Peri-operative outcomes of patients undergoing Atrio-Ventricular Septal Defect repair at a new tertiary/quaternary children's hospital in Johannesburg

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Declaration

I, Marwa Mahdi, student number 0703071k, declare that
this is my original work, submitted for the Master of Medicine in Paediatrics degree for the University of the Witwatersrand. It has not been submitted to any other university or for any other degree or examination. It has not yet been submitted for publication in any journal or presented at any conferences. It is written according to the South African Journal of Child Health guidelines as stipulated below.

Preparation notes by article type

- ***Research***

  Guideline word limit: 3 000 words (excluding abstract and bibliography)
  Research articles describe the background, methods, results and conclusions of an original research study. The article should contain the following sections: introduction, methods, results, discussion and conclusion, and should include a structured abstract (see below). The introduction should be concise – no more than three paragraphs – on the background to the research question, and must include references to other relevant published studies that clearly lay out the rationale for conducting the study. Some common reasons for conducting a study are: to fill a gap in the literature, a logical extension of previous work, or to answer an important clinical question. If other papers related to the same study have been published previously, please make sure to refer to them specifically. Describe the study methods in as much detail as possible so that others would be able to replicate the study should they need to. Where appropriate, sample size calculations should be included to demonstrate that the study is not underpowered. Results should describe the study sample as well as the findings from the study itself, but all interpretation of findings must be kept in the discussion section, which should consider primary outcomes first before any secondary or tertiary findings or post–hoc analyses. The conclusion should briefly summarise the main message of the paper and provide recommendations for further study.

  May include up to 6 illustrations or tables.

  A max of 20 – 25 references

  Structured abstract

  - This should be no more than 250 words, with the following recommended headings:

    - **Background:** why the study is being done and how it relates to other published work.
    - **Objectives:** what the study intends to find out
    - **Methods:** must include study design, number of participants, description of the intervention, primary and secondary outcomes, any specific analyses that were done on the data.
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    - **Conclusion:** must be supported by the data, include recommendations for further study/actions.

    - Please ensure that the structured abstract is complete, accurate and clear and has been approved by all authors. It should be able to be intelligible to the reader without referral to the main body of the article.

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  Here is an example of a good abstract.

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Abstract

Background: Atrioventricular septal defect (AVSD) is a common cardiac malformation, representing about 7% of congenital heart diseases and frequently associated with Down syndrome (DS). There is limited South African literature on the perioperative management of this condition.

Aim and objectives: This study aimed to identify perioperative factors affecting AVSD repair outcomes, assess patient outcomes, and evaluate repair success at a new center in a low/middle-income country.

Methods: This was a retrospective, descriptive record review that included 41 patients with a confirmed diagnosis of AVSD, who underwent corrective surgery over a four-year period.

Results: Complete AVSD (CAVSD) was found in 26/41 (63.4%) patients, primum atrial septal defect in 11/41 (26.8%), and inlet ventricular septal defect in 4/41 (9.8%). The median age at diagnosis was five months (IQR 2-13), and at repair was 22 months (IQR 10-30). The overall mortality rate was 7.3%. Patients with CAVSD had longer cardiopulmonary bypass times (median 155 mins, IQR 126-223), cross-clamp times (median 118 mins, IQR 89-174), and ICU stays (median 7 days, IQR 4-12), compared to those with a partial AVSD. Patients with DS had longer ICU stays (median 8.5 days, IQR 4-14) and ventilation times (median 5 days, IQR 2-9.5), compared to those without DS. Post-operatively, 12/41 (29%) patients experienced a pulmonary hypertensive crisis**.**

Conclusion: This study found that CAVSD was the most prevalent type of AVSD and was frequently associated with DS. Operative repair was performed relatively late, particularly in cases of CAVSD. Although mortality rates were similar to other studies, post-operative morbidity was higher, especially in CAVSD and DS. These findings highlight the need for earlier AVSD diagnosis and repair to reduce complications and improve long-term outcomes in these patients**.**

Keywords: Atrioventricular septal defect, Down Syndrome, Congenital heart disease, Pulmonary hypertension

Abbreviations

ASD	Atrial septal defect
AVSD	Atrio-ventricular septal defect
CAVSD	Complete atrioventricular septal defect
CCT	Cross clamp time
CHD	Congenital heart disease
CPB	Cardiopulmonary bypass
DORA	Double outlet right atrium
DS	Down Syndrome
ECMO	Extracorporeal membrane oxygenation
ICU	Intensive care unit
IQR	Interquartile Range
LAVV	Left atrio-ventricular valve
LAVVR	Left atrio-ventricular valve regurgitation
LMIC	Low and middle income countries
LOS	Length of stay
PAH	Pulmonary arterial hypertension

PDA	Patent ductus arteriosus
PHT	Pulmonary hypertensive crisis
RAVVR	Right atrio-ventricular valve regurgitation
TOF	Tetralogy of Fallot
TR	Tricuspid Regurgitation
VSD	Ventricular septal defect

Introduction

Atrioventricular septal defects (AVSD) are a wide spectrum of acyanotic congenital heart disorders constituting about 7% of all congenital heart disease (CHD) with an incidence of 4-5.3 per 10000 live births.[1] Approximately 50% of children with Down syndrome (DS) have CHD, and 50% of those have AVSD. Conversely, up to 50% of patients with AVSD have DS.[1] Moreover, 50% of AVSD patients also have a patent ductus arteriosus (PDA), making pre-surgical PDA identification critical.[2] The International Paediatric and Congenital Cardiac Code classifies AVSDs into four main groups: complete (CAVSD), partial (or incomplete), intermediate (or transitional), and AVSDs with ventricular imbalance (unbalanced AVSD).[1]

Outcomes of children undergoing surgical repair of AVSD around the globe have improved dramatically over the past decade. This is due to refinements of the surgical technique, improved pre- and post-operative management and timing of surgical intervention - with the repair carried out within the first six months of life in most centres, and within the first year of life in the majority of cases.[3] Prematurity, low birth weight, pulmonary hypertension, complex AVSD, Atrio-Ventricular valve regurgitation and heart block were all factors associated with mortality in other studies.[4] Interestingly, studies conducted in developed countries have found that DS is not a risk factor for increased reoperation rates or increased mortality when compared to patients without DS. Therefore, it is recommended that the repair of CAVSD should be performed before six months of age to avoid complications related to increased left to right shunt, while partial AVSD surgery is recommended between ages two and five.[3] The main cause for morbidity at presentation is signs of heart failure and high pulmonary vascular resistance.[3] In studies done in other developing nations (Uganda and Ethiopia), arrhythmia was identified as the commonest post-operative complication.[5-6]

Due to the paucity of data in Low and Middle Income Countries (LMIC), we aimed to evaluate the age at time of diagnosis and surgery, type of AVSD being repaired, surgical technique used, complications in the peri-operative and post-operative period and how all these factors affect the management surrounding AVSD repair.

Materials and Methods

This retrospective, descriptive study reviewed records of 41 patients who underwent AVSD repair at Nelson Mandela Children's Hospital (NMCH) from July 1, 2018, to June 30, 2022. We included all 41 patients who underwent surgery at NMCH who had a confirmed diagnosis of AVSD. NMCH, a tertiary/quaternary referral hospital in Johannesburg, South Africa, provides specialized cardiac care to pediatric patients from Johannesburg, Gauteng, and neighboring provinces in a LMIC.

Patients with total anomalous pulmonary venous connection, double outlet right ventricle, single ventricle conditions, or those undergoing palliative procedures like pulmonary artery banding were under the exclusion criteria.

The objectives were to evaluate demographics, clinical factors and timing of surgery in patients that had AVSD repair, focusing on perioperative factors and complications, post-operative echocardiography and other factors that would contribute to improved outcomes.

Data Collection

The patient characteristics collected included age at diagnosis and age at the time of surgery, as well as anthropometric data, gender, race/ethnicity, type of atrioventricular septal defect and any associated noncardiac congenital anomalies and syndromes. Preoperative data were gathered, including oxygen saturation levels, cardiac catheterization reports detailing the presence of a shunt, pulmonary vascular resistance, and its reversibility. Additionally, preoperative echocardiograms were reviewed to assess the extent of valvular involvement, tricuspid regurgitation gradient, and the size of the defect.

Procedural factors included cardiopulmonary bypass (CPB) and cross-clamp times (CCT), any concurrent procedures, operative findings, perfusion records, the drainage site of the coronary sinus, the type of surgical repair performed, and whether a patent foramen ovale was preserved.

Postoperatively, data from Intensive Care Unit (ICU) discharge records were used to assess ventilation duration, length of ICU stay, number of re-intubations, and various complications such as respiratory, hemodynamic, renal, neurological, and gastrointestinal issues. The presence and severity of postoperative valvular dysfunction were also evaluated, along with the need for extracorporeal membrane oxygenation (ECMO) and the rate of in-hospital mortality.

Statistical Analysis

Descriptive statistics were presented for the various variables mentioned above. Median values along with the interquartile ranges were given. To compare between different outcome variable groups, the Mann-Whitney U test was used. A p-value of <0.05 was considered statistically significant. All analyses was done in Statistica version 14.0.0.15.

Ethics Approval

This study was approved by the WITS Human Research Ethics Committee (no M220963).

Results

A total of 41 patients fitting the study criteria having undergone corrective AVSD repair over a period of four years were enrolled and there were no patients that needed to be excluded. As seen on Table 1, the male to female ratio was 1.16:1, with 22/41 (54%) patients being male. Many of the patients, comprising 39/41, were of Black ethnicity, while one patient was Caucasian, and another was of mixed-race ethnicity. Seventeen percent of the patients were wasted, whilst 7% were stunted and 54% were both stunted and wasted.

The median age at diagnosis was five months (IQR 2-13), median age at repair was 22 months (IQR 10-30) and the median weight at time of surgical repair was 9kg (IQR 6.5-15.5). The median waiting time for surgery was three months (IQR 1-11). Statistical analysis revealed significant differences in both age at diagnosis and age at surgery when comparing the CAVSD and ASD primum groups, with CAVSD patients being diagnosed earlier and undergoing surgery at a younger age than those with ASD primum. This corresponds to the

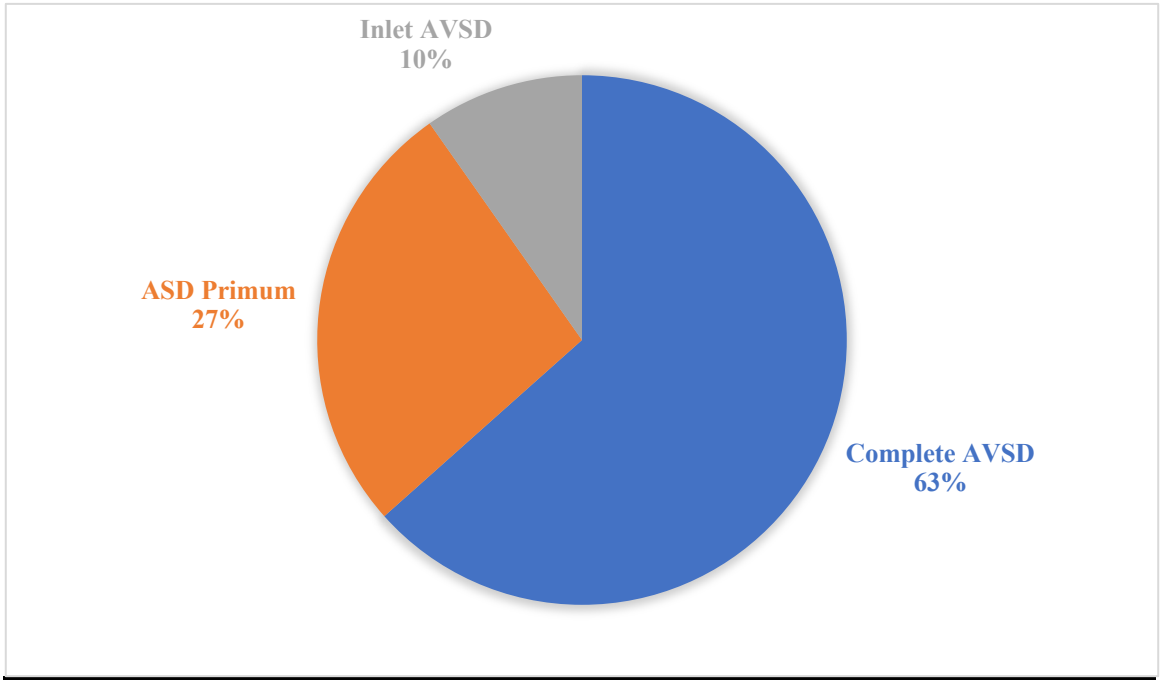
lower weight and height measurements when comparing the CAVSD group and ASD primum.

Most patients had DS (78%). One patient had Noonan syndrome with an associated ASD primum.

Table 1: Basic demographics for the combined cohort study

		Total	
		n	MEDIAN / % (IQR)
Demographics			
Age at diagnosis	(months)	41	5 (2-13)
Weight	(kg)	41	9 (6.5-15.5)
Height	(cm)	41	80 (70-100)
Age at surgery	(months)	41	22 (10-30)
Waiting period from presentation to surgery	(months)	41	3 (1-11)
Down Syndrome	Yes	32	78.05%
Gender	Male	22	53.65%
	Female	19	46.34%
Anthropometry	Normal	9	21.95%
	Wasted	7	17.07%
	Stunted	3	7.32%
	Wasted and Stunted	22	53.66%

Figure 1: Distribution of AVSD Types



As illustrated on Figure 1, among the types of AVSD, 26/41 (63.4%) cases were classified as CAVSD, 11/41 (26.8%) as ASD primum, 4/41 (9.8%) as inlet ventricular septal defect (VSD). All CAVSD's fit the type A Rastelli classification. Twenty-six (63.4%) patients had an associated cardiac abnormality.

Of the 41 patients, 32/41 (78%) had DS, while 9/41 (22%) were non-DS. Among the 32 DS patients, 16/32 (50%) had an PDA. CAVSD was highly prevalent in the DS group, occurring in 24/32 (75%) of patients, while 4/32 (12.5%) had an associated ASD primum. Notably, all 16/16 (100%) patients with an associated PDA had DS.

As per Figure 2, PDA was the most common concomitant associated cardiac anomaly (39%) followed by ASD secundum (34.1%) then double outlet right atrium (DORA) (12.2%). One patient (2.4%) had an associated Tetralogy of Fallot (TOF).

Most patients exhibited preoperative left atrio-ventricular valve regurgitation (LAVVR) and right atrio-ventricular valve regurgitation (RAVVR). Specifically, in the LAVVR group, 39% had mild, 39% had moderate, and 12% had severe LAVVR. In the RAVVR group, 24% had mild, 46% had moderate, and 12% had severe RAVVR.

Figure 2: Presence of associated cardiac lesions

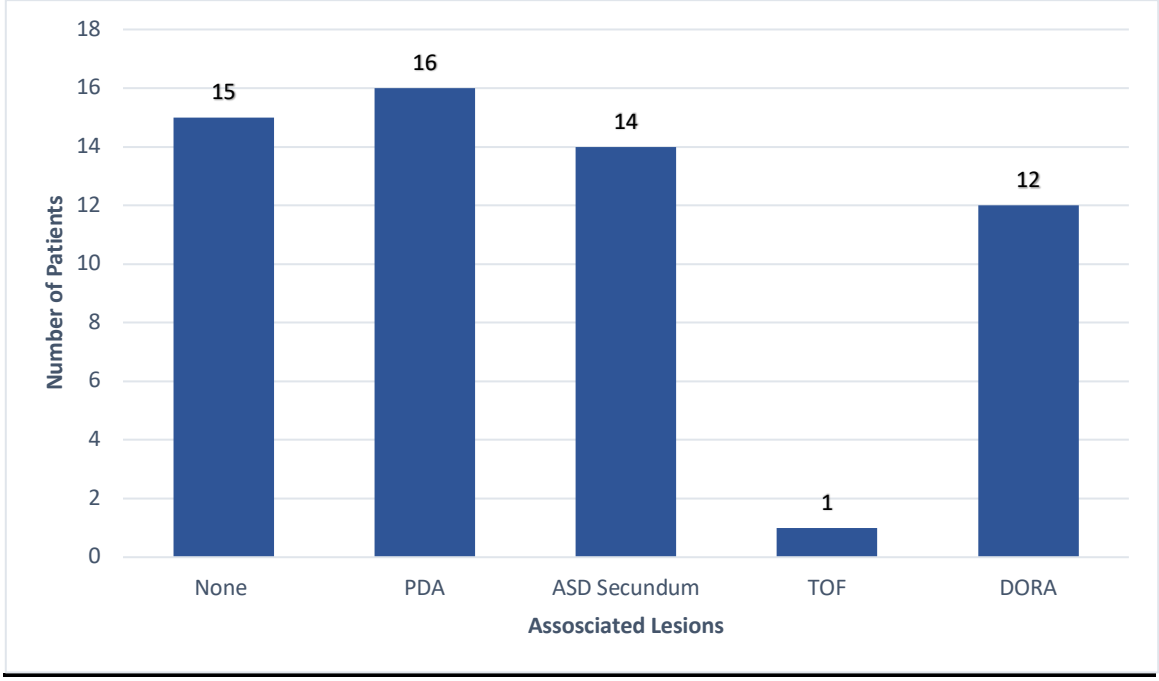


Table 2 indicates significant statistical differences in perioperative outcomes, revealing that CCT, CPB time, ICU length of stay (LOS), and duration of ventilation were all longer for CAVSD repair procedures compared to repairs for ASD primum and inlet VSD.

There were no noteworthy significant differences observed in postoperative biochemistry results, requirement for fluid boluses, or use of inotropes across all the various groups.

Table 2: Peri-operative factors for the combined cohort study by AVSD Type

		Total		Complete AVSD Group		ASD Primum and Inlet VSD Group		
		N	MEDIAN (IQR)	N	MEDIAN (IQR)	N	MEDIAN (IQR)	2 sided P- Value
Perioperative Findings								
Cross clamp time	(mins)	41	118 (89-174)	26	165.5 (118-192)	15	89 (81-94)	0.000025
Cardiopulmonary bypass time	(mins)	41	155 (126-223)	26	210.5 (151-238)	15	123 (113-146))	0.000083
Ventilation length	(days)	41	3 (1-8)	26	5.5 (2-10)	15	1 (1-2)	0.000274
ICU length	(days)	41	7 (4-12)	26	10 (4-14)	15	4 (3-7)	0.003132
Urea	mmol/L	41	4 (2.9-5.7)	26	4.3 (3.1-5.9)	15	3.4 (2.7-5.7)	0.494582
Creatinine	umol/L	41	33 (26-40)	26	33 (26-41)	15	33 (23-35)	0.398332
Albumin	g/L	41	32 (28-35)	26	33.5 (29-37)	15	30 (23-33)	0.107904

Table 3 shows no significant differences in age at AVSD diagnosis between patients with and without DS. However, those with DS had surgery at a median age of 19 months (IQR 9-57.7), whereas those without DS had surgery at a median age of 66 months (IQR 22-92). Notably, only four patients without DS were in the ASD Primum group.

In accordance with the data presented in Table 3, when comparing patients diagnosed with AVSD, significant statistical disparities were found between those with and without DS. Patients without DS had larger ASD sizes, while those with DS experienced considerably

longer postoperative ventilation duration and ICU stays. In patients with DS, there was a longer length of ICU stay of >7days in the CAVSD group, when compared to the ASD primum group.

ASD patch closure was the most frequent procedure performed in patients without DS. This observation aligns with the higher prevalence of ASD primum in non-DS patients, whereas CAVSD was more prevalent among those with DS. All patients with an ASD primum had ASD primum patch closure.

Of the 26 patients diagnosed with CAVSD, none underwent single patch repair. Thirteen received double patch technique, while 12 had modified single patch repair, indicating no distinct preference between the two methods. Additionally, one patient underwent VSD patch closure along with TOF repair. All four patients with an inlet VSD underwent a VSD patch closure and all patients with an ASD primum underwent an ASD primum patch closure.

Table 3: Patient factors for the combined cohort study for those with and without Down Syndrome

		Non-Down Syndrome Group		Down Syndrome Group		2 sided P-Value
		N	Median (IQR)	N	Median (IQR)	
Demographics						
Age at surgery	(months)	9	66 (22-92)	32	19 (9-57.5)	0.024
Age at diagnosis	(months)	9	8 (5-84)	32	5 (1.5-12)	0.133
Weight	(kg)	9	15 (8.3-17.2)	32	8 (6.5-14.25)	0.085
Height	(cm)	9	100 (75-115)	32	78.5 (69.5-99.5)	0.097
Waiting period from presentation to surgery	(months)	9	1 (0.75-11)	32	3 (1.5-11)	0.289

Echo findings						
RAVVR gradient	(mmhg)	3	36 (33-43)	29	50 (40-60)	0.136
RAVVR gradient +5	(mmhg)	3	41 (38-48)	29	55 (45-65)	0.136
Total size of defect	(mm)	2	17.5 (11-24)	23	18 (15-25)	0.650
Inlet VSD size	(mm)	3	4 (3-11)	28	10.5 (7.5-11.5)	0.120
ASD primum size	(mm)	8	14 (10-19)	27	8.5 (5-11)	0.017
Peri-operative						
Cross clamp time	(mins)	9	92 (81-100)	32	133 (90.5-182.5)	0.138
Cardiopulmonary bypass time	(mins)	9	119 (116-146)	32	175 (136-229.5)	0.075
Ventilation length	(days)	9	1	32	5 (2-9.5)	0.0003
ICU length	(days)	9	4 (3-5)	32	8.5 (4-14)	0.049
Urea	mmol/L	9	4.4 (2.9-4.6)	32	3.95 (2.95-5.8)	0.974
Creatinine	umol/L	9	28 (23-43)	32	33.5 (27.5-40)	0.403
Albumin	g/L	9	32 (29-33)	32	31.5 (28-36)	0.837

Figure 3 represents some of the most common post-operative complications experienced in our cohort. The most common finding was that most of the patients (98%) required the use of inotropes postoperatively. Regarding post-op LAVVR, there were no patients that needed early re-repair, although most patients did have a degree of post-op RAVVR. Out of the 41 patients, 10/41 (24.4%) showed no signs of postoperative LAVVR, 13/41 (31.7%) developed mild LAVVR, and 15/41 (36.6%) developed moderate LAVVR.

Twelve (29%) patients out of the total 41 had a pulmonary hypertensive crisis. According to the data, patients who underwent CAVSD repairs exhibited a higher incidence of pulmonary

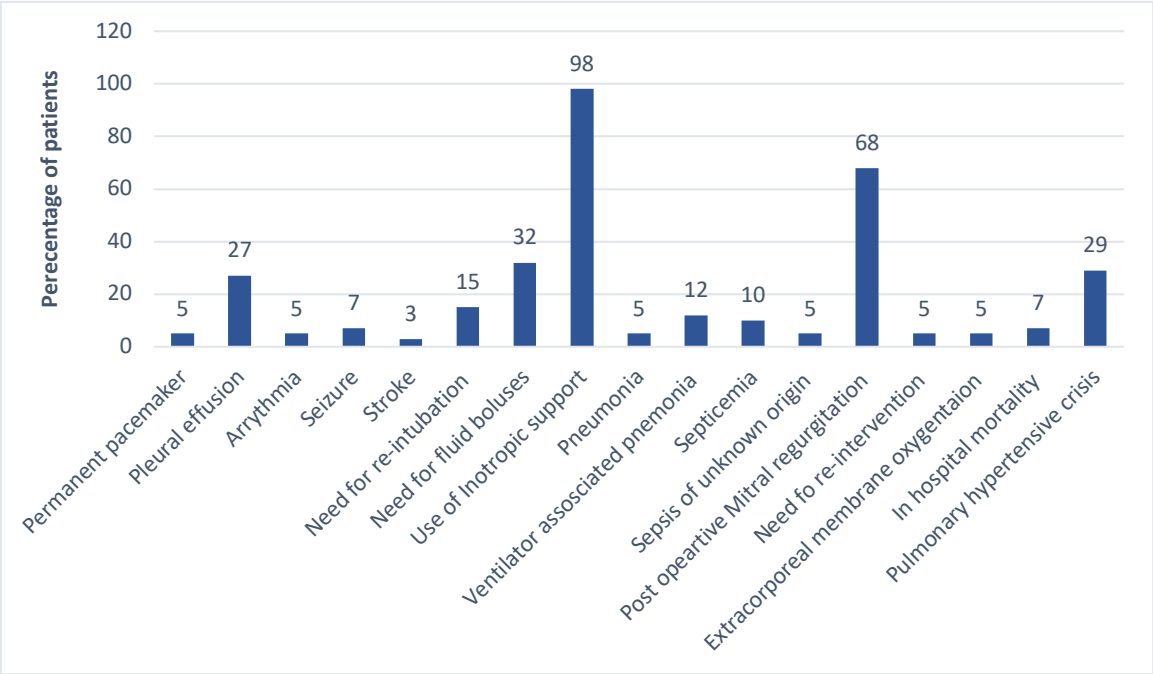
hypertensive crises and a longer length of ICU stay of >7 days. Three patients experienced seizures and only one patient had a stroke with complete neurological recovery prior to discharge. The sepsis rate was low with only two patients developing pneumonia, five patients developing a ventilator associated pneumonia, four patients developing septicemia and two patients had sepsis where the source could not be identified. Two patients were severely unstable post-op requiring ECMO, one of those patients demised.

The analysis of other complications, such as the necessity for re-intervention, re-intubations, or postoperative (ECMO), among other factors delineated in Table 3, did not reveal any distinct significance, between the CAVSD and ASD primum groups, nor between the DS and non-DS groups.

Median ICU length of stay (LOS) was 7 days (IQR 4-12) and median ventilation duration was three days (IQR 1-8). There were only two patients who experienced arrhythmias post-operatively, with only one of them needing a pacemaker for complete atrioventricular block.

Three deaths were reported, constituting 7.3% of the cases. Among them, two deaths were attributed to cardiogenic causes, namely severe pulmonary hypertensive crisis with residual severe post-operative LAVVR, and intractable arrhythmias, respectively. The cause of death for the remaining case remained unknown.

Figure 3: Post-operative Complications



Discussion

This study found CAVSD to be the most common type of AVSD in DS patients, with repairs done at an older age compared to other studies, especially in CAVSD. Mortality rates were similar to other studies, but post-operative morbidity, particularly in the CAVSD group, was higher. While DS doesn't affect early mortality, it increases the risk of complications like prolonged ventilation and longer ICU stays.

More than half of the patients in this cohort were both wasted and stunted at the time of the surgery, a finding similar to that in other published studies.[7] This could be attributed to the observed complications of recurrent illness, congestive cardiac failure and pulmonary infections that are associated with AVSD.[8] Median weight at surgery was 9kg which is higher than that reported in literature, since our patients were also operated at an older age.[3-4,9-10]

DS is commonly associated with AVSD, a correlation supported by our findings, wherein 32/41 (78%) AVSD patients also had DS, similar to that found in current literature.[3-4,7,11] The prevalence of CAVSD was 63.4% which is slightly less than in other published articles [4,7], but similar to a finding found by Formigari.[9] CAVSD was more common in DS,

comprising 75% of the DS group, whilst partial AVSD was more common in non-DS group, a fact similar to that in other published literature.[3,7]

The one patient who had another associated genetic syndrome; was diagnosed with Noonan's syndrome and a partial AVSD. This corresponds with the findings of the study conducted by Marino Bruno *et al.*, which indicated an increased risk associated with Noonan syndrome for partial AVSD.[7] The most common cardiac associated conditions were found to be PDA and ASD secundum and this was consistent with other studies.[7,10]

There was no significant difference in age at diagnosis between DS and non-DS groups, consistent with findings from another study conducted in Romania.[7] Despite guidelines for routine cardiac screening in newborns with DS, our study found a median diagnosis age of five months, later than in other studies.[7] This delay highlights deficiencies in prenatal and postnatal screening, suggesting a need for improved screening services for all patients, particularly those with DS.

Our patients had their repair at an average age of 22 months, despite a median diagnosis age of five months. In LMICs with limited resources and long surgical waiting lists, surgery cannot be performed immediately upon diagnosis. Patients typically undergo cardiac catheterization (especially if older than six months) before being added to a waiting list for surgery. This older age at repair aligns with findings from other studies on congenital heart disease and cardiac surgery in developing nations.[5,12-13] For instance, a study at the Instituto do Coração from April 2001 to December 2007 in which 196 patients were operated, reported a median age at diagnosis of four years and a mean age at surgery of eight years, which is much later than our results.[12] Similarly, a study in Addis Ababa, Ethiopia in which 76 patients were examined mean age at repair of seven years was found.[5]

For CAVSD, surgical correction is ideally performed before the age of six months.[4,7,9,14] However, our cohort was operated on at a median age of 14 months, which is significantly later than recommended. This could be due to the aforementioned reasons. The median age for partial AVSD repair was 19 months in DS patients and 66 months in non-DS patients, which is later than other studies. This delay may be due to later diagnosis and the lower risk of complications in partial AVSD cases.[7]

In children with unrepaired CAVSD, irreversible pulmonary vascular disease affects nearly all by age two.[8] Prompt correction can reduce this risk.[15] Sharma *et al.* found that 5/10 children with DS develop pulmonary arterial hypertension (PAH), with the risk rising to nearly 8/10 in those with DS and AVSD.[16] Our study supports this, as 12/32 DS patients experienced a pulmonary hypertensive crisis, compared to none in the non-DS group. The overall rate of developing pulmonary hypertensive crisis was 29% in our cohort which is significantly higher than that published in other studies.[17-18] This may explain why DS patients in our cohort had a longer ICU and ventilation stays, with a median ventilation length of five days for DS patients compared to one day for non-DS patients. These results are consistent with other published studies.[13,18]

When looking at the CAVSD group, the median ICU LOS was 10 days and median ventilation length was four days, this is longer than that reported in another study.[10] In our cohort, patients with CAVSD had longer CPB times, CCT, ICU stays, and ventilation durations. This differs slightly from the study by Carlos M *et al.*, which reported significant differences in CCT and CPB times between CAVSD and partial AVSD, but no notable difference in ICU length of stay.[19] Our findings match those of Ozlem S *et al.*, a study looking at long term outcomes of AVSD repair that looked at 314 patients with CAVSD and 181 with partial AVSD. Both studies found that CAVSD patients had longer CCT, CPB times, and ICU stays compared to partial AVSD patients.[13] There is currently a paucity in literature directly comparing outcomes between complete and partial AVSD regarding the aforementioned four parameters. Many studies have rather investigated complete AVSD and partial AVSD separately, with some focusing on comparisons based on age at repair or surgical techniques.[11,20-22] The overall median CPB time was slightly higher than in other studies and the median ICU stay of seven days aligns with current literature.[3,7,11]

Most of our patients needed post-operative inotropic support, consistent with the department's goal to discontinue it within 48 hours. Only one patient required a pacemaker, similar to the findings from a Romanian study and other supporting literature.[3,7,19]

A study by Atz AM *et al*. that looked at 120 patients in which 80% of the patients had DS, showed that there was no difference in surgical technique between AVSD in DS and non DS group.[11] Although this was not our finding as we had no patients that required re-operation,

multiple studies indicate that DS patients typically require reoperation for LAVVR later than those without DS.[4,23] Whilst other studies suggest that the complication risk after AVSD surgical repair in DS patients is generally not elevated, except for the risk of PAH. In the study by Formigari, that looked at the impact of DS on outcomes of AVSD repair, it was confirmed that DS patients have a lower probability of re-intervention.[9] This finding is crucial as it emphasizes the importance of not withholding this treatment from patients with DS due to concerns about potential adverse outcomes. Long-term follow-up is essential as the need for reintervention may arise. [10,19]

The overall mortality was 7.3%, which is marginally higher than reported in other literature. [3,5-6,10-11,19] However, it was similar to the findings of a study published by Formigari *et al*., 2004.[9]

Study Limitations

The study's limitations include its single-center, retrospective design, small sample size, and short four-year period, which may impact accuracy. Larger studies are needed for better outcome assessment. Delayed diagnoses were due to healthcare challenges in South Africa, and social factors were not evaluated. Nonetheless, the findings are valuable for understanding AVSD in developing economies and could enhance cardiology referral rates.

Conclusion

The findings from this study emphasize the critical need for earlier surgical intervention in patients with AVSD, particularly LMIC where delayed presentation is common. Clinicians must be aware that patients with CAVSD and those with DS tend to have a more complicated postoperative course compared to those with partial AVSD or patients without DS. Clinicians should carefully manage these patients, anticipate complications, and prioritize early intervention to improve outcomes and reduce risks in these high-risk populations.

Conflicts of Interest

There were no competing interests.

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References

1. Calkoen EE, Hazekamp MG, Blom NA, et al. Atrioventricular septal defect: From embryonic development to long-term follow-up. Int J Cardiol. 2016; 202:784-95.

2. Szulc M, Ritter SB. Patent ductus arteriosus in an infant with atrioventricular septal defect and pulmonary hypertension: Diagnosis by transesophageal color flow echocardiography. J Am Soc Echocardiogr. 1991; 4(2):194-8.

3. Louis JDS, Jodhka U, Jacobs JP, et al. Contemporary outcomes of complete atrioventricular septal defect repair: Analysis of the society of thoracic surgeons congenital heart surgery database. J Thorac Cardiovasc Surg. 2014; 148(6):2526-31.

4. Schleiger A, Miera O, Peters B, et al. Long-term results after surgical repair of atrioventricular septal defect. Interact Cardiovasc Thorac Surg. 2019; 28(5):789-96.

5. Ejigu YM, Amare H. Pediatric cardiac surgery in ethiopia: A single center experience in a developing country. Ethiop J Health Sci. 2023; 33(1)

6. Aliku TO, Lubega S, Lwabi P, et al. Outcome of patients undergoing open heart surgery at the uganda heart institute, mulago hospital complex. Afr Health Sci. 2014; 14(4):946-53.

7. Olariu I-C, Popoiu A, Ardelean A-M, et al. Challenges in the surgical treatment of atrioventricular septal defect in children with and without down syndrome in romania-a developing country. Front Pediatr. 2021; 9:612644.

8. Calabrò R, Limongelli G. Complete atrioventricular canal. Orphanet J Rare Dis. 2006; 1:1-5.

9. Formigari R, Di Donato RM, Gargiulo G, et al. Better surgical prognosis for patients with complete atrioventricular septal defect and down's syndrome. Ann Thorac Surg. 2004; 78(2):666-72.

10. Xie O, Brizard CP, d'Udekem Y, et al. Outcomes of repair of complete atrioventricular septal defect in the current era. Eur J Cardiothorac Surg. 2014; 45(4):610-7.

11. Atz AM, Hawkins JA, Lu M, et al. Surgical management of complete atrioventricular septal defect: Associations with surgical technique, age, and trisomy 21. J Thorac Cardiovasc Surg. 2011; 141(6):1371-9.

12. Mocumbi AO, Lameira E, Yaksh A, et al. Challenges on the management of congenital heart disease in developing countries. Int J Cardiol. 2011; 148(3):285-8.

13. Sarısoy Ö, Ayabakan C, Tokel K, et al. Long-term outcomes in patients who underwent surgical correction for atrioventricular septal defect. Anatol J Cardiol. 2018; 20(4):229.

14. Al-Hay AA, MacNeill SJ, Yacoub M, Shore DF, Shinebourne EA. Complete atrioventricular septal defect, down syndrome, and surgical outcome: Risk factors. Ann Thorac Surg. 2003; 75(2):412-21.

15. Marder L, Tulloh R, Pascall E. Cardiac problems in down syndrome. Paediatr Child Health. 2015; 25(1):23-9.

16. Sharma M, Khera S, Sondhi V, Devgan A. A study to determine the prevalence of pulmonary arterial hypertension in children with down syndrome and congenital heart disease. Med J Armed Forces India. 2013; 69(3):241-5.

17. Cai Y, Chen R, Chen G, et al. Middle to long-term outcomes of surgical repair for atrioventricular septal defect: A single-center study. J Thorac Dis. 2022; 14(10):3706.

18. Tumanyan MR, Filaretova OV, Chechneva VV, et al. Repair of complete atrioventricular septal defect in infants with down syndrome: Outcomes and long-term results. Pediatr Cardiol. 2015; 36:71-5.

19. Mery CM, Zea-Vera R, Chacon-Portillo MA, et al. Contemporary outcomes after repair of isolated and complex complete atrioventricular septal defect. Ann Thorac Surg. 2018; 106(5):1429-37.

20. Sena A, Sheytanov V, Liebrich M, et al. Twelve years of complete atrioventricular septal defect repair. Port J Card Thorac Vasc Surg. 2019; 26(3):187-93.

21. Suzuki T, Bove EL, Devaney EJ, et al. Results of definitive repair of complete atrioventricular septal defect in neonates and infants. Ann Thorac Surg. 2008; 86(2):596-602.

22. Xuan NT, Hung NX, An TH, et al. Treatment of isolated complete atrioventricular septal defect: The hue central hospital experience. Open Access Surgery. 2020; 13:39-46.

23. El-Najdawi EK, Driscoll DJ, Puga FJ, et al. Operation for partial atrioventricular septal defect: A forty-year review. J Thorac Cardiovasc Surg. 2000; 119(5):880-90.