

Outcomes of surgical repair of complete atrioventricular canal defect in patients younger than 2 years of age

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BACKGROUND: Early surgical management of complete atrioventricular (AV) canal defect is the optimal treatment option. Since the published evidence on outcomes is inconclusive, we retrospectively studied the outcomes of patients in our institution.

OBJECTIVE: Study outcomes of complete AV canal repair.

DESIGN: Retrospective, descriptive.

SETTINGS: Single institute.

PATIENTS AND METHODS: Medical records of patients under 2 years of age who underwent complete AV canal repair from January 2004 to December 2014 were retrospectively reviewed.

MAIN OUTCOME MEASURES: Pre- and postoperative morbidity and mortality.

SAMPLE SIZE: 140 patients.

RESULT: The median (IQR) age at the time of surgery was 5.4 (3.9-8.2) months. Down syndrome was diagnosed in 98 (70%) of patients. AV valve regurgitation was found preoperatively in 129 (92%) and postoperatively in 135 (96%) patients. There was a significant association between preoperative pulmonary hypertension and the development of pulmonary hypertension in the postoperative period ($P=.04$). Thirty-three patients needed reoperation. Arrhythmia was found in 19 patients, 16 of whom required pacemaker insertion. Seven patients died (5%).

CONCLUSION: The presence of preoperative and postoperative AV valve regurgitation was common in this cohort but did not significantly affect patient survival. Our findings suggest an acceptable outcome for repair of complete AV septal defect with few complications postoperatively.

LIMITATION: Retrospective in single institute.

CONFLICT OF INTEREST: None.

Atrioventricular (AV) canal defects arise from defective development of endocardial cushions during the prenatal period. AV defects involve the atrial septum and the ventricular septum as well as both AV valves. It is estimated that this anomaly affects 2 of every 10000 live births and accounts for 3% of all cardiac malformations.¹ Management is primarily surgical for most patients since the first successful repair of a complete AV septal defect in 1954.² Outcomes depend on the presence of preoperative risk factors and the age of the child at the time of repair. Patients with a preexisting dysplastic left AV valve or a coexisting coarctation or double orifice AV valve have a high hospital mortality rate.^{3,4} Some studies have shown no significant differences in the outcome of surgery between normal children and those with a combined birth defect such as Down syndrome.⁴ Nowadays, it is possible to correct the defect during early infancy to prevent progressive pulmonary vascular disease.⁵ Studies have shown similar outcomes in patients younger and older than 3 months of age.⁶ Furthermore, the reoperation rate is reduced in younger patients.⁷ However, although the mortality rate has declined in the past decade, reoperation rates have not changed.⁸ The leading causes of death in AV canal repair are pulmonary hypertensive crisis, acute ventricular failure, sepsis, arrhythmia and multisystem organ failure.⁹ Since the published evidence is inconclusive, we retrospectively studied the outcomes of patients under the age of 2 years who underwent complete AV canal repair from 2004 to 2014.

PATIENTS AND METHODS

We reviewed the medical records of all pediatric patients younger than two years of age who underwent complete AV canal repair from January 2004 to December 2014 at King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia. Patients were identified through the department database. The research ethics committee at our institute approved the study. The statistical analysis of data for this study was done using SAS version 9.4 software package (SAS Institute Inc., Cary, NC, USA). Descriptive statistics for all the continuous variables are reported as mean and standard deviation or median and interquartile range (IQR) while categorical variables are reported as frequencies and percentages. The categorical variables were compared by using chi-square test, while continuous variables were compared by t test and ANOVA. The statistical level of significance was set at $P < .05$. Since the quantitative aspects of most of the clinical outcomes of patients who underwent surgery for complete AV canal repair are unknown, it is important to

report these outcomes with certain levels of accuracy. We used a margin of error in estimation of 5% with a 95% confidence interval.

RESULTS

We identified 140 patients younger than two years of age who were diagnosed with complete AV canal defect and underwent repair during the period from January 2004 until December 2014. Ninety-one of the patients (65%) were males. Of 140 patients, 98 patients (70%) were diagnosed with Down syndrome while only 3 patients (2%) had tetralogy of Fallot (**Table 1**). Median (IQR) age at the time of surgery was 5.4 (3.9-8.2) months (**Figure 1**) with a mean (SD) weight of 4.7 (1.7) kg.

Preoperatively, 129 patients (92%) had AV valve regurgitation and 19 (14%) had pulmonary hypertension (**Table 1**). The double-patch surgical technique was used in repair in the majority of patients ($n=93$, 67%), while single-patch single patch was used in 40 patients (29%). A modified single patch was used in only 6 patients (4%) and in one patient we could not determine the repair type. In the postoperative period, AV valve regurgitation was found in 135 patients (96%) with the majority in both valves while only 3 patients developed AV valve stenosis. Pulmonary hypertension was found in 11 patients (14%) preoperatively and in 9 patients (6%) during the early postoperative period. There was a significant association between preoperative pulmonary hypertension and the development of pulmonary hypertension in the postoperative period ($P=.04$) (**Table 2**). Seven patients (5%) died with a mean follow-up period of 4 years; 2 died within one month after surgery. Thirty-three patients (24%) underwent reoperation; 23 were diagnosed with mild AV regurgitation, 7 with moderate and 3 with severe AV regurgitation after the first surgery. Among the patients who underwent reoperation (33 patients), there were significantly lower reoperation rates among Down patients versus non-

Table 1. Preoperative characteristics of the 140 patients.

Variable	N
Down syndrome	98 (70)
Tetralogy of Fallot	3 (2)
Atrioventricular valve regurgitation (pre-operative)	129 (92)
Pulmonary hypertension	19 (14)
Parachute mitral valve	6 (4)

Data are number (%).

Down patients (18.37 vs 35.71 $P=.032$). Forty-seven patients (34%) were readmitted for different reasons during the follow up period. Postoperative arrhythmia was found in 19 patients (14%) and 16 (11%) of those required pacemaker insertion (Table 3). There were no differences in pacemaker requirements by surgical technique.

DISCUSSION

Complete AV septal defect (CAVSD) is considered one of the complex heart anomalies that is characterized by variable defects including both atria and ventricles, the septum and the AV valves. CAVSD accounts for about 3% of all cardiac anomalies.¹ Rastelli's classification of CAVSD (Figure 2) includes three types according to the insertion of the chordae and the structure of the superior bridging leaflet of the common AV valve. Surgical management of these complex anomalies can be achieved through different procedures that include using a single patch, double patch, or a modified single patch technique. In this study we analyzed 140 patients who underwent CAVSD repair. Our study did not show any significant association between the patient gender and postoperative valve competency, regurgitation or stenosis. The same results were found in the literature.³ Down syndrome is considered one of the most common genetic disorders that is associated with CAVSD;¹ 70% of our patients were diagnosed with Down syndrome. Many patients had AV regurgitation postoperatively, but we found no significant association between Down syndrome and postoperative AV regurgitation. The significantly lower reoperation rates among Down patients versus non-Down patients was most likely because Down patients have more redundant tissues than non-Down patients, which helps with the repair. Masuda et al reported 64 infants with CAVSD who underwent repair; 34 were diagnosed with Down syndrome. The mean (SD) follow-up period of all patients was 99 (47) months. Masuda concluded that Down syndrome does not affect the long-term results of CAVSD when the defect is repaired during the first year of life.¹⁰ Rizzoli et al reported 87 patients with CAVSD with 74% diagnosed with Down syndrome who underwent repair. Rizzoli concluded that Down syndrome was not an independent risk factor for operative mortality, nor reoperation.¹¹

Double-patch repair was used in the majority of our patients (67%). The type of repair technique did not show any significant differences in valve regurgitation and severity of regurgitation postoperatively. Follow-up data indicate the same mortality rates in single- versus double-patch technique. Pan et al reported on CAVSD repair in 98 infants in which 46 patients underwent modified single-patch repair while 59 patients underwent double patch repair.¹² No significant difference was found between the two techniques in terms of mortality, postoperative AV valve regurgitation, left ventricular outflow tract obstruction or residual ventricular septal defect except for a shorter cross clamp time for the modified single-patch group. Pan et al also reported that two patients developed heart block that required a

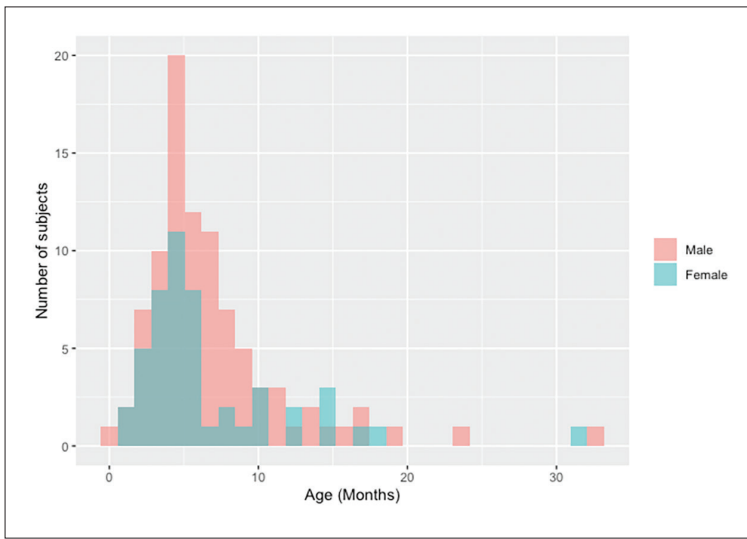


Figure 1. Age distribution of the 140 patients.

Table 2. Postoperative findings.

Variable	Number of patients
Atrioventricular regurgitation	135 (96)
Normal pulmonary artery pressure	131 (94)
Reoperation	33 (24)
Readmission	47 (34)

Data are number (%).

Table 3. Postoperative complications in the 140 patients.

Variable	Our findings	Reported finding
Renal failure	4 (2.8)	5/121 (7.6) ¹³
Pulmonary hypertensive crisis	11 (7.9)	5/121 (7.6) ¹³
Absence of sinus rhythm	19 (13.6)	17 /121(25.8) ¹³
Infection	32 (22.9)	6/121 (9.1) ¹³
Pacemaker insertion	16 (11.4)	11/ 172 (6.4) ³

Data are number (%).

pacemaker insertion but there was no statistically significant difference between the two techniques.

We found that there was a significant association between preoperative pulmonary hypertension and the development of pulmonary hypertension in the postoperative period ($P=.04$). In our study, 129 patients (92%) were diagnosed with AV regurgitation while 135 patients (96%) were diagnosed with valve regurgitation despite the preoperative condition of the valve. The preoperative condition of the AV valve (competent, regurgitation, stenosis) did not have any significant impact on the postoperative valve competency or patient survival. The presence of regurgitation in the left or right or both valves had no effect on reoperation rate. Nineteen patients (14%) developed postoperative arrhythmia, 16 patients (11%) of whom required pacemaker insertion. Furthermore, no statistically significant association was found between the patch used during the operation and pacemaker insertion. Gunther et al reported 16 patients out of 320 who needed pacemaker insertions while Crawford et al reported 11 patients out of 172.^{3,8} We reported 32 patients (23%) who developed infectious complications with prolonged use of antibiotics while 11 of our patients (8%) developed neurologic complications that required a neurocognitive CT scan for diagnosis. Gunther et al reported 52 patients out of 274 in which 10 patients died from infectious complications

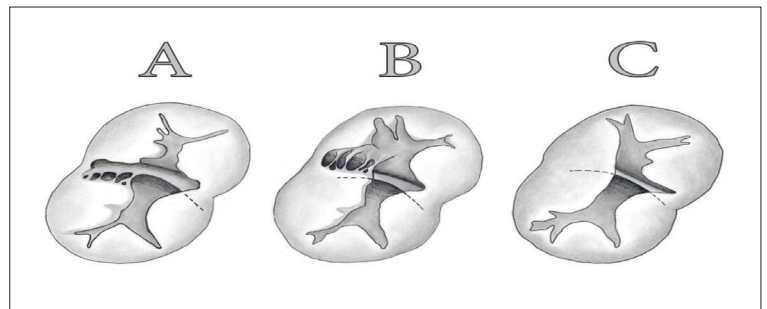


Figure 2. Rastelli's classification of complete AV septal defect.

while 1 only died from stroke.³

In conclusion, we confirmed the results of previous studies regarding the association between complete AV septal defect and Down syndrome. The presence of preoperative and postoperative AV valve regurgitation was common in this cohort but did not significantly affect patient survival. The majority of our patients were treated with the double-patch technique. The existence of preoperative pulmonary hypertension correlated with the presence of postoperative pulmonary hypertension. Our findings suggest an acceptable outcome for complete AV septal defect with few complications postoperatively which is consistent with other findings.¹³ Limitation is that the study was retrospective and from a single institution.

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