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Early Outcome of Surgical Repair of Complete Atrioventricular Canal Defects (Comparative Study Between Children before and after 6 Months of Age)

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ABSTRACT

Background: Complete atrioventricular septal defect (CAVSD) is composed of a primum atrial septal defect, nonrestrictive inlet ventricular septal defect and common atrioventricular (AV) valve.

The aim of the work: to compare the result of surgical repair of complete atrioventricular (CAVC) before and after six months of age.

Patients and Methods: Between January 2018 and December 2019 at Al-Azhar University and Atfal Misr hospital, 80 patients underwent CAVC repair. These patients were divided into 2 groups according to the age and each group is of 40 patients. These patients were managed by modified patch and double patch techniques.

Results: In-group A, the mortality was four operative deaths, four patients with severe mitral regurgitation and mean pulmonary artery pressure 30 – 55 mmHg. While in the group B, there were seven operative deaths, two patients with severe mitral regurgitation and mean pulmonary artery pressure 50 – 65 mmHg.

Conclusion: Early repair improve the pulmonary artery pressure with high incidence of surgical reintervention for Lt AV valve, while late repair improve the results for Lt AV valve but has a lower effect on the pulmonary artery pressure.

Keywords: Repair; CAVC; Pediatric; Six; Months.

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INTRODUCTION

Complete atrioventricular septal defect (CAVSD) is composed of a primum atrial septal defect, nonrestrictive inlet ventricular septal defect and common atrioventricular (AV) valve.¹

Down syndrome patients have high incidence of pulmonary vascular obstructive disease (PVOD)² that increase the morbidity and mortality rate. This syndrome is a predisposing factor to increase the intraoperative complications, the need for mechanical ventilation and long ICU and hospital stay.³

Early in life, these patients are complaining of failure to thrive and recurrent chest infection which is life threatening, not responding to medical treatment and may contraindicate the primary repair.⁴

Echocardiography allows direct visualization of the defect spectrum and often a large defect of the midline heart structures are seen. Color Doppler often aids in further visualization of the central opening. An AVSD can give a classical "Gooseneck" sign on a lateral left ventricular angiogram.⁵

Over the last years, there is an advancement of repair of CAVSD starting from two staged technique by

pulmonary artery banding then total repair to primary total repair in early life. Accurate diagnosis of the pathology with perfect orientation of the surgical anatomy played an important role in the development of the surgical result. Also, how to manage the pulmonary hypertensive crisis, myocardial protection during surgery and the postoperative care are important factors in reducing the mortality and reoperation rate.⁶

The ideal timing and the operative technique for the management of CAVSD is still debate. Many authors preferred early intervention as it avoids the pulmonary hypertension development.⁷

PATIENTS AND METHODS

This study is prospective, comparative, non-blinded and multicenter. In the department of cardiothoracic surgery at AL – Azhar university and Atfal Misr hospital, the study was done on 80 patients divided into 2 groups; group A included 40 patients below six months of age and group B included 40 patients after six months of age.

This study included all patients presented with isolated complete atrio-ventricular canal and excluded

cases of complete atrio-ventricular canal associated with repairable congenital cardiac anomalies, unbalanced complete atrio-ventricular canal and redo cases.

In the preoperative period, we followed gender, age, body weight, laboratory tests, chest x-ray, echocardiography (size of VSD, degree of left AV valve regurgitation & degree of pulmonary hypertension) and any medication used for heart failure and pulmonary hypertension.

Intraoperative, under general anesthesia, median sternotomy incision was the standard incision, The pericardium is opened, full dose of Heparin (300 unit/Kg) was administrated, cardiopulmonary bypass was instituted through aortic and bi-caval cannulation, PDA is looked for routinely and ligated if present prior to bypass and CPB was commenced with systemic cooling to moderate levels of hypothermia (28C). The aorta was cross-clamped and cold blood cardioplegia was administered into the aortic root. An oblique incision was made in the right atrium; cold saline is injected into the ventricles to approximate the A-V valve in its closed position. The most anterior point of the left superior leaflet and left inferior leaflet opposing edges was found and a double-armed 6-0 polypropylene suture is placed through it and the cleft was repaired with interrupted sutures. In patients repaired with modified single patch technique, direct suture of the ventricular component, with placement of U sutures, with 5-0 or 6-0 polyester tied on a Dacron fabric to the right side of the ventricular septum. Then the sutures are passed through the antero-superior and postero-inferior valvular leaflets at appropriate sites, demarcating the limits of the right and left atrial outflow tracts, thus determining the size of the right and left AV valve orifices. After tying the ventricular stitches that close the ventricular component and fix the AV valves, the suture of the patch was completed continuously at the atrial level, with 5-0 or 6-0 polypropylene. In patients repaired with double patch technique, the interventricular patch (pericardial or synthetic) was sutured to the right side of the crest of the ventricular septum with continuous 5-0 or 6-0 polypropylene suture. The interatrial patch was then trimmed to appropriate shape and size, and the first part of its insertion is accomplished, then sutured along the edge of the defect. The heart is then re-warmed and de-aired, the Aorta is de-clamped, de-airing of the heart is continued through an aortic root vent. The RA is closed with 5/0 poly-propylene, both caval snares are taken off, and rewarming completed. After stabilization of haemodynamics, patients were weaned from CPB, the venous cannulae are removed, Heparin was neutralized with protamine, and then the arterial cannula was removed. Two retrosternal drainage tubes were inserted, as well as two temporary pace-maker wires which are attached to the RV wall and further two wires to the RA wall if dual chamber pacing was required. We recorded invasive measurement of pulmonary pressure, time of aortic cross clamp, time of total cardiopulmonary bypass, type of surgical technique and any complication occurring during the operation.

In the postoperative period, we followed dose and type of inotropes, period of ventilation, time of ICU and hospital stay, clinical examination, chest x-ray, echocardiography and any postoperative complications before discharge as surgical bleeding, heart block and residual mitral regurgitation.

Follow up of the patients during the first 6 months after discharge by clinical examination, CXR and echocardiography with median follow up duration of 17 months.

Death during hospital stay or within 30 days after the operation was defined as (Early mortality).

Statistical analysis:

Statistical studies of preoperative, operative and postoperative data would include three main items, (1) Descriptive statistics: The main characteristics and parameters of the 80 patients in the 2 groups are summarized with the maximum, minimum, mean value and standard deviation for each variable is presented. (2) Analysis of variance: Most of the preoperative, operative and postoperative data are analyzed, the mean value of each variable of the two groups of patients is presented, the F-ratio (ANOVA test) and the probability (p value) are shown to compare between different means. In most of the variables tested, there is significant difference between the two groups of patients. (3) Correlation of data: Most of the preoperative, operative and postoperative data are correlated with each other. The data of all patients (80) will be correlated with each other. Correlation matrix was done for the whole group of patients. The coefficient of correlation (r) was calculated for each correlation in all variables. The critical value for correlation was calculated in each correlation matrix. If the (r) for a certain variable is above the critical value, the correlation is statistically significant, and if the (r) below the critical value, the correlation is statistically insignificant. A positive correlation means that when one variable increases, the other variable also increases and vice versa. A negative correlation means that when one variable increases the other decreases and vice versa.

RESULTS

In group A, the age of cases was ranging between 3 – 6 months with mean age 4.85 ± 1.02 month; the weight was ranging between 4 – 7.5 kg with mean weight 4.93 ± 1.03 kg. While in group B, the age of cases was ranging between 6.5 – 11 months with mean age 7.47 ± 1.68 month, the weight was ranging between 6 – 8.5 kg with mean weight 6.87 ± 1.54 kg. Table (1)

The p value between the two groups was significant (p value < 0.05)

		Age (m)	Weight (kg)
A	Range	3 – 6	4 – 7.5
	Mean	4.85 ± 1.02	4.93 ± 1.03
B	Range	6.5 – 11	6 – 8.5
	Mean	7.47 ± 1.68	6.87 ± 1.54

Table 1: Age and weight of patients.

31 patients (77.5%) were with the features of Down syndrome in the group A, while they were 27 patients (67.5%) in group B.

According to Rastelli classification, in group A there were 31 patients (77.5%) with type A, 9 (22.5%) type B and no patient with type C. While in group B, they were 34 (85%) with type A, 6 patients (15%) with type B and no cases with type C.

Symptoms of patients in group A were dyspnea on effort (during feeding) which was 33 patients (82.5%), while dyspnea at rest was present in 2 patients (5%). Recurrent chest infection occurred in 33 patients (82.5%) and asymptomatic patients were 5 patients (12.5%). While in group B there was dyspnea on effort (during feeding) in 38 patients (95%), while dyspnea at rest was present in 2 patients (5%). Recurrent chest infection occurred in 37 patients (92.5%) and asymptomatic patients were 1 patient (2.5%). Table (2)

Symptoms	A		B	
	No.	%	No.	%
Dyspnea on effort (feeding)	33	82.5	38	95
Dyspnea on rest	2	5	2	5
Recurrent chest infection	33	82.5	37	92.5
Asymptomatic	5	12.5	1	2.5

Table 2: Symptoms of patients.

All patients were investigated with plain chest x-ray that showed increased cardiothoracic ratio in 100% of patients.

We diagnosed all patients with preoperative echocardiography that revealed:

In group A:

The range of ASD diameter was 12 – 18 mm with mean 13.24 ± 1.74 mm. The range of VSD diameter was 6 – 11 mm with mean 8.25 ± 1.53 mm. Systolic PAP ranges 45 – 80 mmHg with mean 61.00 ± 10.07 mmHg. The degree of right AV regurgitation was mild in 22 patients (55%), moderate in 13 patients (32.5%) & severe in 5 patients (12.5%). The degree of left AV regurgitation was mild in 15 patients (37.5%), moderate in 18 patients (45.0%) & severe in 7 patients (17.5%). Table (3)

In group B:

The range of ASD diameter was 11 – 19 mm with mean 12.02 ± 1.32 mm. The range of VSD diameter was 7 – 13 mm with mean 9.34 ± 1.89 mm. Systolic PAP ranges 60 – 90 mmHg with mean 74.21 ± 15.01 mmHg. The degree of right AV regurgitation was mild in 14 patients (35%), moderate in 18 patients (45%) & severe in 8 patients (20%). The degree of left AV regurgitation was mild in 8 patients (20%), moderate in 18 patients (45.0%) & severe in 15 patients (37.5%).

The p value between the two groups according to pulmonary artery pressure and AVV valve regurgitation was significant (p value < 0.05)

		A	B
ASD (mm)	Range	12 – 18	11 – 19
	Mean	13.24 ± 1.74	12.02 ± 1.32
VSD (mm)	Range	6 – 11	7 – 13
	Mean	8.25 ± 1.53	9.34 ± 1.89
PAP (mmHg)	Range	45 – 80	60 – 90
	Mean	61.00 ± 10.07	74.21 ± 15.01
Rt AV valve	Mild	22	14
	Moderate	13	18
	Sever	5	8
Lt AV valve	Mild	15	8
	Moderate	18	18
	Sever	7	15
Rastelli Classification	A	31	34
	B	9	6
	C	0	0

Table 3: Preoperative echocardiography data

In group A, the aortic clamp time was ranged between 45 – 75 minutes with mean 56.62 ± 9.29 minutes; the total cardiopulmonary bypass (CPB) time was ranged between 90 – 140 minutes with mean 112.87 ± 11.48 minutes. While group B, the aortic clamp time was ranged between 50 – 90 minutes with mean 62.64 ± 14.21 minutes; the total cardiopulmonary bypass (CPB) time was ranged between 90 – 160 minutes with mean 121.78 ± 18.21 minutes. Table (4)

		A	B
Aortic cross clamp time (min)	Range	45 – 75	50 – 90
	Mean	56.62 ± 9.29	62.64 ± 14.21
CPB time (min)	Range	90 – 140	90 – 160
	Mean	112.87 ± 11.48	121.78 ± 18.21

Table 4: Aortic cross clamp and CPB time

In patients repaired with double patch technique (17 patients), the VSD was closed with GorTex patch and the primum ASD was closed with autologous pericardial patch that may extend to close also the secundum ASD

In patients repaired with single patch technique (63 patients) the VSD and ASD components were closed with autologous pericardial patch.

Left AV valve was repaired with interrupted 5/0 prolene sutures in 76 patients and the other 4 patients were not repaired as they had double valve orifices.

During cardiopulmonary bypass conventional hemofiltration was done for all patients while modified ultrafiltration (MUF) was done only for 7 patients (2 patients in group A and 5 in the group B)

The inotropes and vasodilators used for the patients are described in Table (5). Milrinone was used for all patients at a dose 0.7 mic/kg/min

		A	B
Adrenaline (mcg/kg/min)	Range	50 – 140	100 – 220
	Mean	85.64 ± 29.18	140.54 ± 70.01
Noradrenaline (mcg/kg/min)	Range	50 – 120	100 – 150
	Mean	74.23 ± 27.30	120.32 ± 25.47
Dobutamine (mcg/kg/min)	Range	5 – 15	10 – 15
	Mean	8.80 ± 2.69	12.32 ± 2.54

Table 5: Postoperative inotropes and vasodilators.

		A	B	P value
Ventilation time (h)	Range	21 – 72	36– 92	0.002
	Mean	17.73 ± 8.21	44.26 ± 37.04	
ICU stay (day)	Range	2 – 10	6 – 15	0.012
	Mean	4.07 ± 2.22	9.32 ± 5.12	
Hospital stay (day)	Range	7 – 19	11 – 23	0.004
	Mean	10.16 ± 4.29	16.11 ± 7.54	

Table 6: ICU and hospital stay

In our study, we observed patients complicated with chest infection (three in group A and nine in group B) that was managed by antibiotics and chest physiotherapy, superficial wound infection (one in group A and one in group B) that was managed by frequent dressing and antibiotics, one case of surgical bleeding in group A that was transferred again in operating room for exploration, impaired renal function (two in group A and four in group B) that was managed by peritoneal dialysis, temporal heart block (five in group A and three in group B) that was managed with atropine and dexamethasone and recovered within 8 days, one case with permanent heart block in group B that needed permanent pace maker and pulmonary hypertensive crisis (two in group A and eight in group B) that was managed by sedation, muscle relaxant, hyperventilation, correction of electrolytes and pulmonary vasodilators drugs. Table (7)

In group A, the time need for ventilation ranged between 21– 72 hour with mean 17.73 ± 8.21 hour, the period of stay in the ICU ranged between 2 – 10 days with mean 4.07 ± 2.22 days and the total period of stay in the hospital ranged between 7 – 16 days with mean 8.16 ± 3.29 days.

While in group B, the time need for ventilation ranged between 36– 92 hour with mean 44.26 ± 37.04 hour, the period of stay in the ICU ranged between 6 – 15 days with mean 9.32 ± 5.12 days and the total period of stay in the hospital ranged between 11 – 23 days with mean 16.11 ± 7.54 days. Table (6)

The p value between the two groups according to ventilation time, ICU and hospital stay was significant (p value < 0.05)

Complications	A	B
Chest infection	3	9
Superficial wound infection	1	1
Bleeding	1	0
Impaired renal function	2	4
Temporal heart block	5	3
Permanent heart block	0	1
Pulmonary hypertensive crisis	2	8

Table 7: Postoperative complications

Postoperative echocardiography showed:

No cases with residual ASD and 4 cases (10%) with residual VSD ranging between 2- 4 mm in the first group, while in the second group there was no residual ASD or VSD.

In group A, there was 4 cases (10%) with mild tricuspid regurgitation, 8 cases (20%) with moderate tricuspid regurgitation, 9 cases (22.5%) with mild mitral regurgitation 4 cases (10%) with moderate mitral regurgitation and 4 cases (10%) with sever mitral regurgitation. But in group B we found 3 cases

(7.5%) with mild tricuspid regurgitation, 6 cases (15%) with moderate tricuspid regurgitation, 1 case (2.5%) with sever tricuspid regurgitation, 4 cases (10%) with mild mitral regurgitation, 2 cases (5%) with moderate mitral regurgitation and 2 cases (5%) with sever mitral regurgitation. Table (8)

The p value between the two groups was significant (p value < 0.05)

	Degree	A	B	P value
Tricuspid regurgitation	Mild	4	3	0.015
	Moderate	8	6	
	Sever	0	1	
Mitral regurgitation	Mild	9	4	0.008
	Moderate	4	2	
	Sever	4	2	

Table 8: Postoperative AV valve regurgitation

The range of postoperative systolic PAP was between 30 – 55 mmHg with mean 37.23 ± 2.57 mmHg in group A, while in group B the range was between 50 – 65 mmHg with mean 56.82 ± 6.29 mmHg. The p value between the two groups was significant (p value < 0.05)

In group A, the mortality rate was 4 cases (10%), 2 of them were due to low cardiac output, one due to pulmonary hypertensive crisis and one due to chest infection. While in group B, the mortality rate was 7 cases (17.5%), 2 of them due to chest infection and 5 due to pulmonary hypertensive crisis. The p value between the two groups was significant (p value < 0.05).

Kaplan–Meier curve for freedom from death comparing children before and after six months of age is shown in figure 1.

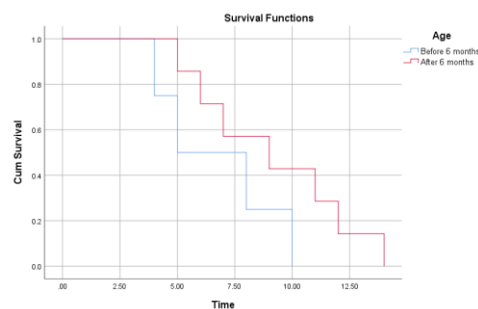


Fig. 1: Kaplan–Meier curves for freedom from death

Follow up:

All cases were normal as regard there hemodynamics, chest condition and wound scar.

No cases with residual ASD or VSD except for 1 case with residual VSD 3 mm in the first group.

In group A, 10 cases with moderate tricuspid regurgitation, 1 case with sever tricuspid regurgitation, 5 cases with mild mitral regurgitation, 8 cases with moderate mitral regurgitation and 7

cases with sever mitral regurgitation (3 of them need surgical intervention for mital repair) and pulmonary artery pressure ranged between 15 – 35 mmHg with mean 20.69 ± 5.49 mmHg.

While in group B, 5 cases with tricuspid regurgitation, 5 cases with mild mitral regurgitation, 4 cases with moderate mitral regurgitation and 2 cases with sever mitral regurgitation that need follow up with medical treatment and pulmonary artery pressure ranged between 25 – 40 mmHg with mean 28.59 ± 9.83 mmHg.

DISCUSSION

The improvement in the operative techniques and ICU management played a great role in decreasing the rate of morbidity and mortality.⁸

Early repair decrease the risk of development of pulmonary vascular obstructive disease. It is preferred in patients with refractory heart failure that not respond to medical treatment.⁹

Historically, CAVSD was successfully repaired by C. Walton Lillehei for the first time in 1955.¹⁰

There is debate about the timing of repair of CAVSD in spite of the benefits of early repair before the age of six months that do not give the same results in all patients⁹. In our study, we operated on 80 patients with age ranged between 3 - 11 months (mean 6.45 ± 2.17 month), while Dawary et al operated on 140 patients with age below 2 years¹¹, while Atz et al operated on 120 patients ranging between 9 days to 1.1 years (median age at surgery was 3.7 months).¹²

This series of patients didn't include cases of complete atrio-ventricular canal associated with repairable congenital cardiac anomalies, unbalanced complete atrio-ventricular canal and redo cases.

We studied all data about pre-operative, intra-operative and post-operative course that affect prognosis of the repair. Full analysis of immediate and early post-operative results was done.

Patients with Down syndrome have higher incidence of morbidity rate that affect the duration of ICU stay, hospital stay and the expected life style even after excellent repair.¹³

In our study 58 patients are Down syndrome that represents 72.5 % of patients, while it was 80% in the study of Atz et al¹². In Deraz et al it was 89.6 % of patients.¹⁴

Several studies confirmed that there is a relationship between the weight and the surgical result. Waiting for gaining weight may be risky as the clinical status of the patient need urgent surgical intervention. These patients are complaining of congestive heart failure and pulmonary vascular obstructive disease that they cannot coexist with it. In a study done by Prifti and colleagues¹⁵ about the relation between weight and the surgical result, they noted high incidence of reoperation for left AV valve regurgitation if the weight at time of repair was < 5 kg. Suzuki and colleagues noted high rate of left AV valve regurgitation if repair was done before age of 3 months.¹⁶

Many studies revealed that primary repair of CAVSD at young age are associated with poor outcomes. Others preferred the age of 3 – 6 months as the ideal timing for intervention.¹⁷

The used surgical technique in our study was modified single patch technique in 63 patients that represents 78.75 % and double patch technique in 17 patients that represents 21.25 %. Whie Ugaki et al used 29 children (56.8 %) operated with modified single-patch technique and 22 patients with double patch technique (43.2%)¹⁸. Abdel-Raouf et al used single patch technique in 30 patients (60%) and double patch technique in 20 patients (40%).¹⁹

In our study there was 4 cases (5%) with residual VSD ranging between 2- 4 mm that are in the first group that revealed also 17 cases mitral regurgitation [9 cases with mild mitral regurgitation 4 cases with moderate mitral regurgitation and 4 cases with sever mitral regurgitation], while the second revealed 8 cases mitral regurgitation [4 cases with mild mitral regurgitation 2 cases with moderate mitral regurgitation and 2 cases with sever mitral regurgitation] Dawary et al found 33 patients with mitral regurgitation [23 cases (16.4%) with mild AV regurgitation, 7 (5%) with moderate and 3 (2.1%) with severe AV regurgitation]¹¹. Atz et al found 56 (48%) with residual VSD, 59 patients (49.1%) with mitral regurgitation (12). Deraz et al found 5 patients (3.4%) with residual VSD, 12 patients (8.2%) with sever mitral regurgitation¹⁴. Ugaki et al found 4 patients (7.8%) with residual VSD and 6 patients with mitral regurgitation¹⁸. Abdel-Raouf et al found 8 patients (16%) with moderate mitral regurgitation and 2 patients (4%) with sever mitral regurgitation.¹⁹

In our first group we had 5 cases (12.5%) with temporal heart block that recovered within 8 days

and none of them needed permanent pace maker, while in the second group we had 4 cases with heart block but only one of them needed insertion of permanent pace maker in comparison to Deraz et al that found 5 patients (3.4%) with complete heart block that needed placement of a permanent pace maker¹⁴. Only 1 patient needed permanent pace maker according to Ugaki et al¹⁸. Pan et al also reported 2 patients (2.04%) developed heart block that required a pacemaker insertion²⁰. Abdel-Raouf et al reported 2 cases (4%) with transient heart block which resolved well with no need for pacemaker implantation.¹⁹

In comparison to Atz et al who reported 2.5% (3 cases) mortality¹², we had 13.75% (11 cases: 4 in the first group and 7 in the second group) mortality. Deraz et al reported 6.2% (9 cases)¹⁴, while Abdel-Raouf et al reported 6 % (3 cases).¹⁹

CONCLUSION

Repair of CAVC in the first group give better results in improvement of the pulmonary artery pressure with higher incidence of reoperation for Lt AV valve repair.

While repair in the second group give bad results in improvement of the pulmonary artery pressure with better results for the Lt AV valve.

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