Postoperative cardiac intensive care outcome for Down syndrome children

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ABSTRACT

Objective: The purpose of this study is to review the postoperative intensive care unit (ICU) course, complications and outcome of Down's syndrome children undergoing surgical repair of congenital heart diseases (CHD).

Methods: A retrospective chart review analysis of intensive care course and outcome of Down syndrome children undergoing surgical repair of congenital heart diseases (CHD) from May 2000 to May 2004. The study was conducted in the Pediatric Cardiac ICU, King Abdul-Aziz Cardiac Center of National Guard Hospital, Riyadh, Kingdom of Saudi Arabia. All Down's syndrome children who had surgical repair during the study period were included.

Results: During the study period, 80 patients (31 males and 49 females) with Down's syndrome had surgical repair of CHD. Their average weight was 5.8±0.3 kg and age was 11.7±1.5 months. Their primary surgical interventions were as follow: atrio-ventricular septal defect repair (44), ventricular septal defect closure (24), patent ductus arteriosus ligation (6), secundum atrial septal defect closure (3), and tetralogy of Fallot repair (3). Postoperative complications occurred in 19 patients (23%). Few patients had more than one complication. The compilations were as follow: 8 patients (10%) had sepsis, 3 (4%) required permanent pacemaker insertion, 4 (5%) had chylothorax, 2 (2.5%) had life threatening pulmonary hypertensive crisis with full recovery after proper managements, 5 (6%) needed prolong intubation, more than 7 days, and one patient (1.2%) required tracheostomy. All patients survived and were discharged home except one (1.2%) who expired 8 weeks after surgery from sepsis and multi-organ failure.

Conclusion: Patients with Down's syndrome undergoing CHD repair had an acceptable postoperative morbidity and low mortality. Their results are comparable to non-Down's cardiac patients. From an ICU perspective, the majority of these patients do well postoperatively with good ICU outcome.

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Congenital heart defects (CHD) are present in 30-50% of children with Down's syndrome (DS). The most common types of CHD in these patients are atrioventricular septal defects (AVSD), ventricular septal defects (VSD), secundum atrial septal defect (ASD), patent ductus arteriosus (PDA) and tetralogy of Fallot (TOF).[1] In the majority of cases, the presence of cardiac lesion has significant effects on the child's life leading to the development of cardio-respiratory symptoms, increasing the infection risk, and increasing the need for repeated hospitalizations.[2] In spite of the obvious needs to repair the cardiac defects in DS children, these patients have not always reached the optimal surgical correction they need, or in some instance they were offered less equal care in comparison to other non-DS children.[3,4] The reasons for this disparity in the management of DS children are

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multi-factorial and may have medical, socio-economical, or even ethical backgrounds. From an intensive care perspective, DS children were frequently labeled as high-risk patients, particularly due to the morbidity and mortality during the perioperative period of the cardiac surgery.[5] This in return has influenced the decision to refer DS patients to surgery or in some instances has lead to delay or even disqualify them completely from surgical correction. In our era of modern medicine, many changes have occurred in the intensive care unit (ICU) management of sick children including new modalities of mechanical ventilation, usage of new systemic and selective vasodilators and invasive and non-invasive support systems. This impressive development in critical care, cardiac surgery and cardiology fields has improved the ICU outcome following cardiac surgery in general and of CHD in particular. Many recent reports reflected the improvement in the ICU course and outcome following the repair of many cardiac lesions. However, the information available regarding the ICU outcome following CHD repair in DS patients is still limited and often does not reflect the contemporary results. In order to have a current assessment of the preoperative risk, complications and ICU outcome following cardiac surgery in DS patients, we conducted a retrospective chart review and database analysis of all DS children who underwent surgical repair of CHD in the last 4 years in our institution. Our specific aims are to determine the ICU course, postoperative complications and ICU outcome of DS children undergoing surgical correction of CHD.

Methods. A retrospective chart review and database analysis was performed on all DS patients who underwent surgical repair of CHD between May 2000 and May 2004 at King Abdul-Aziz Medical City, Riyadh, Kingdom of Saudi Arabia. Our medical facility receives referral patients from all areas of Saudi Arabia and the surrounding Middle Eastern countries. During the initial 2 years of the study period, all postoperative cardiac patients were admitted to 11 beds in the general Pediatric ICU. Starting from January 2002 and thereafter, all our postoperative cardiac patients were admitted to a separate 8 bedded pediatric cardiac ICU. A team of 2 full-time pediatric cardiac intensivists consultants and one part-time general pediatric intensivist consultant covers the pediatric cardiac unit. Additionally, the pediatric cardiac ICU has 24 hours coverage by dedicated junior staff that are either pediatric cardiac fellows, pediatric ICU fellows, or a qualified pediatrician trained in the management of critical cardiac patients. Medical information about the patients was obtained from the database and from the medical records of the patients. Diagnosis of DS was made on the basis of clinical examination and genetic testing. The presence and the type of heart lesions were established on the basis of clinical assessment, electrocardiography, echocardiography, radiology and hemodynamic studies. Patients were followed until they were discharged from the ICU. Demographic data including age, gender, weight at the time of surgery and data regarding diagnosis, type of surgical repair, bypass time, cross clamp time, ventilation hours, maximum number of inotropes, maximum duration of inotropes infusion, and number of extubation trial, postoperative complications and mortality were recorded. Pre-existing medical conditions that might affect surgery or postoperative course and outcome were also identified. Perioperative complications were defined as events or conditions that were not present before surgery and occurred as a result of anesthesia, surgery or during postoperative ICU care. All major complications that have consequences on the patient health, affecting the ICU course, or outcome were identified. Any death occurred during the postoperative ICU course or within 30 days following the surgery was reported as mortality.

Results. During the study period 80 children (31 males and 49 females) with DS had surgical repair of CHD. Their weight ranged between 2.3 and 23 kg with a mean of 5.8±0.3 kg. Their age ranged between 0.33 month and 86 months with a mean 11.7±1.5 months. Classification of the cardiac lesions with their percentage is summarized in **Table 1.** In 15 cases (18%), there were pre-existing health problems prior to surgery, primarily of respiratory origin in 9 patients (respiratory failure, respiratory infection, reactive airway disease, tracheomalacia), 4 patients had congenital malformation of bowel that required colostomy early in life. Additionally, sepsis occurred in 4 cases prior to surgery. In lesions associated with left to right shunting, different degree of pulmonary hypertension was observed pre-operatively. This pulmonary hypertension was judged clinically or by hemodynamic catheter measurement (in 4 cases [5%]) to be reversible before surgery. Severe pulmonary hypertension with irreversible pulmonary vascular disease unresponsive to either 100% oxygen supplement or other pulmonary vasodilators such as nitric oxide was found in a small number of cases referred to our institute. They were declared inoperable and were disqualified for entry to our pool of patients included in the study. The primary surgical repair performed in the 80 patients were as follows: 44 AVSD repair (54%), 24 VSD closure (30%), 6 PDA ligation (8%), 3 secundum ASD closure (4%), 3 TOF repair (4%). Two patients had redo mitral valve repair for significant residual mitral regurgitation.

Measurements of different variables related to surgery and postoperative ICU course are summarized in **Table 2**. All patients who had open heat surgery had central venous and left atrial pressure monitoring lines at the time of arrival to ICU postoperatively. Pulmonary arterial catheter was not routinely inserted intraoperatively and only in exceptional cases if the patient's pulmonary pressure remained high at the end of surgery. Postoperative complications occurred in 19 patients (23%). The average age of the patients with complication was 14±4 months. Based on their age at the time of surgery, we divided the patients with complications into 3 groups. **Table 3** summarizes the number of patients and the incidence of complication in each age-group category. The complications that were encountered were as follows: 8 patients (10%) had sepsis mainly caused by *Staphylococcus species*; 3 patient (4%) required permanent pacemaker insertion for complete heart block; 4 patients (5%) had chylothorax, managed conservatively; 2 patients (2.5%) known to have significant pulmonary hypertension before surgery developed postoperatively life threatening pulmonary hypertensive crisis. Their pulmonary arterial pressure was postoperatively equal to 2/3 the systemic arterial pressure. In one of those 2 patients, the pulmonary hypertensive crisis lead to cardio-respiratory arrest from severe pulmonary hypertensive crisis and was successfully resuscitated; 5 patients (6%) remained intubated and ventilated more than 7 days post surgery and then they were all extubated successfully except one (1.2%) who needed tracheostomy. All patients survived and were discharged home except one (1.2%) late death, 8 weeks after surgery from sepsis and multi-organ failure.

Discussion. Down's syndrome (Trisomy 21) is the most common chromosomal abnormality in childhood. The incidence of this syndrome is 1:800 live births.[1] Congenital heart defects form important features of this syndrome, present in 30-50% of the patients. The common cardiac defects in DS are AVSD, VSD, secundum ASD, and PDA. Most of these cardiac lesions lead to significant left to right shunt with the development of congestive heart failure, increased pulmonary blood flow, recurrent respiratory infections and failure to thrive. If the CHD is left without repair, the disease will progress to severe heart failure or pulmonary hypertension which tends to have an accelerated and violent course in Down's patients.[1] The consequences of CHD on DS children are manifested by increase in morbidity, mortality and shortening of their life span when compared to those with DS and normal heart.[2] Although repairing the cardiac defect in DS patients is frequently indicated early in life, corrective surgery is not always achieved. Many factors contribute to the delay or even withholding

Table 1 - Type of cardiac defects in 80 patients with Down's syndrome.

Cardiac lesions	Classification	n	(%)
AVSD	Complete	44	(54)
VSD	VSD	24	(30)
PDA	Isolated	6	(8)
ASDII	Isolated	3	(4)
TOF	Isolated	3	(4)

AVSD - atrioventricular septal defect, VSD - ventricular septal defect, PDA - patent ductus arteriosus, ASD - atrial septal defect, TOF - tetralogy of fallot

Table 2 - Summary of demographic, surgical and ICU data related to the 80 patients with Down syndrome who underwent surgical repair of congenital heart diseases. Data are presented as mean ± standard error of the mean (SEM).

Demographic data	Mean±SEM
Male to female ratio	31:49
Age (months)	11.7± 1.5
Weight (kg)	5.8± 0.3
Length of ICU stay (days)	6.8±1.2
Length of hospital stay (days)	20±3
Bypass time (minutes)	78± 4
Cross-clamp time (minutes)	53 ± 2.8
Mechanical ventilation needed during PO (hours)	70 ± 19
Extubation trial (hours)	1.2 ± 0.06
Maximum number of inotropes	1.5 ± 0.1
Maximum duration of inotropes infusion (hours)	43 ± 8
N of patients with more than one extubation trial	11

PO - postoperative, ICU - intensive care unit

Table 3 - Summary of patient's distribution into 3 categories according to age and incidence of complication in each age-group category.

Age groups (months)	N of patients who underwent cardiac surgery	N of patients with complication n (%)
0-6	37	9 (24)
7-12	29	6 (21)
>12	14	4 (28)

of this surgical option in these patients. While some of these factors are indigenously related to the natural history of the disease and the presence of genetic abnormalities, others are related to the risk associated with the surgery and postoperative care. Early reports that were published between 1980 to 1990 on the surgical outcome in some of the Down patients particularly following atrioventricular canal repair were not very favorable. That led some authors to question the benefits and the wisdom of carrying out major cardiac surgery in such patients since it added no substantial advantages to the medical management alone.[6] Furthermore, a few published studies in the early 1990 suggested that, the presence of DS affects negatively the surgical results by increasing the postoperative complications, morbidity and mortality.[5,7] In view of such reports and similar studies, a general impression emerged that DS patients "do not do well" after cardiac surgery and as a result many health care providers were reluctant to refer or carry out surgical cardiac correction in DS patients.

In the last 15 years, there has been a gradual improvement in the results of DS children undergoing cardiac surgery. In a review published in 1989, Baciewicz et al[8] reported 55 patients with DS with (16.4%) 30 days mortality following surgical cardiac repair. In 1990 Sheehan et al[9] reported 42 children with DS with overall postoperative mortality of 9%. In 1999 Malec et al[1] reported 100 patients with DS who underwent surgical repair with 38% morbidity and 6% ICU mortality. Furthermore, Parvathy et al[10] reported in a study published in 2000, 22 DS patient who underwent cardiac surgery with no early mortality, 28.5% postoperative complications and 9.5% late death. Also in 2000, Kashima et al[11] reported 48 DS patients who had VSD repair, with postoperative mortality of 2%. In our series of 80 patients with DS, we observed nil mortality in the first 30 days after surgery and only one late death from sepsis 8 weeks following VSD closure. Postoperative complications were observed in 19 of 80 patients (23%). The most common were infection and respiratory complications. Sepsis occurred in 8 patients (10%) mainly caused by *Staphylococcus* and *Pseudomonas* organisms. In 7/8 cases, it occurred early in the postoperative period and in one case, sepsis developed late and led to death. Five patients (6%) required prolonged ventilator support (>7 days) and in one case (1.2%), the patient required tracheostomy. Pulmonary hypertensive crisis occurred in 2 cases (2.4%) and led to cardio-respiratory arrest in one of them. They were managed with sedation, muscle relaxation, mild hyperventilation, electrolytes corrections, and nitric oxide inhalation with subsequent full recovery. Chylothorax occurred in 4 patients (5%) that were cured by conservative management. Complete atrioventricular block occurred in 3 (4%) patients postoperatively and required permanent pacemaker insertion in all. The overall incidence of post-operative complications in our series was 23%. The mean ventilation hours and ICU length of stay postoperatively were 70 $\pm$ 19 hours and 6.8 $\pm$ 1.2 days. Due to the unique characteristic features of DS patients, it was difficult to match our patients with similar control non-Down patients. The majority of the ICU complications seen in our DS patients were manageable with nearly 100% ICU survival. The 30 days nil mortality in our DS cases were similar to the nil mortality seen in all non-Down VSD pediatric cases (n=60) that underwent surgical repair in our hospital during the same period of the study.

In conclusion, ICU morbidity and mortality in DS children undergoing cardiac repair have improved significantly in the last 10 years, attributed to advances in surgical techniques and ICU management strategies. Most DS patients do well and are discharged with ordinary ICU course following surgical repair of CHD. Postoperative complications and poor ICU outcome should no longer be considered as a reason for withholding or delaying repair in DS children. Positive medical attitude and unequivocal equal opportunity of care should be granted for all children with DS.

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