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Original Research Paper

Cardiology

ATRIOVENTRICULAR CANAL DEFECTS RETROSPECTIVE ANALYSIS OVER THE LAST TEN YEARS IN OUR INSTITUTE

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ABSTRACT

Heart, characterized as 'the youngest, most diverse, most fluid, most changeable, most versatile part of creation' is the first organ to form and start functioning in vertebrate embryos. Beginning about embryonic day 20 in the human foetus, progenitor cells within the lateral plate mesoderm become committed to a cardiogenic fate.[1]

Cardiac malformations constitute a major percentage of clinically significant birth defects, with an estimated prevalence of 4 to 50 per 1000 live births. An incidence of congenital cardiac defects of 50 per 1000 live births is a conservative estimate.[2] Atrioventricular (AV) canal defects occur in two out of every 10,000 live births accounting for approximately 3% of all major congenital cardiac defects.[3] Both sexes are equally affected, with a slight female preponderance (female/male ratio -1.3/1).3 A striking association with Down syndrome is noted with about 50- 75% of patients with complete AV canal defect and about 10% of patients with partial AV canal defect have associated Down syndrome. In children with Down syndrome, AV canal defects represent the most common congenital heart anomaly and are seen in 20-25% representing a 1000-fold higher rate when compared to the incidence in the general population.[3,4,5]

KEYWORDS :

INTRODUCTION

In the course of human embryonic development, formation of the AV valves and septa depends upon precise central fusion of the endocardial cushions with the septum primum and primitive interventricular septum. Failure of this process results in a broad spectrum of congenital cardiac anomalies.[3, 6] The most important and complex being complete AV canal defect characterised by a variable deficiency of the AV area (crux cordis), the malformation involving the atrial, ventricular and AV septae and both AV valves resulting in the AV valve leaflets being insecurely attached to the upper margin of the ventricular septum, and the leaflets having clefts, rendering them incompetent.[3, 6] Ostium primum atrial septal defect (OP ASD) represents the simplest form of the spectrum complex.[3,7,8]

Complete AV canal defects have been described and classified depending on the morphology of the common anterior leaflet and the degree of its 'bridging' over the ventricular septum and the pathophysiological consequence of these defects is severe left-to-right shunting. A 'partial' variant of AV canal defect also exists (also known as OP ASD).[3,4,7-9]

Complete AV canal defect is frequently associated with other congenital cardiac anomalies. A large percentage (about 80%) of patients with this defect have associated Down syndrome.[3,4,5]

The natural history of AV canal defect is ultimately fatal; 80% of children born with this condition die within 2 years.[3, 6, 7] Pulmonary vascular obstructive disease (PVOD) usually develops, within the first year of life.[3,4,7,9]

Symptoms of complete AV canal defect usually appear in infancy and include failure to thrive, recurrent respiratory infections and congestive heart failure.[3,4,5,7]

Treatment of AV canal defects requires either palliative or curative surgery. Long-term medical therapy is ineffective and largely restricted to control of congestive cardiac failure,essentially serving as a bridge to surgery.[3, 4-7] Palliation, achieved by pulmonary artery (PA) banding, reduces the degree of interventricular shunting and protects against development of PVOD.[4, 7, 10, 11] PA banding produces unsatisfactory results in the presence of persistent AV valve regurgitation and hence, it should be reserved for patients with a relatively competent AV valve but with a large left-to-right shunt.[3, 7, 10,11]

Corrective surgery may be performed as a primary procedure or as a 2nd-stage operation following PA banding.[7, 10, 11] With improvements in surgical techniques, anesthetic and postoperative management, the risk of corrective surgery in infancy has steadily decreased. As a result, primary corrective repair is preferred over staged repair well before the onset of irreversible pulmonary hypertension.[3,4,5,11]

We hereby describe our experience of the repair of AV canal defects and present results of our retrospective analysis of patients operated upon during a 10 year period.

AIMS AND OBJECTIVES:

To review and retrospectively analyse data on patients operated for atrioventricular canal defects in our institute (between August 2006 and July 2016) and to determine patient and operative variables influencing survival.

MATERIALS AND METHODS:

PATIENT POPULATION:

Twenty- eight patients referred to us with a diagnosis of AV canal defects (partial or complete) were operated upon after the necessary investigations under a senior surgeon. One patient was excluded from this study because of nonavailability of proper records. All other patients were included in the study.

TABLE 1: TYPE OF DEFECT

(n_{-27})	Frequency	Percent	Valid Percent	Cumulative Percent
COMPLETE	9	33.3	33.3	33.3
INTERMEDIATE	2	7.4	7.4	40.7
PARTIAL	16	59.3	59.3	100.0

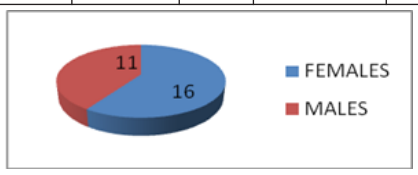

FIGURE 1: Sex Distribution

A female preponderance was noted in the group with 16 out of the 27 being females patients (59%). Female to male ratio was 16 to 11 (1.45:1). (Figure 1). The youngest patient who underwent surgery was 7 months old at the time of surgery. The mean age of the patients in our cohort was 4.67+/- 2.2 years in complete AV canal defects group (range 1.5 and 7 years), 0.58 years in the intermediate AV canal defects and 16.56 +/- 10.35 years (range 3 to 35 years) in the partial group.

The mean weight of patients was 26.74 kgs (range 4.5 kgs and 66.5 kgs). The mean body surface area of the patients was 0.949 (range 0.28 to 1.86).

TABLE 2: Associated Syndrome

ASSOCIATED SYNDROME	Frequency	percent
DOWN SYNDROME	5	8.5

BUTTERFLY D 11 VERTEBRA WITH SCOLIOSIS	1	3.7
ELLIS VAN CREVALD	1	3.7
JUVENILE DIABETES	1	3.7
LAWRENCE MOON BEIDL	1	3.7
MALROTATION OF GUT	1	3.7
POLYDACTYLY	1	3.7
WPW SYNDROME	1	3.7
NONE	15	55.6

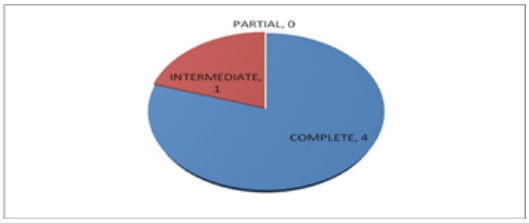

FIGURE 2: Down Syndrome

PERIOPERATIVE MANAGEMENT

Antibiotic protocol followed in all patients included-

Inj. Cefuroxime 25 mg/ kg per dose [(II generation cephalosporin) up to a maximum of 750 mg/kg/ dose] and
Inj. Gentamicin 1 mg/ kg/ dose (aminoglycoside)

These were given with premedication and at induction. It was followed with three daily doses of Inj. Cefuroxime 25 mg/ kg per dose (upto a maximum of 750 mg/kg/ dose) for 2 days and changed to oral form of the same for another 3 days. Inj. Gentamicin 3 mg/ kg once daily was continued for 5 days.

CARDIOPULMONARY BYPASS:

Minimax/ Affinity, Medtronic, USA prime membrane oxygenators were used in our patients. Aortobicaval cannulation was used in all patients. The mean total bypass time was 101.52 mins (range 54 to 179 min) and the cross clamp time was 63.19 min (range 28 to 124 min). Cold blood cardioplegia (St. Thomas I cardioplegia solution with procaine) was used in all patients, with an average of 3.2 cardioplegic doses per patient. All patients were cooled to moderate hypothermia (28-32 deg C).

SURGICAL TECHNIQUE

Midline sternotomy performed. Thymus excised. Pericardial patch harvested in all patients. Systemic heparinization using 3mg/ kg of heparin. Cardiopulmonary bypass commenced in all patients with aortobicaval cannulation. Vena cavae taped. Aortic root catheter was placed. Moderate hypothermia was used in all patients. The aorta was cross clamped and cold blood cardioplegia is given down the root (calculated dose of 20 ml/ kg). Topical ice cold saline was used for surface cooling. Repeated doses of cardioplegia used to maintain the arrest. The complete AV canal defects were repaired by a two- patch technique using Dacron and pericardial patches for the ventricular and atrial septal defects respectively and closure of the left AV valve (mitral valve) cleft. The patients with partial variant of AV canal defects underwent pericardial patch closure of the ostium primum atrial septal defect and repair of the mitral valve cleft in a manner similar to the complete AV canal defects.

A right atriotomy is made anterior to the interatrial groove. The left side of the heart was vented across the septal defect. The anatomy of the AV canal confirmed and typed according to the Rastelli's classification for complete AV canal defects. Cold saline was injected through the common leaflet and the exact area of apposition of the left superior and inferior leaflets noted. A 5-0 prolene suture taken through this zone of apposition along the free edge. Ventricular septal defect is closed with a 'D' shaped patch, taking the sutures on the RV aspect of the septum and keeping all the chordae on the left side. Interrupted pledgetted sutures are then passed through the upper border of the dacron patch, through the left superior and inferior leaflets and then through the pericardial patch and tied down. The cleft in the mitral valve was then repaired with interrupted 6-0 prolene sutures. Valve was then rechecked for leaflet apposition and valvular incompetence. Ostium primum defect closed with pericardial patch using continuous sutures taking the sutures on the rim of the coronary sinus, so as to allow the coronary sinus to drain into the RA in all cases.

Left side of the heart is then deaired and the cross clamp released. Right atriotomy is closed. Partial bypass resumed. Two temporary epicardial pacing wires are placed on the RV surface. Inotropic support (Inj. Adrenaline) started. Patient weaned off bypass and Protamine given for complete heparin reversal.

POSTOPERATIVE MANAGEMENT

Ventilation was accomplished with the Servo Ventilator (Siemens-Elema AB, Solna, Sweden) in all patients with endotracheal tube or nasotracheal tube (for small children). Patients were ventilated in the pressure controlled mode with a tidal volume of 10-12ml/ kg. The minimum inspired oxygen fraction that provides acceptable arterial oxygen saturation was used (usually 50% of oxygen).

Sedation and analgesia consisted of a continuous infusion of injection Fentanyl at a dose of 5-10 microgram/ kg/ hr for all children till 4 hours prior to extubation. For adult patients sedation and analgesia consisted of intermittent boluses of injection Morphine. Neuromuscular blockade was achieved with intermittent boluses of Vecuronium if needed.

Hemoglobin value and serum electrolytes were checked and arterial blood gases analysed in all patients at arrival to the ICU and appropriately corrected. A bed side chest roentgenogram was obtained to confirm the correct position of the endotracheal tube, drains, invasive monitoring lines and also to rule out pleural or pericardial collection or pneumothorax and acted appropriately. Inotropic dosage was adjusted according to the hemodynamic status and all the inotropes were usually continued till the time of extubation. All the inotropes gradually tapered, twelve hours after extubation if the patient was hemodynamically stable.

STATISTICAL ANALYSIS

The various variables considered to have a possible influence on the survival including, presence or absence of a syndromal association, perioperative, data were studied for statistical significance. The variables distributed along the Gaussian curve were analyzed by the 'T' test for statistical significance and those not following the normal curve of distribution evaluated by the Mann- Witney U test. The various Chi square tests (Pearson's Chi square and Fisher Exact test) were then used to validate the statistical significance thus obtained. The statistical analysis was performed using the SSPS software, version 16, Chicago, Illinois, USA.

RESULTS

The patients who presented to us with the diagnosis of AV defects were operated after a thorough evaluation. The types of the AV canal defects and the occurrence of various features regarding their distribution in each of the three groups of patients was studied.

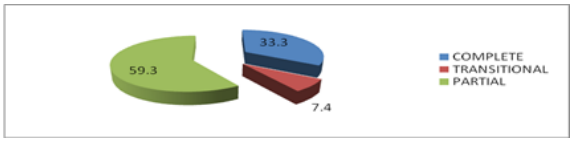

FIGURE 3: Type Of Av Canal Defect

Of the 27 patients in the study group, 16 patients belonged to the partial AV canal defects type accounting for 59.3% of the patients. Of the remaining, 9 (33.3%) patients were of the complete AV canal defect type and 2(7.4%) belonging to the transitional/ intermediate AV canal defect type (Table 1 and Figure 3).

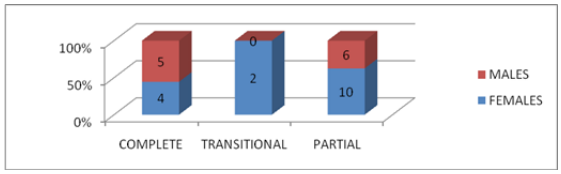

FIGURE 4: Sex Distribution

In the complete AV canal defects group, 44.6% (4 of 9) of the patients were female patients. They accounted for 100% (2 of 2) and 62.5% (10 of 16) of the patients in the transitional and partial groups respectively. Males comprised 55.6% of the patients (5 of 9) in the complete AV canal defects group and 37.5% (6 out of 16) in the partial AV canal defects group. (Figure 4)

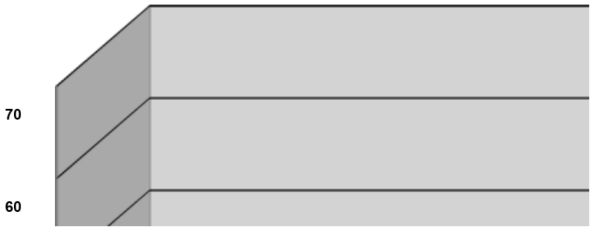

FIGURE 5: Weight Of Patients (in Kilograms)

The mean body weight of the patients in our study was 11.33+/- 3.9 kgs in complete AV canal defects group (range 7 to 17 kgs), 4.8+/- 0.28 kgs in the intermediate AV canal defects and 8 +/- 19.18 kgs (range 8 to 66 kgs) in the partial AV canal defects group. The smallest child weighed 5 kgs in the intermediate AV canal defects group. (Figure 5)

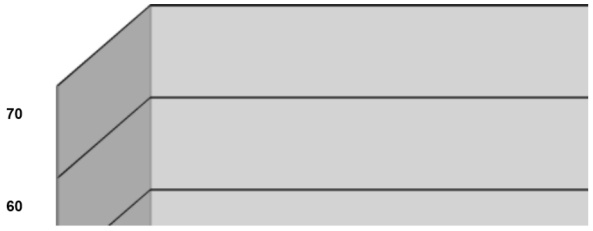

FIGURE 6: Post Operative Hospital Stay

The median post operative hospital stay in the present set of patients was 10days. The mean hospital stay was 11.33+/- 5.679 days in complete AV canal defects group (range 1 to 22), 11.50+/- 2.12 days in the intermediate AV canal defects (range 10 to 13 days) and 11.56+/- 7.83 days (range 0 to 31 days) in the partial AV canal defects group. (Figure 6)

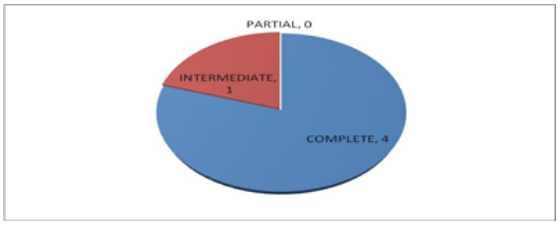

FIGURE 7: Down Syndrome

Down syndrome was the most common associated syndrome in our patents occurring in 4 complete AV canal defects patients (4 of 9 patients) and one infant with interstitial AV canal defects (one of the 2 infants).(Figure 7)

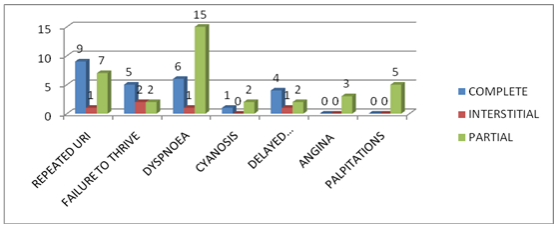

FIGURE 8: Distribution Of Symptomatology

Dyspnoea on exertion was the most common mode of presentation seen in 22 patients (84.6%), the majority of the whom (15, 68%) belonged to the partial type of AV canal defects. Recurrent URI was the next most common symptom seen in 17 of the 26 patients (65%), 9 patients belonging to the complete variant of AV canal defects. Other symptoms were equally distributed in the three groups.(Figure 8)

TABLE 3: Distribution Of Radiographic Features

	COMPLETE	INTERMEDIATE	PARTIAL
CARDIOMEGALY	9 (100.0)	2 (100.0)	10 (62.5)
PLETHORA	9 (100.0)	2 (100.0)	10 (62.5)
NORMAL RADIOGRAPH	0	0	4 (25)
PROMINENT PAs	3 (33.3)	0	2(12.5)
SKELETAL ABNORMALITY	0	0	1(6.2)

Cardiomegaly and pulmonary plethora, as expected, were the most common radiographic features seen in 21 patients (80.76 %), both seen in all patients with complete and interstitial variants of AV canal defects, and in 10 (62.5%) of the patients with partial AV canal defects. Four patients with partial AV canal defects had a normal chest radiograph. (Table. 3)

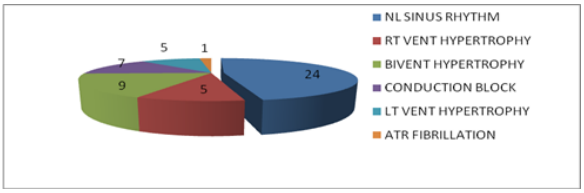

FIGURE 9: Electrocardiographic Findings

Twenty four patients (92.3 %) had a normal sinus rhythm, seen in all patients with complete and interstitial variants of AV canal defect and in 10 (62.5%) of the patients with partial AV canal defect. Biventricular hypertrophy in 9 patients (33%) comprised the next common finding. Conduction block was noted in 7 patients (25.9%), 1 in the complete AV canal defect group (bifascicular block with normal heart rate) and 6 in the partial AV canal defect group (4 had a first degree AV block and the other two had incomplete right bundle branch block). Atrial fibrillation was noticed in one patient. (Figure 9)

TABLE 4: Cardiac Catheterization

CARDIAC CATHETERIZATION	COMPLETE	INTERMEDIATE	PARTIAL
DONE	8 (88.9)	1(50)	7(43.8)
NOT DONE	1(11.1)	1(50)	9(56.2)

Sixteen out of 27 patients (61.5 %) underwent a preoperative cardiac catheterization and angiography. It was done in most of the patients with complete variant of AV defects (8 of 9 patients, 88.9 %). One of the 2 patients with interstitial AV canal defects and 7 of 16 (43.8%) patients with partial AV defects underwent this study to assess the anatomy and state of pulmonary hypertension.(Table 4) Among the findings of the 16 catheterisation studies, AV valve regurgitation was the most common finding seen in 14 patients (87.5 %). OP ASD was the next most common feature noted in 13 (81.25%), followed by the other characteristic findings of the AV canal defect, the gooseneck deformity of the LVOT tract (13, 81.25%), AV valve cleft (11, 68.75%) and the inlet ventricular septal defect (7,43.75%). Additional VSDs were seen in 5 patients.

TABLE 5: Distribution Of Pa Pressures (n=16)

	COMPLETE	INTERMEDIATE	PARTIAL
Minimum (mmHg)	6	31	6
Maximum (mmHg)	56	31	47
Mean (mmHg)	33.625	31	23.85
Standard deviation	17.524	00	13.158

Mean PA pressures in these patients were 33.624 +/- 17.524 mmHg, 31 mmHg and 23.85 +/-13.158 mmHg in the complete, interstitial and partial defects respectively. (Table 5)

TABLE 6: Distribution Of Magnitude Of Shunt (n=16)

Qp/Qs	COMPLETE	INTERMEDIATE	PARTIAL
Minimum	1	3.8	1.6
Maximum	16.27	3.8	1.6
Mean	4.574	3.8	1.6

Mean Qp/Qs was 4.57+/- 5.27 l/mins, 3.8 l/mins and 1.6 l/mins in the complete, interstitial and partial AV canal defect groups respectively. (Table 6)

The mean pulmonary vascular resistance in the complete AV canal defect group was 5.0 +/- 9.56 units. This high value was due to the skewed nature of the data, with one high PVR value distorting the final mean values for the entire group. The mean values in the interstitial and partial groups being 0.61 units and 2.6 units respectively.

Likewise the mean pulmonary vascular resistance index for the complete AV canal defect group was .68+/-2.36. For the transitional and partial groups it was 0.25 and 1.2 respectively.

TABLE 7: Bypass Time And Cross Clamp Time (minutes)

The total bypass time for our group of patients was 101.52+/- 30.40 mins and the mean cross clamp time 63.19 +/- 23.21 min.(Table 7) The mean bypass time was 122.26+/- 25.19 mins, 90+/-21.21 mins and 91.06+/-28.82 mins in the complete, interstitial and partial groups respectively. The corresponding mean cross clamp times were 81.67+/- 24.53 mins, 48+/-8.48 mins and 54+/-17.19 mins in the three groups. Patients in the complete AVCD group had the maximum mean bypass and cross clamp time for obvious reasons of the need for an additional step of ventricular septal defect patching.(Table 8)

TABLE 8: Bypass Time And Cross Clamp Time In The Three Groups (minutes)

AVCD	(IN MINUTES)	MI	MAX	MEAN	STD DEVIATION
COMPLETE	BYPASS TIME	67	153	122.67	25.199
	CROSS CLAMP TIME	42	124	81.67	24.536
INTERMEDIATE	BYPASS TIME	75	105	90	21.213
	CROSS CLAMP TIME	42	54	48	8.485
PARTIAL	BYPASS TIME	54	179	91.06	28.822
	CROSS CLAMP TIME	28	95	54.69	17.196

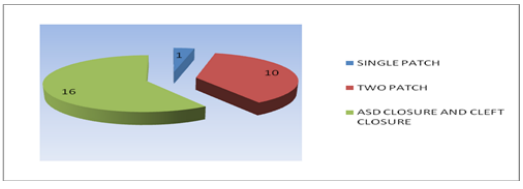

FIGURE 10: Types Of Surgery

Most of our patients belonged to the partial AV canal group (16 patients) and hence underwent ASD patch closure and repair of the cleft. The two patch repair of the AV canal group involving separate closure of the atrial and ventricular defects and repair of the mitral cleft was done in 10 patients. The conventional single patch technique was done in only one case. (Figure 10)

TABLE 9: Duration Of Ventilation And Inotropic Support

DURATION (DAYS)	Minimum	Maximum	Mean	Std.
VENTILATION	1	4	1.5	0.76158
INOTROPIC SUPPORT	1	3	1.653	0.6288

The mean duration of post operative mechanical ventilation was 1.5 +/- 0.7 days. The mean post operative mechanical ventilation in the three groups was 1.5+/- 0.76 days, 1 and 1.6+/- 0.91 days respectively. The mean duration of inotropic support was 1.65+/- 0.62 days. The mean post operative inotropic support in the three groups was 1.65 +/- 0.62 days, 1 and 1.8+/- 0.67 days respectively. (Table 9 and Figure 11)

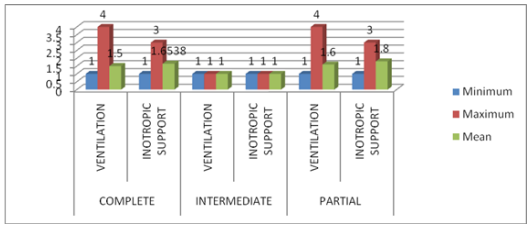

FIGURE 11: Duration Of Ventilation And Inotropic Support In The Three Groups

TABLE 10: Associated Cardiac Lesion

ASSOCIATED CARDIAC LESION	FREQUENCY
LSVC	5
OS ASD	4
PAPVC	1
SINGLE ATRIUM	1
MILD LPA STENOSIS	1
PFO	1

Intraoperatively, a left SVC was encountered in 5 patients which was dealt with separately. An associated OS ASD was noted in 4 patients which was joined with the primum defect and patched using the same pericardial patch. Partial anamolous pulmonary venous connection involving the right superior pulmonary vein was noted in one case which was rechanneled into the left atrium during the ASD closure. Single atrium was noted in one case and partitioned using the pericardial patch. Mild left pulmonary artery stenosis and patent foramen ovale were noted in one case each and left alone. (Table 10)

TABLE 11: Early Post Operative Complications

COMPLICATION	COMPLETE	INTERSTITIAL	PARTIAL
TEMPORARY PACING	4	0	7
HEART BLOCK	3	0	1
FEBRILE ILLNESS	3	1	5
PERICARDIAL EFFUSION	1	0	3
PNEUMOTHORAX	2	0	0
PULMONARY HYPERTENSIVE	1	0	0
ACUTE ABDOMEN	0	1	0

The most common complication noted in our patients after surgery was the need to temporarily pace for a conduction block and bradycardia. It was seen in 11 patients (4 with complete AV canal and 7 with partial AV canal defect). These patients were started on isoprenaline infusion initially and later on Tab. Alupent (Orciprenaline, analogue of isoprenaline) 10-20 mg thrice daily.

Febrile illness was noted in 8 patients. One of them diagnosed to have filariasis and managed accordingly. Rest had non- specific febrile illness which was managed conservatively. Four of our patients had mild pericardial effusion which was managed conservatively. Two patients developed pneumothorax and was managed with tube thoracostomy.

One patient in the complete AVCD group with severe pulmonary arterial hypertension preopertively and developed severe pulmonary hypertensive crisis on the first post operative day while on ventilator and succumbed to it. (Table 11)

In the post operative period the cardiomegaly and pulmonary plethoric changes were noted to have significantly regressed and noted in 8(32%) and 5(20%) patients respectively as against 21 patients each (80.76%) preoperatively.

TABLE 12: Post Operative Left Av Valve Regurgitation (post Op Day 5)

SEVERITY	FREQUENCY	PERCENT
TRACE	10	40
MILD	10	40
MODERATE	5	20

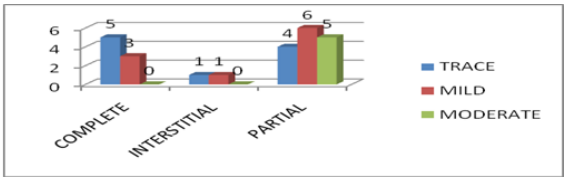

FIGURE 12.postoperative Left Av Valve Regurgitation

Our patients had an effective repair of the AV canal defect with only trace or mild mitral regurgitation noted in 20 out of the 25 (80%) patients postoperatively. Moderate mitral regurgitation was noted in 5 patients. None of the patients had severe mitral regurgitation. The patients of the partial AV canal defect group demonstrated a higher frequency of AV valve regurgitation 4,6 and 5 patients having trace, mild and moderate regurgitation respectively. None of the patients with complete AV canal defect repair had more than mild regurgitation. (Table 12, Figure 12)

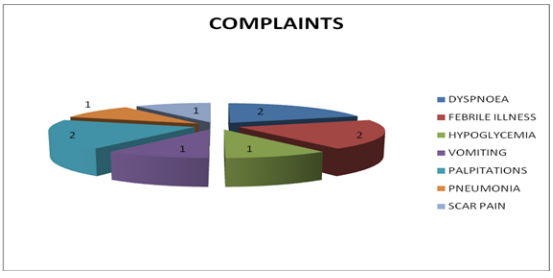

FIGURE 13. Complaints At Follow Up

Four of our patients were lost to follow up with no post operative reviews. At follow up, the duration of which ranged from 1 month to 7.6 years, 11 patients had no significant complaints. Dyspnoea on exertion and transient febrile illness was noted in 2 patients each. One patient presented with features of digitalis toxicity. One child presenting with bronchopneumonia, right lung, and another case of type I diabetes mellitus who presented with recurrent hypoglycemia were managed in child health department. (Figure 13)

Normalization of the radiographic features was noted in most of the patients and 13 of 21 (61.9%), presented at the time of follow up at 6 months, with a normal chest radiograph. Cardiomegaly was still noted at 6 months in 6 patients (28.5%). Pulmonary plethora and right lobar pneumonia was noted in one patient each at the 6th month.

Most of the patients had a normal sinus rhythm at the time of follow up (20/ 21, 95.2%). None had complete heart block. 12 patients had right bundle branch block (7 and 5 in complete and partial AV canal defects group). Five patients had bifascicular block with controlled heart rate (4 and 1 in complete and partial AV canal defects groups respectively). One patient of the partial AV canal defects group had atrial fibrillation.

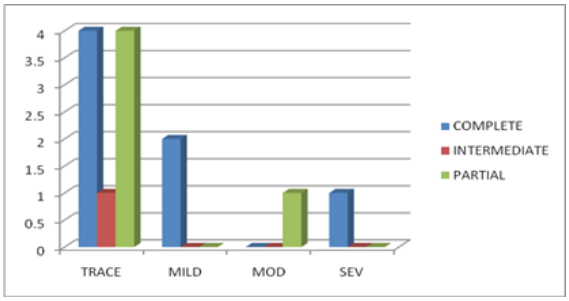

FIGURE 14 : Left Av Valve Regurgitation During Follow Up

The patients improved with respect to their left AV valve regurgitation with 6 patients having trace (4) to mild (2) regurgitation and only one having severe regurgitation in the complete AV canal defect group. One patient had trace regurgitation in the interstitial group. 4 patients had trace and 1 had moderate regurgitation in the partial AV canal defect group. (Figure 14)

MORTALITY DATA

Early mortality :

Two of our patients died after the surgery within the first 24 hours and accounted for an early mortality rate of 7.4%.

The first patient operated during the early part of study period was a 7 years female child with a partial AV canal defect. Clinical examination revealed features of an atrial septal defect with pulmonary hypertension. She had cardiomegaly and pulmonary plethora on radiographic evaluation. Electrocardiography revealed a normal sinus rhythm with features of biventricular hypertrophy and first degree conduction block. A preoperative echocardiography revealed features of partial AV canal defect with moderate left AV valve regurgitation. Cardiac catheterization was not done. Intraoperatively she was found to have a partial AV canal defect. The valve tissue looked deficient with thickening of the leaflets and severely regurgitant. The PA was severely tense. A primary valve repair and pericardial patch closure of ASD was done. There was difficulty in coming off bypass. A revision was done and valve was found to be severely leaking and mitral valve replacement was done. The total bypass time was 179 minutes and cross clamp time was 95 minutes. These were definitely in excess of the mean timings for this group of patients. The patient was transferred on high inotropic support. She remained hemodynamically unstable throughout the early postoperative period and finally succumbed to a possible biventricular failure within 12 hours of surgery.

The second case was a 19 months female child with Down syndrome and had a complete AV canal defect, type A. On examination she had features of VSD with severe pulmonary hypertension. Radiographic evaluation revealed cardiomegaly and pulmonary plethora and had a normal sinus rhythm with features of biventricular hypertrophy. A preoperative echocardiography revealed features of complete AV canal defect with moderate AV valve regurgitation. A catheterization was done which revealed features of complete AV canal, type A, with significant AV valve regurgitation. Her PA pressures were 56 mmHg with Qp /Qs of 1 which increased to 2.5 post oxygenation. The pulmonary vascular resistance was 22.5 units which reduced to 8.74 units post oxygenation and the pulmonary vascular resistance index falling from 5.87 to 3.05 defining a very high pulmonary arterial resistance with reversibility.

Considering as the only option, a surgical repair was offered. She underwent a single- patch repair with the total bypass of 120 minutes and cross clamp time of 61 minutes, timings well within the range for the mean total timings for the complete AV canal defects group. Postoperatively the patient was managed with pulmonary arterial hypertension lowering measures. The patient however had an episode of severe pulmonary arterial hypertensive crisis and succumbed to it on the first post operative day.

Late mortality:

We had one late death but it was due to a non cardiac cause. This death resulted in a late mortality rate of 4%.

This 13 years female child was a case of partial AV canal defect with juvenile diabetes. She underwent two patch repair at the age of 5 years and had a uneventful recovery. She had a well functioning mitral valve repair with only trace to mild AV valve regurgitation. She was under the follow up of child health division for the management of the insulin dependent diabetes till recently when she expired after a follow up period of 93 months.

MORBIDITY DATA:

We studied the follow up of the patients in the surviving group of patients for the various morbidity data. Four of our patients were lost to follow up with no subsequent follow up after initial surgery and no response to recent correspondence.

We looked into the following factors in the follow up data of the 21 surviving patients in the three groups to evaluate the morbidity -

1. Worsening of the left AV valve regurgitation requiring reoperations.
2. Need for insertion of a permanent pacemaker for conduction block.
3. Occurrence of left outflow tract stenosis requiring reoperations.
4. Any surgical procedures for associated left sided obstructive lesions.

None of our surviving patients underwent any reoperations left AV valve regurgitation or for left ventricular outflow tract obstruction. One patient operated for complete AV canal defect, Type A, had a small residual ventricular septal defect and presented with right lower lobe bronchopneumonia at follow up and was managed conservatively. Another patient was suspected to have coarctation of aorta at follow up which however was ruled out in the absence of significant left ventricular to aortic gradient on a cardiac catheterization and angiography.

One of our patients required a permanent pacemaker after 9 months of surgery for symptomatic bifascicular block and has since been doing well. This patient was operated for a complete AV canal defect type A, at the age of 7 years and was discharged after an uneventful recovery in normal sinus rhythm. At 6 months follow up she was found to have a bifascicular block (RBBB and LAHB). She was kept observation under the cardiologists care and underwent a permanent pacemaker insertion for symptomatic bifascicular block after 3 months.

The rate of pacemaker insertion and residual ventricular septal defects cannot be deduced accurately due to the patients lost to follow up.

The various variables were checked for statistically significant inter-group differences withrespect to their occurrence in the two following ways.

1. Between the three types of AVCD- complete, interstitial and partial.
2. Between the mortality group and the surviving group.

1. Between the three types of AV canal defects- complete, interstitial and partial.

None of the demographic variables were noted to be significantly different in the three groups. However a trend towards an earlier repair was noted in the three subsets over the duration of study.

- **Mean bypass time and mean cross clamp time:**

When the Anova test was applied to study for the statistically significant difference, the mean bypass time and mean cross clamp time were found to be significantly different among the three groups of patients with a 'p' value of 0.031 and 0.008 respectively (The mean difference is significant at the 0.05 level, 'p'< 0.05). The mean bypass time was 122.26+/-mins and 91.06+/-28.82 mins in the complete and partial groups respectively.The corresponding mean cross clamp times were 81.67+/- 24.53 mins and 54+/-17.19 mins.

TABLE 13: Anova

		Sum of Squares	df	Mean Square	F	Sig.
BYPASS TIME	Between	6039.803	2	3019.902	4.029	**0.031**
	Within Groups	17990.938	24	749.622		
	Total	24030.741	26			
CROSS CLAMP TIME	Between	4690.637	2	2345.318	6.037	**0.008**
	Within Groups	9323.438	24	388.477		
	Total	14014.074	26			

(Between groups: Between the complete, intermediate and partial types. Within groups: Within patients of the same group)

When this was checked by the Bonferroni correction for paired observations (between the complete and the partial AV canal defect groups) it was noted to be significant at the 0.05 level with 'p' values of 0.032 and 0.009.

TABLE 14 Multiple Comparisons (Bonferro ni)

Dependent Variable	COMPLETE/ COMPLETE/ INTERMEDIATE INTERMEDIATE / / PARTIAL PARTIAL	Mean Difference (I-J)	Std. Error	Sig.	95% Confidence Interval	
					Lower Bound	Upper Bound
BYPASS TIME	**COMPLETE** INTERMEDIATE	32.667	21.403	.420	-22.42	87.75
	PARTIAL	31.604*	11.408	**.032**	2.24	60.96
	INTERMEDIATE COMPLETE	-32.667	21.403	.420	-87.75	22.42
	PARTIAL	-1.062	20.534	1.000	-53.91	51.79
	PARTIAL **COMPLETE**	-31.604*	11.408	**.032**	-60.96	-2.24
	INTERMEDIATE	1.062	20.534	1.000	-51.79	53.91
CROSS CLAMP TIME	**COMPLETE** INTERMEDIATE	33.667	15.408	.117	-5.99	73.32
	PARTIAL	26.979*	8.212	**.009**	5.84	48.11
	INTERMEDIATE COMPLETE	-33.667	15.408	.117	-73.32	5.99
	PARTIAL	-6.688	14.782	1.000	-44.73	31.36
	PARTIAL **COMPLETE**	-26.979*	8.212	**.009**	-48.11	-5.84
	INTERMEDIATE	6.688	14.782	1.000	-31.36	44.73

*. The mean difference is significant at the 0.05 level.

Difference in the total bypass times and cross clamp times between the complete and partial groups of patients was statistically significant.

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- **Type of surgery**

When the types of surgery were studied for their distribution in the three groups by the univariate analysis, a statistically significant association was noted ('p'= 0.018). This due to the larger fraction of the patients being of the partial AV canal type and undergoing mitral cleft repair and pericardial patching of the atrial septal defect.

2. Between the patients survival group and the mortality group

When the patients in the surviving and mortality groups were considered, there was a statistically significant difference in the type of the surgical repair in the two groups (p= 0.003). The 2 patients who expired underwent single patch repair in one and pericardial patching of the atrial septal defect and mitral cleft closure followed by mitral valve replacement in the other, surgeries not performed in the surviving group of patients.

The observation of one patient in the mortality group with a partial AV canal defect having a normal sinus rhythm with biventricular hypertrophy and first degree conduction block was found to be statistically significant with mortality with a 'p' value of 0.003 (p< 0.05) but this wasn't noted in the multivariate analysis.

TABLE 15: Variables And P Values

VARIABLES	p value
POST OP HOSPITAL STAY	0.014
HEMOGLOBIN	0.343
PA PRESSURE (mm Hg)	0.127
Qp/Qs	0.12
PVR	0.134
PVRI	0.134
BYPASS TIME	0.078
CROSS CLAMP TIME	0.195

FLOW RATE(LTS/MIN)	0.228
ECHO- AVV REGURGITATION	0.198
CATH - AV V REGURGITATION	0.052
POD5- ECHO MR	0.351
ECHO- AV VALVE REGURGITATION	0.089

None of the others were found to have any association of statistical significance with respect to their distribution in the three AV canal defect types or the survival and mortality groups namely - demographic variables, postoperative hospital stay, clinical features, haemoglobin levels (preoperative or post operative), radiographic features, echocardiographic features, features on cardiac catheterisation- pulmonary artery pressures, Qp/Qs, pulmonary vascular resistance, pulmonary vascular resistance index, mean cross clamp time and mean bypass time (in the mortality and surviving groups) , flows rates on bypass, post operative duration of mechanical ventilation, duration of inotropic support, post operative radiographic, electrocardiographic or post operative echocardiographic features, follow up symptoms, radiographic, electrocardiographic or echocardiographic features. (Table 15)

A regression analysis could not be applied to further validate the statistical significance noted above due to the limited number of variables found to have a statistical association.

DISCUSSION

The early days of AV canal defect repairs were associated with operative mortalities as high as 50%. Over the past 4 decades, operative mortality has declined to 3% to 6% in most series. Factors cited for improved results include improved accuracy of preoperative diagnosis (including associated congenital heart disease as well as AV valve morphology and ventricular size), improved intraoperative support including myocardial protection, and improved postoperative management. [3,4-7,10,11,23-41]

Our early mortality rate was 7.4% is well within the range of mortality rates reported from various centres. Our late mortality rate of was 4% (but the cause of death in that was non cardiac).

Coincident with a decrease in mortality over time is an increase in the rate of repair of AV canal defect during infancy. This transition to routine early repair accomplished without an increased risk of mortality, and also avoiding the use of palliative procedures. [3,6,34,37-41,47-53,55-57]

The youngest patient who underwent surgery in our study was 7 months old at the time of surgery and belonged to the intermediate AV canal defects group. The mean age of the patients in our study cohort was 4.67+/- 2.2 years in complete AV canal defect group (range 1.5 and 7 years), 0.58 years in the intermediate AV canal defect and 16.56 +/- 10.35 years (range 3 to 35 years) in the partial canal defect group. Though the mean age of patients is higher than noted in most series for the complete and partial groups, the youngest kid operated with complete AV canal defect was 1.5 years and that in the partial group 3 years.

We routinely prefer primary repair of all patients presenting to us with no palliative procedure carried before the definite repair.

A female preponderance was noted in our patients with 16 out of the 27 being female patients (59% of patients). Female to male ratio is 1.45:1. This is perhaps the case with most studies. [3]

Ostium primum defects or the partial AV canal defects occur more frequently than once thought. Partial AV defects represent approximately 25% of the AV canal defects. [41] In our study however 59.3% (16 of 27 patients) of the AV canal defects were of the partial variant.

We did not obtain cardiac catherterization in our patients. Mean PA pressures in the 16 catheterization studies performed on our patients was 33.624 +/- 17.524 mmHg, 31 mmHg and 23.85 +/- 13.158 mmHg in the complete, interstitial and partial defects respectively. The mean pulmonary vascular resistance was 5. +/- 9.56 units in the complete AVCD group. Likewise the mean PVRI for the complete AVCD group was 1.68+/-2. 36. For the transitional and partial groups it was 0.25 and 1.2.

Progressive ventricular dilatation from volume overload with progressive AV valve annular dilatation may be avoided by correction during early infancy, thus decreasing the complexity of the repair and the potential for residual left AV valve regurgitation. [3,6,34,37-41,47-53,55-57]

Problems related to persistent PAH have largely been overcome by operating on infants in the first 6 months of life. Ventilator time, ICU length of stay, and overall length of hospital stay have continued to decrease significantly. [74-77]

The median post operative hospital stay in the present set of patients was 10 days also noted in most studies.

Down syndrome was the most common associated syndrome in our patents (18.5% of patients) occurring in 4 complete AV canal defect patients (4 of 9 patients, 44.4%) and one infant with interstitial AV canal defect (one of the 2 infants, 50%). These figures well noted in the literature which report that 50- 75% of patients with complete AV canal defect and about 10% of patients with partial AV canal defect have associated Down syndrome. [3,4,5]

Conduction block was noted in 7 out of the 27 patients (26.9%), 1 in the complete AV canal defects group and 6 in the partial AV canal defects group preoperatively. The patient in the complete AV canal defect had a bifascicular block with normal heart rate. 4 of the patients in the later group had a first degree AV block and the other two had incomplete right bundle branch block. Most of the patients had a normal sinus rhythm at the time of follow up (20/ 21, 95%). Five patients had associated bifascicular block (4 and 1 in complete and partial AV canal defects groups respectively). One patient (4.7%) had symptomatic bifascicular block and required a pacemaker insertion after 9 months of surgery. None had complete heart block. This rate though a little higher has been noted in previous studies

Patients in the complete AV canal defect group having higher mean bypass and cross clamp time for obvious reasons of the need for a additional step of VSD patching. The mean bypass time for our group of patients was 101.52+/- 30.40 mins and the mean total bypass time cross clamp time being 63.19 +/- 23.21 min.

The mean duration of post operative mechanical ventilation was 1.5 +/- 0.7 days. The mean duration of inotropic support was 1.65+/- 0.62 days.

Patients with moderate to severe preoperative left AV regurgitation can be expected to have a significant reduction in post repair left AV regurgitation with long-term survival similar to patients who undergo operation with mild or less severe left AV regurgitation. [28,33-35,37,39,40,59,60]

Our patients had an effective repair of the AV canal defect with only trace or mild mitral regurgitation noted in 20 out of the 25 (80%) patients postoperatively. Moderate mitral regurgitation was noted in 5 patients. None of the patients had severe mitral regurgitation. The patients of the partial AV canal defect group demonstrated a higher frequency of AV valve regurgitation 4,6 and 5 patients having trace, mild and moderate regurgitation respectively. None of the patients with complete AV canal defect repair had more than mild regurgitation.

However, persistent postoperative left AV (mitral) valve regurgitation continues to be a problem and consistently identified in most series. [3,4-7,9-11,23-41] Because the incidence of late postoperative left AV valve regurgitation requiring reoperation is so remarkably constant in most series using both single and double-patch techniques, one possibility may be the 5% to 10% of patients with complete AV canal defect have such abnormal AV valves that long

term competence may be difficult or impossible to obtain in this particular subset.

During follow up it was noted that our patients had improved with respect to their left AV valve regurgitation with 6 patients having trace (4) to mild (2) regurgitation and only one having severe regurgitation in the complete AV canal group. One patient had trace regurgitation in the interstitial group. 4 patients had trace and 1 had moderate regurgitation in the partial AV canal group.

The incidence of reoperation for mitral regurgitation ranges between 3.5% and 19.7%.[28, 37, 40] None of our patients had a reoperation for their left AV valve regurgitation.

It is essential that everything possible be done at the time of the initial operation because reoperation for early left AV valve regurgitation is clearly associated with poorer outcomes.[37,40]However, continued refinement of surgical technique in conjunction with careful pre- and intra-operative TEE should hopefully decrease this incidence over time.

It is not clear whether the single-patch (conventional or modified) or double-patch techniques is superior to the other. It is clear that excellent results may be obtained using either technique and that the most important factor is likely the individual surgeon's skill and familiarity with a particular technique as opposed to the technique itself.[28,37,40,66]

Whether a single-patch or two-patch technique is superior in repair of complete AV canal defects with a significant ventricular component is a technical issue of controversy.[66]

We at our institute continue to prefer the two-patch technique because it avoids the need to divide and reattach the leaflets thereby preserving valve tissue, particularly important in small infants with a limited quantity of AV valve tissue. A nd also as one becomes comfortable with a certain approach and has what are felt to be acceptable results, it is often difficult to change the paradigm of the surgical approach to a certain lesion.

STUDY LIMITATIONS:

This was a retrospective study with a small number of patients. We as a tertiary centre cater to a patient population from many adjacent states and centres with varying referral patterns. Therefore the difficulty in extrapolating the results with respect to the true incidence of AV canal defects in the population. It also makes follow up difficult with possibility of patients being lost to follow up to accurately assess the late results. This indeed was the case with our study where in 4 patients were lost to follow up.

CONCLUSIONS

In AV canal defects like other cardiac lesions sufficient relief from the symptoms of cardiac failure, with prevention of secondary changes in the heart and lungs, may not be achieved by medical treatment alone. Early surgical intervention, despite the complexity of the lesion, to induce immediate improvement and minimise long term changes in both myocardial and pulmonary vasculature is the clear message in the literature.

AV canal defect is a rare congenital anomaly which can be repaired with low mortality and good early and long-term results. The type of repair whether one patch or two patch can be accomplished with similar results but requires that the surgery be performed early to avoid the complications of irreversible pulmonary obliterative changes and worsening AV valve regurgitation.

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