remain asymptomatic well beyond that age.

Their clinical presentation is virtually identical to that of patients with the more common fossa ovalis ASD.

Associated Left AV valve regurgitation in these patients may produce symptoms earlier, and progressive severe heart failure may require treatment in infancy. They will typically have a loud apical pansystolic murmur, and the apex of the LV may be palpable. Tachypnea and hepatomegaly can also be seen.

4) Management

Due to the improvement in diagnostics for congenital heart disease, Partial AVCDs are diagnosed early in life and surgery can be planned early. Surgery in these patients is preferred to be done in the preschool years. However, some patients with partial AVCD present later in life. They usually present with arrhythmias such as atrial fibrillation and late surgery usually brings more pre-operative complications [12].

Surgery involves repair of the cleft of the left atrioventricular component of the valve to correct the regurgitation if any and patch repair of the primum ASD with either an autologous or heterologous pericardial patch. Surgery is performed via median sternotomy and on cardiopulmonary bypass as in all the other types of AVCD.

Through a right atriotomy, the heart is opened to access the defect. The valve is assessed. The left component can be visualized through the primum ASD. The

cleft in the left component is then repaired as below and valve tested for competency.

This is then followed by closure of the ASD with pericardium and the closure of the right atrium.

5) Risk factors and outcomes of surgery of partial AVCD

Generally, unless associated with severe left-sided AV valve regurgitation or some other significant cardiac lesion, PAVSDs can be repaired with an operative mortality below 1% [10].

As in all the types of AVCD, the risk of damage to the AV node during surgery is higher due to its displacement postero-inferiorly into a new nodal triangle bounded by the annulus of the right AV valve, the coronary sinus, and the lower edge of the interatrial septum as described by Anderson [13].

For the patients that present late in adulthood, surgery still has good outcomes with low morbidity. One study showed repair of Partial AVC in adults may be performed at low risk and may improve patient longevity and functional status [14]. Another study showed that though adult operations bring more pre-operative complications, mortality and reoperation rates were excellent [12].

Another risk factor of reoperations and mortality irrespective of age at operation was found to be preoperative severe regurgitation of the mitral component of the atrioventricular valve [15].

6) Intermediate/transitional AV canal defects

The intermediate types usually include presence of two AV valve orifices and a restrictive inlet ventricular septal defect, with dense chordal attachments to the ventricular septum. This is also commonly associated with Down syndrome. Presentation is as in partial AV canal defect because the VSD is usually restrictive.

Surgery is usually as in the partial AVCD with sometimes direct closure of the restrictive VSD or patch repair of the VSD depending on the size.

1.4. Complete Atrioventricular Canal Defects

Complete Atrioventricular canal defect (CAVCD) is the most complex form of the spectrum of endocardial cushion defects that are seen in children. Complete atrioventricular defect includes ostium primum atrial septal defect (ASD), a common atrioventricular valve and a confluent posterior ventricular septal defect (VSD) usually located in the inlet portion of the ventricular septum [4].

CAVCD is further classified into 3 types: Type A, Type B and Type C according to Rastelli's classification [5].

1) Epidemiology and risk factors

CAVCD accounts for about 3-7% of all cardiac malformations and can occur as an isolated or as part of a genetic syndrome [1] [4]. Fulton and colleagues showed that a third of the patients with a complete atrioventricular canal defect have Down syndrome [16]. Another study showed that in n patients with Down syndrome, the complete form of atrioventricular canal was found to be prevalent, with a high frequency of associated Fallot's tetralogy [6]. Maternal diabetes has

also been found to be a risk factor for the most severe form of AVCD [17].

2) Pathophysiology of CAVCD

CAVCD is characterised by an ostium primum atrial septal defect, a common atrioventricular valve and a variable deficiency of the ventricular septum inflow. It was classified by Rastelli into types A, B and C.

Rastelli classification of complete atrioventricular canal defects. Type A defect, extensive chordal attachments of leaflet to septum; Type B defect, left superior leaflet attached to the right side of the septum; Type C defect, with free floating superior leaflet [18]. CAVCD is also further divided into balanced or unbalanced types.

The common atrioventricular valve may be positioned equally over the ventricles making it balanced or unequally over the right or left ventricle making it unbalanced with varying degrees of associated ventricular hypoplasia [19].

In CAVCD there is both interatrial and interventricular shunting, leading to right ventricular pressure and volume overload and pulmonary hypertension [5]. There can also be an association of insufficiency of the common atrioventricular valve further complicating the presentation.

Children present earlier on with features of heart failure as a result of the ventricular pressure and volume overload with symptoms such as failure to thrive, feeding difficulties, respiratory problems, and hepatomegaly. It is very rare for a child to survive beyond the first year of life without serious morbidity. Eventually, if unoperated irreversible pulmonary resistance develops, leading to reversal of the intra-cardiac shunt and cyanosis develops, further reducing the exercise capacity [20]. When untreated, with time the children with CAVCD develop complications of pulmonary over circulation making surgery impossible [5].

3) Clinical presentation

Patients with complete AVCD usually present with symptoms early in life, before 6 months of age due to pulmonary overcirculation.

Tachypnea and difficulty gaining weight are the first signs noticeable in patients with AV canal defects. The occurrence and severity of symptoms depend on the degree of AV valve regurgitation and other associated CHD, which can contribute to the early development of congestive heart failure (CHF) [2] [20]. Symptoms of Congestive Heart Failure include increased work of breathing, sweating while feeding, poor feeding, and lethargy. Signs of Congestive Heart Failure include tachypnea, tachycardia, failure to thrive, wheezing or rales on lung auscultation, S3 gallop rhythm, apical displacement of the apical impulse, hepatomegaly, or increased jugular venous pressure (JVP).

A careful and complete cardiac exam should be done to look for:

- a) Wide and fixed split S2 heard due to left-to-right shunting across the ASD, causing increased blood flow in the right side of the heart irrespective of the phase of respiration.
- b) S3 due to increased flow of blood splashing across the left ventricular walls which are still compliant.

c) Additional murmurs such as:

- Holosystolic murmur secondary to left AV valve regurgitation is heard best at the apex.
- Mid diastolic murmur if the shunt is large or if there is significant AV valve regurgitation.

4) Management of CAVCD

Surgery is the optimal management for CAVCD and has evolved over the years with improvement in the mortality and other outcomes [21].

There are two widely used techniques used and these are the Two patch repair and the modified single patch technique. However, surgery for CAVCD has also evolved, it started with suturing the common AV superior and inferior leaflets to the crest of the ventricular septum using interrupted mattress sutures. The inter-atrial communication was closed with patch material, or pericardium before the Rastelli classification. It then moved on to the use of the classical single patch repair after the improved understanding of the AVSD anatomy. This involved creating an anterior mitral valve leaflet by attaching the common AV valve leaflets together using interrupted stitches. The septal defects were closed with a single patch of either autologous pericardium or Teflon. The technique of AV valve reconstruction described “splitting” the common superior and inferior valve leaflets (when necessary) from the free edge to the annulus, slightly to the pulmonary ventricular side of the midline, and then attaching the reconstructed leaflets to the septation patch [22].

Now we have moved to the modified single patch and double patch repair with different institutions following what suits them best. However, both techniques continue to be studied and so far show comparable results, especially with mortality [23] [24].

The two-patch repair is performed as described below by a number of authors [25] [26]. It is done through a right atriotomy. After inspection of the common valve, cold saline is used to fill the ventricular chambers and float the AV valve tissue into a closed position to establish the line of coaptation between the superior and inferior components of the valve and to identify the midpoint that separates the valve into right and left components. This is done by placing a stitch at the mid-point. The VSD is then closed by a Gore-Tex patch onto the right side of the defect with running 5.0 Prolene, taking care of adequate coaptation at the meeting point of the superior and inferior leaflets on both the right and the left sides. Closure of the ASD is begun with a running Prolene using an autologous pericardial patch, starting in the commissure between the right mural leaflet and the inferior bridging leaflet, following the hinge point level of the leaflet and continuing onto the previous suture line at the AV level of the VSD patch. Valve testing is then done to identify the cleft of the left component. Using horizontal interrupted sutures Prolene 6.0, the left-sided cleft is closed.

The modified single patch differs from the two-patch repair in the way the VSD is closed. VSD closure in this technique is achieved by bringing the septal compo-

nents of the LSL and LIL down to the intraventricular septum and the rest of the procedure is completed as in the two-patch repair [27]. This technique was extensively explained by Graham R Nunn and colleagues 2004 [28].

In the current era definitive repair is the choice of management with palliation procedures only done in a few cases of high-risk patients [29] [30]. The choice of technique to use is based on institution protocol and personal preference and comfort.

5) Outcomes and risk factors of surgical repair of CAVCD

CAVCD repair has evolved over the years as has all congenital cardiac surgery. This has been facilitated by improvement in techniques, diagnostics and anesthesia services. The outcomes and risk factors have been studied and continue to be studied as there's still a lack of consensus on a number of aspects.

Timing of surgery is an important aspect in the management of CAVCD and contributes to the outcome of the surgery. Uli and colleagues noted that an optimal age for operation of these patients is between 3 and 6 months. They noted that repair before 3 months of age and the need for cleft closure were associated with higher degree of (left atrioventricular valve regurgitation) LAVVR at discharge [31].

However, another study done by stellin and colleagues showed that operating children below 3 months of age gave better results with lower mortality rates and higher freedom from reoperation for left AV regurgitation [32].

It was also noted that in symptomatic patients there was no need to delay surgery since mortality and reoperation outcomes were comparable between the younger and older infants [33]. Other Studies also showed that the outcomes of the repair largely depend on age at surgery [34].

Associated defects and associated syndromes were also seen to be risk factors in CAVCD surgery [35]. However, others have noted that in the current era Down syndrome is not a risk factor for reoperation [31] [36]. Another study done by Danielle and colleagues also showed that Down syndrome had no effect on timing of surgery, other outcomes and reoperation [37].

The overall mortality rate has been seen to go down over the years with improved survival of these patients with the highest cause of re-operations being LAVVR. Mortality has been defined early if it occurs within 30 days or before discharge and late if it occurs 30 days post-surgery [38].

Other outcomes that have been studied over the years include hospital and ICU stay and rhythm complications. Weight at operation has been noted to contribute to morbidity of patients as well. Yosef and colleagues found that children operated on at a weight under 4 kg had longer ICU and ward stay as well a higher incidence of atrioventricular regurgitation postoperatively [39].

Also low weight at operation was associated with mortality and still long hospital stay by another study [37].

This study looked at the experience of our center in the outcomes of the surgical repair of AVCD.

1.5. Research Questions

What are the outcomes of surgical repair of AVCD?

What are the associated risk factors of mortality and reintervention surgery post repair of AVCD?

1.6. Objectives

To determine the outcomes of surgical repair of AVCD.

To assess the associated risk factors of mortality and reintervention surgery post-surgical repair of AVCD.

2. The Study

2.1. Patients and Methods

Pediatric and congenital cardiac surgery in Italy is carried out by a number of centers of which our center Niguarda hospital is part. We retrospectively reviewed the medical records of patients who had AVCD repair at Niguarda hospital from January 2003 to December 2023. The operation notes and follow up notes were studied. Of the 126 patients identified, 110 met the required criteria. 16 patients were excluded because they were not fit for biventricular repair or had missing study information.

The following parameters were investigated: preoperative age in months at repair, sex, weight in kilograms, type of AVCD, associated syndromes, associated co-anomalies, initial palliative procedure, competency of univentricular valve, operative (X-clamp time and cardiopulmonary by-pass time in minutes). All the patients were subjected to follow-up and post-operative outcomes, *i.e.* mitral regurgitation (MR), arrhythmias such as atrioventricular block (AV block) and mortality were recorded. The mean follow up time of the patients was 9 years.

2.2. Surgical Technique

During the study period for complete AVCD, the 2-patch repair technique was the standard technique used. The heart is opened through a right atriotomy and anatomy is appreciated. Saline testing is done to assess the anatomy and symmetry of the common AV valve during the valve repair. The VSD is closed with a polytetrafluoroethylene patch. The left atrioventricular valve component was always closed without consumption of valve tissue to avoid stenosis of the left AV component. The stitches are placed in such fashion as maintaining as much coaptation height as possible when the LAVV cleft is closed. During valve repair, caution was taken not to create unnecessary tension in the leaflets, preserve maximum amount of valve tissue, avoid cutting secondary chords, and respect the natural symmetry of the valve. The primum atrial septal defect was then closed using an autologous or bovine pericardial patch. Generally, the coronary sinus was maintained on the right side of the heart, and in all the patients, the cleft in the LAVV was completely closed.

For the partial AVCD, the procedure was as follows: the defect is accessed as above. The cleft repaired as above and the primum ASD closed with heterologous pericardium.

In the intermediate defects, the restrictive VSD was examined and if need be, repaired primarily or with a bovine patch and the rest of the repair as above.

2.3. Study Outcome Measures

The primary outcome measures were in hospital mortality and reintervention surgery.

Hospital mortality was defined as death before discharge after operation.

Reoperation was defined as reoperation less than 30 days after initial AVCD repair, and late reoperation was defined as reoperation more than 30 days after initial AVCD repair or after discharge.

Permanent pacemaker placements are not included as re-operations but discussed separately. Only pacemakers placed post operatively were considered.

2.4. Data Analysis

Data was analyzed using STATA version 14.1 (Stata Corp LP, Texas, USA). Continuous variables were summarized using means, standard deviations if normally distributed and medians and interquartile ranges if skewed. Categorical variables were summarized using frequencies, proportions, and percentages. We visualized the data using bar graphs and pie charts. For the different outcomes of mortality, AVPM insertion, reintervention, we summarized these as proportions of individuals who had the outcome as compared to the total sample size. The hospital length of stay was summarized as median with its interquartile range.

Logistic regression was used to determine the factors associated with mortality and reintervention separately. Odds ratios with their 95% confidence intervals and p-values were calculated. Multivariate analysis was conducted using logical model building taking into consideration variables with p values ≤ 0.2 at bivariate analysis, those with biological plausibility and known confounders. We assessed interaction by building two-way interaction terms and testing their significance using the likelihood ratio test. None of the interaction terms were significant. We assessed for confounding by considering a 10% change in the odds ratio between the crude estimate and the adjusted estimate for each variable in the model. We assessed the goodness of fit of the model using the Hosmer and Lemeshow goodness of fit test and presented the final model using the adjusted odds ratios and their 95% confidence intervals. A p-value of less than ≤ 0.05 was considered as statistically significant.

3. Results

3.1. Descriptive Statistics

We identified 110 patients diagnosed with AV canal defects and underwent repair

during the period from January 2003 until December 2023 as shown below in **Table 1**. Fifty-nine of the patients (53.6%) were females. Of the 110 patients, sixty-two (56.4%) had complete AVCD, thirty-one (28.2%) had partial AVCD and seventeen (15.4%) had intermediate AVCD.

The commonest associated defect of all defects was PDA (12.7%) followed by additional ASD (11.8%). Considering all defects, the double patch technique was the commonest used procedure and Custodial cardioplegia was used in 96 of the patients (87.3%). In all patients, the mitral cleft was repaired.

Most of the children were operated on between the age of 3 months and 12 months.

Table 1. Baseline clinical characteristics N = 110.

Characteristic	Frequency	Percent
Sex		
Female	59	53.6
Male	51	46.4
Cardioplegia		
Custodiol	96	87.3
Del Nido	14	12.7
Ass Malformation (n = 110)		
TOF	5	4.5
PDA	14	12.7
Additional ASD/foramen ovale	13	11.8
hypoplastic aortic arch	1	0.9
coarctation of Aorta	2	1.8
None	68	61.8
Coronary sinus syndrome	2	1.8
azygous continuation of IVC	2	1.8
valvular heart disease	3	2.7
Ass. Surgery (n = 110)		
additional Secundum ASD + foramen ovale closure	13	11.8
PDA	12	10.9
TOF	5	4.5
coarctation of the aorta	1	0.9
hypoplastic aortic arch	1	0.9
None	71	64.5
Tricuspid valve procedures	4	3.6
Others	4	3.6

Continued

Rerouting pulmonary veins	2	1.8
Technique		
Modified single patch and other techniques	50	45.5
Double patch	60	54.5
Chest closure		
No*	9	8.2.
Yes	101	91.8
Age at surgery (months)		
>3 months	13	11,8
3 - 6 months	41	37.3
7 - 12 months	38	34.5
>12 months	18	16.4

*3 died on table.

Table 1 shows the baseline factors looked at in the study. PDA-patent ductus arteriosus, TOF-tetralogy of fallot, ASD-Atrial septal defect, IVC-inferior vena cava.

We were able to show the different syndromes associated with AVCD in our cohort, as shown in **Figure 1**. Out of the 110 patients, 45 patients (41%) were diagnosed with Down syndrome. The other syndromes noted were Noonan, VACTERL and pluriformal syndromes.

In **Figure 2** above, 10 (9.1%) of the patients had to undergo palliative procedures

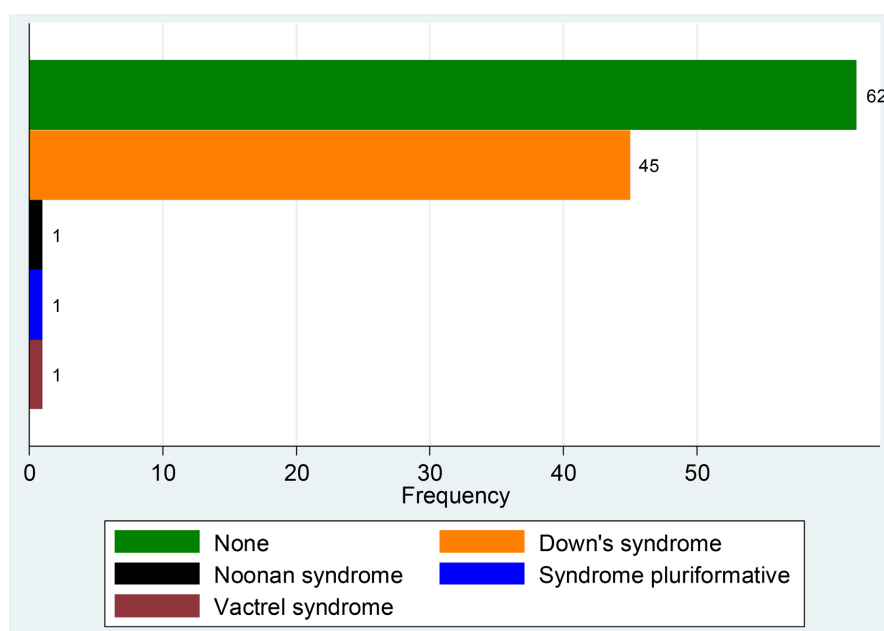


Figure 1. Associated syndromes.

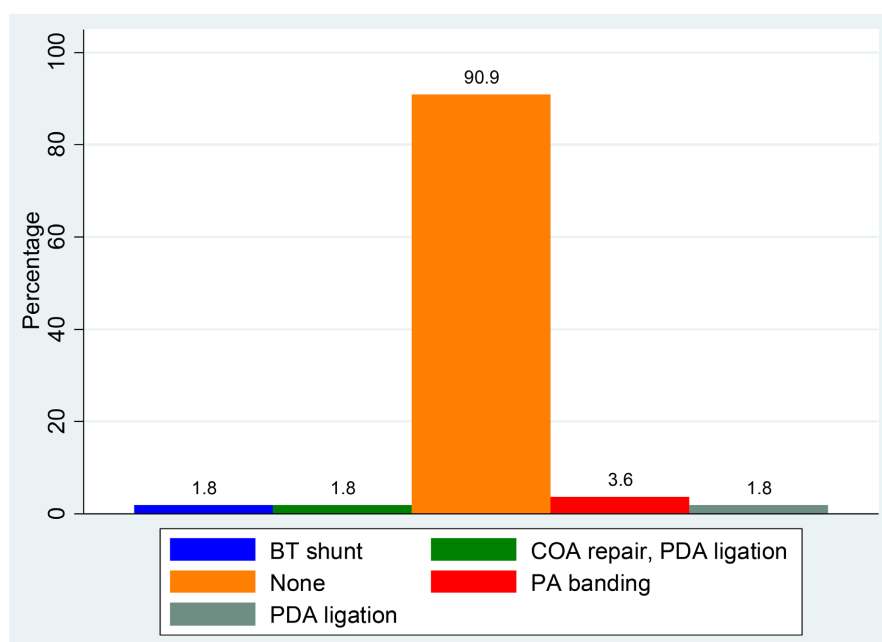


Figure 2. Bar graph of palliative procedures.

before the definitive surgery. Two of these had TOF with right ventricular outflow obstruction and underwent modified BT shunt, four underwent PA banding, two had PDA ligation and another two had both coarctation repair and PDA closure.

Analysis of the continuous outcomes was done as shown in **Table 2**.

Table 2. The continuous outcomes. Median (IQR) age at the time of surgery was 6 (4 - 9) months with a mean (SD) weight of 12.11 (16.6) kg.

Characteristic	Minimum	Maximum	Mean	Sd	Median	Interquartile range (1 st -3 rd quartile)
Age	2	42	8.25	7.96	6	4 - 9
Weight	2.3	92	12.11	16.6	6	5 - 11
ECC	49	346	128.07	52.2	124	91 - 154
Xclamp	25	235	87.47	36.1	85	60 - 110

3.2. Outcomes

The mortality in the study was calculated, giving a percentage of 3.6% as depicted in **Table 3** below.

Table 3. Mortality, *3 died on table N = 110.

Characteristic	Frequency	Percentage
Mortality		
No	106	96.4
Yes	4*	3.6

In the postoperative period, we had 4 (6.5%) deaths in hospital, with 3 dying on table in the operating room and the 4th child died in ICU on day 3 post operation. Of the 4, three were less than 6 months at time of operation and 2 (50%) of them had had prior palliative surgery. Also, notably for all the 4 patients Custodiol cardioplegia was used. The 3 patients who died in theatre were reported to have had unsuccessful weaning off Cardiopulmonary bypass. The cause of death for the patient in ICU was recorded as cardiorespiratory failure.

Table 4. Reoperation rates before and after discharge N = 110.

Characteristic	Frequency	Percentage
Reoperation before discharge		
No	105	95.5
mitral valvuloplasty	3	2.7
repair of residual VSD	2	1.8
Reoperation after discharge (n = 110)		
No	104	94.5
mv valvuloplasty	2	1.8
PDA closure	1	0.9
MV and tricuspid valve plasty	2	1.8
Removal of subaortic membrane and commissurotomy	1	0.9

As shown in **Table 4**, there were 5 (4.5%) reoperations before discharge, 3 for severe LAVVR, 1 for residual VSD and 1 for both residual VSD and severe RAVVR. Of the 5, all four were diagnosed with CAVCD and only one had Partial AVCD. The patients with severe LAVVR included a 2 yr old child with down syndrome who was reoperated on day 5 and had mitral valvuloplasty with edge-to-edge plasty, a 6-month-old child who had undergone modified single patch repair and was reoperated on day 22 post op and also repair was by edge-to-edge stitch of the residual cleft. Both patients had mild regurgitation of the LAVV pre-operatively. The 3rd child who had a mitral valvuloplasty was reoperated on day 2 due to severe MR, was also noted to have severe LAVRR pre-op. The child was 8months at time of operation and was the only one with diagnosis of partial AV canal. The 4th child was reoperated on day 30 post op due to a residual VSD and severe RAVVR. The residual VSD was closed with pledgeted prolene stitches and the valve repaired by closure of the cleft on the right side. The right components of the atrioventricular valve had mild regurgitation preop. The 5th child had CAVCD with associated TOF and was reoperated on day 4 for a residual VSD. All went on to be discharged successfully.

Five patients were readmitted for reoperation with 3 for severe LAVVR, and 1 for PDA closure. Of the 2 patients with severe LAVVR, one was reoperated on after 11years due to severe LAVVR and moderate RAVVR and had mitral valve

replacement with a mechanical valve, Carbomedics Optiform size 29 mm and tricuspid plasty, the second was reoperated after 13 years and had a mitral valvuloplasty with an annular ring of size 32mm. The third patient was reoperated after 8 years due to a small cleft and perforation in the anterior leaflet. All the 3 patients had moderate regurgitation of LAVV before the initial surgery. The 4th patient was found to have a large PDA causing symptoms after 7 years and underwent percutaneous closure of the defect. The last patient was re-operated for aortic valve disease with removal of a subaortic membrane.

The study showed that five patients had permanent pacemakers inserted due to AV block post-operatively, as seen below in **Table 5**.

Table 5. Patients who had permanent pacemakers inserted, AV-atrioventricular, PM-pacemaker.

Av block PM implant	Frequency	Percentage
No	105	95.5
Yes	5	4.5

We also looked at length of hospital stay as in **Table 6** and the average length of hospital stay was 18 days.

Table 6. Length of hospital stay.

Characteristic	Minimum	Maximum	Mean	Sd	Median	Interquartile range (1 st - 3 rd quartile)
Length of hospital stay	7	48	15.86	8.82	13	10 - 21

Looking at the AV canal defects as a whole in terms of outcomes

Table 7. Frequency of the different types of AV canal defects.

Diagnosis	Freq.	Percent
CAVCD/Complete	62	56.4
Intermediate	17	15.4
Partial	31	28.2

Table 7 above, out of the 110 patients studied, most were diagnosed with CAVCD 62 (56.4%), followed by partial AVCD 31 (28.2%) and Intermediate AVCD 17 (15.4%).

In the study as in **Table 8** below, it was noted Down syndrome was commonest in patients with AV canal defects, 37 (59.7%). Of the six patients who had a pacemaker inserted, 5 had surgery for CAVCD, while the one who had a partial AV canal defect had the pacemaker inserted before surgery due to sinus node disease.

The reported deaths (4) mortality of 3.6%, were all patients with CAVCD. Most of the reoperations both before and after discharge were in patients with CAVCD as seen in the table.

Table 8. Numbers of AV canal defects and mortality, reoperations and pacemaker insertion, Presence of Down's syndrome, and comparison of the 3.

Characteristic	Complete N (%)	Intermediate, N (%)	Partial N (%)	P value*
Presence of syndromes				
Down's syndrome	37 (59.7)	7 (41.2)	1 (3.2)	
Noonan syndrome	1 (1.6)	0 (0)	0 (0)	
None	22 (35.5)	10 (58.8)	30 (96.8)	
Syndrome pulir malformation	1 (1.6)	0 (0)	0 (0)	0.001
Vacterl syndrome	1 (1.6)	0 (0)	0 (0)	
AV PM (Pacemaker)				
No	57 (91.9)	17 (100.0)	30 (96.8)	
Yes	5 (8.1)	0 (0.0)	1 (3.2)	0.602
Mortality				
No	58 (93.5)	17 (100.0)	13 (100.0)	
Yes	4 (6.5)	0 (0.0)	0 (0.0)	0.361
Reoperation before discharge				
No	58 (93.6)	17 (100.0)	30 (96.8)	
Mitral valvuloplasty	2 (3.2)	0 (0.0)	1 (3.2)	
Repair of residual	2 (3.2)	0 (0.0)	0 (0.0)	0.914
Reoperation after discharge				
No	58 (93.6)	17 (100.0)	29 (93.6)	
PDA closure	1 (1.6)	0 (0.0)	0 (0.0)	
mv valvuloplasty	2 (3.2)	0 (0.0)	0 (0.0)	
MV and tricuspidplasty	1 (1.6)	0 (0.0)	1 (3.2)	
Removal of subaortic membrane and commissurotomy	0 (0.0)	0 (0.0)	1 (3.2)	0.679

*P values based on Fisher's exact tests.

Table 9. Comparison in age, weight at operation, length of hospital stay, Bypass (ECC) and cross clamp average for each type.

Characteristic	Complete N Median (IQR)	Intermediate, Median (IQR)	Partial Median (IQR)	P value*
Cross clamp time	98 (85 - 98)	65 (51 - 65)	60 (43 - 60)	<0.001

Continued

ECC time	139 (123 - 139)	96 (76 - 96)	84 (75 - 84)	<0.001
Weight at operation	5 (4.3 - 5)	8 (5.9 - 8)	15.5 (20 - 15.5)	<0.0001
Age	5 (3 - 5)	6 (5 - 6)	6 (4 - 6)	0.161
Length of hospital stay	16 (12 - 16)	9 (8 - 9)	10 (8.5 - 10)	<0.001

*Based on the Mann-Whitney U test for non-normally distributed data in **Table 9**, patients undergoing repair for CAVCD had longer cross clamp and in turn bypass time compared to the other patients. Patients with CAVCD were operated earlier, average age of 5 months compared to the others, intermediate and partial both 6 months. The weight at operation also varied with CAVCD operated on when they are smaller (average of 5 kg) and partial AVCD with average weight of 15.5 kg.

Patients who operated on for CAVCD spent a longer time in hospital with an average stay of 16 days.

3.3. Mortality**Table 10.** Factors associated with mortality and reintervention bivariate.

Characteristic	Mortality, No N (%) n = 106	Mortality, Yes N (%), N = 4	OR (95 %CI)	P value
Age in months				
<6	51 (48.1)	3 (75.0)	Reference	
6 - 12	55 (51.9)	1 (25.0)	0.31 (0.04 - 3.71)	0.316
Sex				
Female	56 (52.8)	3 (75.0)	Reference	
Male	50 (47.2)	1 (25.0)	0.37 (0.03 - 3.07)	0.400
Previous palliation				
No	98 (92.5)	2 (50.0)	Reference	
Yes	8 (7.5)	2 (50.0)	12.25 (1.52 - 98.87)	0.019
Cardioplegia				
Custodioid	92 (86.8)	4 (100)	Reference	
Del Nido	14 (13.2)	0 (0)	Cannot be calculated	
Associated syndromes				
No	59 (55.7)	3 (75.0)	Reference	
Yes	47 (44.3)	1 (25.0)	0.42 (0.042 - 4.15)	0.457
Associated surgery				
None	70 (66.0)	1 (25.0)	Reference	
Yes	36 (34.0)	3 (75.0)	5.83 (0.585 - 58.100)	0.133
Associated malformations				
None	68 (64.2)	0 (0.0)		

Continued

PDA	13 (12.2)	1 (25.0)		
TOF	3 (2.8)	2 (50.0)		
Other malformations	22 (20.8)	1 (25.0)	Can not be calculated	
ECC			1.02 (1.01 - 1.04)	0.002
Cross clamp			1.04 (1.01 - 1.07)	0.003
Weight in Kg				
Minimum-3.9	8 (7.6)	2 (50.0)	Reference	
4.0 and above	97 (92.4)	2 (50.0)	0.08 (0.01 - 0.67)	0.019

In **Table 10** above, Low weight (less than 3.9), previous palliation, Extra corporeal circulation (ECC), cross clamp time and associated surgeries seem to be the factors that are associated closely with mortality.

Therefore, the variables considered for multivariate are ECC, cross-clamp, weight, associated surgeries and previous palliation based on P values less than 0.20

Multivariate model

Table 11. Multivariate model looking at weight and ECC.

Characteristic	Adjusted Odds Ratio (95% Confidence interval)	P-value
Weight in kg		
Min-3.9	Reference	
4.0-max	0.008 (0.000 - 0.647)	0.031
ECC	1.035 (1.008 - 1.064)	0.010

We went ahead and analysed the data using a multivariate model that showed ECC to be significant under the multivariate model in **Table 11**.

3.4. Re-Intervention/Re-Operation

Table 12. Factors associated with re-operation/reintervention.

Characteristic	Re-intervention, N (%), No	Re-intervention, N (%), Yes	Unadjusted odds ratios (95% CI)	P-value
Age				
<6 mths	53 (56.9)	1 (20.0)	Reference	
>6 mths	52 (49.5)	4 (80.0)	4.08 (0.44 - 37.71)	0.216
Sex				
Female	58 (55.2)	1 (20.0)	Reference	

Continued

Male	47 (44.8)	4 (80.0)	4.97 (0.56 - 58.41)	0.143
Previous palliation				
No	95 (90.5)	5 (100.0)	Reference	
Yes	10 (9.5)	0 (0.0)	Cannot be calculated	
Associated surgeries				
No	69 (65.7)	2 (40.0)	Reference	
Yes	36 (34.3)	3 (60.0)	2.88 (0.46 - 17.99)	0.259
Associated syndromes				
No	60 (57.1)	2 (40.0)	Reference	
Yes	45 (42.9)	3 (60.0)	2.0 (0.32 - 12.47)	0.458
Cardioplegia				
Custodioid	93 (88.6)	3 (60.0)	Reference	
Del Nido	12 (11.4)	2 (40.0)	5.16 (0.78 - 34.11)	0.088
Associated malformations				
None	66 (62.9)	2 (40.0)	Reference	
Yes	39 (37.1)	3 (60.0)	2.54 (0.41 - 15.86)	0.319
ECC			1.01 (1.00 - 1.03)	0.018
Cross clamp			1.02 (0.99 - 1.03)	0.128
Weight in Kg				
Minimum-3.9	10 (9.6)	0 (00.0)	Reference	
4.0 and above	94 (90.4)	5 (100.0)	Cannot be calculated	

Using the bivariate analysis, re-operation was analyzed as above in **Table 12** and cardioplegia and ECC were significant.

For the multivariate analysis, the variables considered included cardioplegia and ECC in **Table 13**.

Table 13. Multivariate analysis of cardioplegia and ECC in association with re-operation.

Characteristic	Adjusted Odds Ratio (95% Confidence intervals)	P-value
Cardioplegia		
Custodioid	Reference	
del Nido	11.99 (1.233 - 116.609)	0.032
ECC	1.02 (1.005 - 1.033)	0.008

4. Discussion

The aim of this study was to retrospectively assess the outcome of definitive surgery for AVCD for patients at our hospital. The study looked at patient records of

110 patients who were operated on over a period of 20 years. These were patients with congenital AVCD including adults. AVCD accounts for about 3% - 7% of cardiac malformations and can occur in isolation or in patients with syndromes or other associated cardiac malformations [2] [4] [5]. AVCD are classified into 3 categories that is CAVCD, intermediate AVCD and partial AVCD. Complete AVCD includes ostium primum atrial septal defect, a common atrioventricular valve and a confluent posterior ventricular septal defect located in the inlet portion of ventricular septum. Partial AVCD is characterized by ostium primum septal defect and two distinct orifices of the atrioventricular valves with cleft of the antero-medial leaflet of the mitral valve. The intermediate AVCD has a restrictive ventricular septal defect associated with anatomical characteristics of partial AVCD [4] [40].

In this study, 62 (56.4%) patients had CAVCD, 31 (28.2%) had partial AVCD and 17 (15.4%) had intermediate AVCD (**Table 7**). This is a bit different from other studies that had partial AVCD as the majority [41]. This could be because our Centre is a tertiary cardiac Centre that receives most of the complicated cases in the region.

Definitive repair for this malformation, especially the CAVCD is currently carried out early in life, usually between 3 and 6 months but can also be carried out safely below 3 months of age if child presents early with symptoms [33] [42]. The surgery of patients with partial and intermediate AVCD can be safely delayed depending on presentation of patient. In our study the biggest number (37.3%) of patients were operated on also between the age of 3 - 6 months and we also had 13 (11.8%) patients operated on earlier before 3 months of age. Most of these operated on before 6 months were CAVCD (**Table 2**).

The choice of operative procedure for the CAVCD was the double patch technique which is widely used as seen by other authors [26] [39]. The partial AVCD were repaired by patch closure of the ASD and repair of cleft while the intermediate AVCD were repaired with ASD patch repair, LAVV cleft repair and the restrictive VSD was mostly repaired directly and sometimes with a patch as well (**Table 1**).

There were slightly more female patients (female: male ratio of 1.2:1) as seen in other studies [2]. Down syndrome was highly associated with the diagnosis of CAVCD in our cohort with about 59% of the children confirmed for Down syndrome which is comparable to other studies [5] [43]. 42 (38%) of the children had associated malformations including TOF, additional ASDs, aortic coarctation and hypoplastic aortic arch, PDAs and additional VSDs (**Table 1**). Eight of these patients had to undergo palliative procedures before the definitive surgery, 2 had pulmonary systemic shunts, 4 had pulmonary artery banding, 1 had PDA ligation and one had COA and PDA ligation (**Figure 2**). Overall palliative procedures have reduced, especially for CAVCD and corrective surgery is preferred even at our Centre [44]. There were five patients with associated TOF that were corrected as well at the definitive surgery. The median ECC time was 124 minutes and median cross-clamp time was 85 minutes at surgery (**Table 2**).

Out of the 110 patients 4 died before discharge with an in-hospital mortality rate of 3.6% (**Table 3**). All these patients had CAVCD. This is comparable to other studies [45] [46]. Down syndrome was not seen as a risk factor to surgery as in other studies [47]. In our study low weight (less than 3.9), previous palliation, Extra corporeal circulation, cross clamp time and associated surgeries seem to be the factors that are closely associated with mortality (**Table 10**). With multivariate analysis, ECC time is seen to be a major association to mortality (**Table 11**).

There were 5 (4.5%) reoperations before discharge, 3 for severe LAVVR, 1 for residual VSD and 1 for both residual VSD and RAVVR. Of the 5, all four were diagnosed with CAVCD and only one had Partial AVCD (**Table 4**). The patients with LAVVR included a 2 yr old child with Down syndrome who was re-operated on day 5 and had mitral valvuloplasty with edge-to-edge plasty, a 6-month-old child who had undergone modified single patch repair and reoperated on day 22 post op and also repair was by edge-to-edge stitch of the residual cleft. Both patients had mild regurgitation of the LAVV pre-operatively. The 3rd child who had a mitral valvuloplasty was re-operated on day 2 due to severe MR, was also noted to have severe LAVRR pre-op. The child was 8 months at time of operation and was the only one with diagnosis of partial AV canal. The 4th child, 6 months of age, was reoperated on day 30 post op due to a residual VSD and severe RAVVR. The residual VSD was closed with pledgeted prolene stitches and the valve repaired by closure of the cleft on the right side. The right components of the atrio-ventricular valve had mild regurgitation preop. The 5th child, 3 months of age, had CAVCD with associated TOF and was reoperated on day 4 for a residual VSD. All went on to be discharged successfully.

Five patients were readmitted for reoperation (**Table 4**) with 3 for LAVVR, and 1 for PDA closure. Of the 2 patients with LAVVR, one was reoperated on after 11 years due to severe LAVVR and moderate RAVVR and had mitral valve replacement with a mechanical valve, Carbomedics optiform size 29 mm and tricuspid plasty, the second was reoperated after 13 years and had a mitral valvuloplasty with an annular ring of size 32 mm. The third patient was reoperated after 8 years due to a small cleft and perforation in the anterior leaflet. All the 3 patients had moderate regurgitation of LAVV before the initial surgery. The 4th patient was found to have a large PDA causing symptoms after 7 years and underwent percutaneous closure of the defect. The last patient was re-operated for aortic valve disease which was causing symptoms with removal of a subaortic membrane.

The commonest re-operation cause also seen in other studies is LAVVR but has also gone down over the years. The results at our Centre resonate with other Centres as well in this regard [44] [46] [48]. For re-operation, ECC time is seen as a bigger association both at bivariate and multivariate levels in our study (**Table 12** & **Table 13**).

Five (8.1%) patients had to have permanent pacemakers inserted due to AV block post-surgery and all these had CAVCD (**Table 5**). AV block is one of the outcomes seen in other studies as well [41] [49].

Overall, there was less morbidity in the partial AVCD compared to the complete AVCD as compared to other studies [41] (Table 8). Patients undergoing repair for CAVCD had longer cross-clamps and in turn bypass time compared to the other patients (Table 9).

The average hospital stay was 18 days (interquartile range: 12 - 25) (Table 6). Also similar to other studies [49].

5. Conclusion

From our study, Atrioventricular septal defects, especially the CAVCD were safely corrected between 3 - 6 months with less morbidity. The other types were safely operated on later after 1st year of life, especially the partial type. The commonest cause of redo surgery is the regurgitation of LAVV component, especially in patients with CAVCD. AV block occurs more in patients with CAVCD and subsequent permanent pacemaker insertion. Overall, partial AVCD and intermediate AVCD have less morbidity compared to CAVCD with less mortality, shorter hospital stays and complications of AV blocks.

Conflicts of Interest

The authors declare no conflicts of interest.

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