Original Article

A Unique Milieu for Perioperative Care of Adult Congenital Heart Disease Patients at a Single Institution

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Purpose: Adult patients with congenital heart disease presenting for cardiothoracic operation pose special demands for the arrangement of their perioperative care. This study describes the distinctive perioperative setup for adult congenital heart patients.

Materials and Methods: A retrospective review of 48 adult congenital heart disease cases that received operative intervention from July 2000 to October 2005. Operative procedures and postoperative care were all exercised in the same unit by the same staff handling adult and pediatric cardiothoracic surgery cases.

Results: Forty-two patients underwent definitive biventricular repair, and the remainder were subjected to palliative procedures. There were 2 cases of operative mortality: 1 patient died as a result of arrhythmia after Glenn procedure, and 1 died as a result of multiorgan failure secondary to sepsis after ventricular septal defect repair. Complications that prolonged intensive care stay for more than 72 hours were extracorporeal support for systemic right ventricular dysfunction (n = 1); reoperation for ventricular septal patch endocarditis after ventricular septal defect repair (n = 1); respiratory failure (n = 4); renal failure (n = 3); sepsis (n = 2); junctional tachycardia (n = 1); pulmonary hypertension (n = 2); and reperfusion injury of the lung (n = 2). Patients subjected to the Fontan operation had a longer hospital stay (mean = 33 days) as a result of pleural effusions. Forty-six patients were discharged home.

Conclusion: This distinctive environment allows our group to provide the appropriate care for our adult congenital heart disease patients in a well-integrated discipline. The frequent clinical exposure to both adult and pediatric cardiothoracic procedures has helped in facilitating the provision of optimum care to the patients with adult congenital heart disease. (Ann Thorac Cardiovasc Surg 2009; 15: 150–154)

Key words: perioperative care, adult, congenital heart

Introduction

The number of adult congenital heart disease patients is continuously increasing worldwide. Moreover, advances in the surgical and medical management of patients with congenital heart disease have resulted in increased survival to adulthood.[1] Where these patients should receive care is also a problem. It can be difficult for adults to

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Table 1. Surgical procedure list

	Diagnosis	Number
Definitive	Tetralogy of Fallot repair	6
	Rastelli's repair for tetralogy of Fallot	2
	Rastelli's repair for DORV & PS	1
	Subaortic membrane resection	4
	Coarctation repair	2
	Atrial septal defect closure	8
	Partial anomalous pulmonary venous drainage repair	3
	Partial atrioventricular canal defect repair	4
	Ventricular septal defect closure	9
	Ventricular septal defect, sinus of Valsalva fistula, and aortic incompetence repair	2
	Left atrioventricular valve regurgitation and regurgitant pulmonic conduit in corrected transposition of great arteries	1
Palliative	Bidirectional Glenn shunt	1
	Bilateral bidirectional Glenn shunt	1
	Fontan connection	3
	Insertion of epicardial automated defibrillator in Fontan	1

DORV & PS, double outlet right ventricle and pulmonary stenosis.

receive appropriate care in a pediatric setting such as a children's hospital.[2] The special needs of adults with congenital heart disease are also not well addressed by adult institutions where coronary artery disease dominates. Thus adult congenital heart disease patients who have undergone cardiothoracic surgery present a management challenge in the perioperative period.[3] Many groups have expressed their concerns regarding the best hospital setup that will provide well-integrated care to the adult congenital heart disease patient.[2,4]

Our institution, like most of the tertiary-care centers in Saudi Arabia, serves both adult and pediatric patient populations in the same facility. Fourteen years of age and above is the age definition for the adult patient population at our institution. With the exception of a few major disciplines, including oncology, anesthesiology, neuroscience, and cardiovascular diseases, all general specialties and subspecialties are grouped under a main specialty title, such as pediatrics, surgery, and internal medicine. The arrangement was designed in this fashion to maximize the utilization of resources and to strengthen the objectives of the tertiary-care services.

The Department of Cardiovascular Diseases consists of several divisions and directorships: Cardiothoracic Surgery, Adult Cardiology, Pediatric Cardiology, Cardiac Surgery Intensive Care Unit, and Invasive and Noninvasive Cardiac Laboratories. Referred cases of cardiovascular pathology are screened and filtered through the on-call cardiologist and channeled to the proper subspecialty in the department. The assessment and medical management of each adult congenital heart disease case is handled by joint care from pediatric cardiology and adult cardiology. Each case presents to the adult congenital heart disease forum, thus complementing this joint care and crystallizing the plan of management. Two actively involved cardiothoracic surgeons in both adult and pediatric cardiothoracic practice and who are experienced in cardiopulmonary transplantation are participants in the adult congenital heart disease forum.

The purpose of this review is to describe the operative outcome and perioperative course, utilizing a distinctive setup for a cohort of adult congenital heart disease patients who have undergone various cardiothoracic procedures at our institution.

Materials and Methods

From July 2000 to October 2005, 48 adult congenital heart disease cases received operative intervention at our institution. Table 1 demonstrates the surgical list of such patients who received cardiothoracic operative procedures.

All patients were admitted for elective surgeries except for 1 urgent case of tetralogy of Fallot repair resulting from severe cyanosis and 1 emergent repair of aortic dissection resulting from aortic balloon dilatation for coarctation of the aorta. Preoperatively, patients were evaluated by members of the anesthesiology staff, who are involved in the care of both adult and pediatric cardiothoracic surgery cases. Preoperative teaching was provided by members of the nursing staff, responsible for all adult and pediatric cardiac patients in the telemetry cardiothoracic surgery unit.

Operative procedures were carried out by members who are actively involved in both adult and pediatric cardiothoracic surgery and also cardiopulmonary transplantation. These personnel include operating room nurses, perfusionists, technologists, anesthesiologists, and cardiothoracic surgeons. The operating room is equipped with all items needed for both adult and pediatric cardiothoracic surgeries. Nursing personnel in the operating room are familiar with adult and pediatric size patients and are continuously exposed to both types of patient procedures. In general, 1 adult anesthesiologist is involved in the perioperative care of uncomplicated cases, such as atrial septal defect, subaortic membrane, and valvular diseases; otherwise, 2 anesthesiologists are involved in each case: 1 from adult cardiac subspecialty and 1 from pediatric congenital heart disease background. Monitoring lines, parameters, and vasoactive medications are implemented according to the surgical diagnosis and treatment. Arterial pressure and central venous pressure lines are placed with consideration of intracardiac anatomy, previous palliative shunt procedures, and planned single ventricle palliative connections. Perfusionists are acquainted with both adult and pediatric cardiopulmonary support. The anesthesiology team and perfusion team are well experienced in hemodynamic monitoring, cardiopulmonary bypass, cyanosis-related issues, pulmonary hypertension, organ protection, hemostasis, fluid management, and targeted parameters for all adult congenital heart disease cases. However, an exceptional case may require a specific strategy, which will be delineated by the surgeon to the operating room staff. The pediatric echocardiographer handles the intraoperative transesophageal echocardiographic studies.

Our Cardiac Surgery Intensive Care Unit is a combined setup for both adult and pediatric cardiothoracic surgery cases. In this unit, care is provided to all cardiopulmonary transplant, palliative, and corrective cardiothoracic surgical procedures. Patients are admitted to the unit postoperatively where nursing care is provided either by a nurse experienced in taking care of both adult and pediatric cardiac surgical patients, or by an adult cardiac surgical nurse with support from the nurse in charge, who is experienced in caring for adult and pediatric cardiac surgical patients. Medical care is organized mainly through the cardiothoracic surgery team and complemented by the pediatric cardiac intensive care team. Additional management is provided if requested in consultation with other cardiovascular subspecialties and medical services. Acceptable hemodynamic and oxygen saturation parameters are prescribed for each patient with the consideration of surgical diagnosis, body size, and age. Long-term patients in the Cardiac Surgery Intensive Care Unit receive their care mainly from the adult intensive care group, and infrequent assistance is provided by the pediatric congenital heart disease staff.

Results

All but two patients survived their operations. Forty-six patients were discharged home. Table 2 summarizes in days the duration of mechanical ventilatory support, intensive care unit stay, and hospitalization for the entire cohort.

Short-term patients (less than 72 hours) in the Cardiac Surgery Intensive Care Unit were managed by the cardiothoracic surgery team and the pediatric cardiac intensive care team. Longer-term patients in the intensive care unit were initially managed by cardiothoracic surgery and the pediatric cardiac intensive care team, then transitioned to adult intensive care service with partial involvement of the pediatric congenital heart disease staff. Rastelli's repair, emergency repair of dissecting aorta postballooning of coarctation, use of extracorporeal life support, and ventricular septal defect closure constitute the main groups requiring prolonged stay in the intensive care unit.

Morbidity

Complications requiring a prolonged intensive care unit stay exceeding 72 hours are listed in Table 3. Only 2 events required the involvement of the cardiothoracic surgeons: institution and decannulation of extracorporeal support for systemic right ventricular dysfunction in a congenitally corrected transposition patient and reoperation for ventricular septal patch endocarditis postventricular septal defect repair. The adult intensive care unit team handled nonsurgical issues such as respiratory failure, renal failure, and sepsis, as in all the other adult patient population. Pulmonary hypertension was treated with intravenous pulmonary vasodilators and nitric oxide, as in most of the pediatric pulmonary hypertensive congenital heart disease cases. Reperfusion injury of the lung was experienced in 2 cases of postoperative Rastelli's operation where no evidence of significant aortopulmonary collaterals was illustrated. It was managed with prolonged intubation utilizing high positive end expiratory pressure, postextubation use of noninvasive positive pressure ventilation support, and intravenous steroids. This strategy was adopted to minimize pulmonary hemorrhage and to allow lung recruitment. One occasion of junctional tachycardia was experienced in one of the mortality cases

Table 2. Duration of ventilatory support, intensive care unit stay, and hospitalization

Surgical procedure	Mechanical ventilation (day)	Intensive care unit (day)	Hospital stay (day)
Tetralogy of Fallot repair	1/2 (1/4–1)	1.8 (1–3)	6.8 (6–9)
Rastelli's repair for tetralogy of Fallot	10 (3–17)	16.5 (5–28)	31 (17–45)
Rastelli's repair for DORV & PS	6	7	16
Subaortic membrane resection	3/4 (0–1)	1.25 (1–2)	6.5 (5–9)
Coarctation repair	2 1/2 (1/4–4)	4 (1–7)	14 1/2 (8–21)
Atrial septal defect closure	0.38 (0–1)	1.38 (1–3)	6.63 (3–12)
Partial anomalous pulmonary venous drainage repair	2/3 (0–1)	1.33 (1–2)	6 (5–7)
Partial atrioventricular canal defect repair	3/4 (1/2–1)	1.25 (1–2)	7.8 (6–10)
Ventricular septal defect closure*	4.8 (0.17–35)	5.8 (1–35)	11.4 (5–37)
Ventricular septal defect, sinus of Valsalva fistula, and aortic incompetence repair	1/2 (0.13–1)	1.5 (1–2)	10.5 (8–13)
Left atrioventricular valve regurgitation and regurgitant pulmonic conduit in corrected transposition of great arteries	14	20	39
Bidirectional Glenn shunt*	5	5	6
Bilateral bidirectional Glenn shunt (Kawashima)	1/4	3	11
Fontan connection	1 (1)	3 (1–6)	33 (13–46)
Insertion of epicardial automated defibrillator in Fontan	0	0	7

Number of days is expressed as mean and (range) for all and as an absolute number for single cases.
DORV & PS; double outlet right ventricle and pulmonary stenosis; *, a single mortality in the group.

discussed below. Patients subjected to the Fontan operation had a longer hospital stay because of pleural effusions.

Mortality

Two patients died in this series during the same operative hospitalization. Patient 1: a 23 year-old-female who underwent bidirectional Glenn shunt procedure for Ebstein tricuspid valve with hypoplastic right ventricle and normal sinus rhythm. Postoperatively, she remained intubated and developed sustained junctional tachyarrhythmia followed by ventricular tachycardia and died 5 days after her operation as a result of ventricular fibrillation. Patient 2: a 27 year-old-male who underwent patch repair of a ventricular septal defect with fenestration at the atrial level resulting from pulmonary hypertension. The patient received multimodal medical treatment for his pulmonary hypertension during the postoperative period, but his course was complicated by renal dysfunction, sepsis, and endocarditis. Two weeks after his first operation, he underwent a second operation for ventricular septal defect patch endocarditis. He remained critical and died as a result of multisystem failure secondary to sepsis.

Table 3. Complications of prolonged intensive care unit stay

Complication	Number of events
Respiratory failure	4
Renal failure	3
Pulmonary hypertension	2
Sepsis	2
Pulmonary reperfusion injury	2
Arrhythmia	1
Extracorporeal life support	1
Reoperation for endocarditis	1

Discussion

Adults with congenital heart disease who may be referred for surgery fall into two general categories: those without previous surgery and those with previous palliative/corrective surgery.[5] These patients will be subjected to surgery either in a children's hospital with the help of adult staff members or in an adult hospital with the assistance of a pediatric congenital heart disease group.[4] Consequently, the operating room arrangement is integrated between the adult and pediatric cardiothoracic surgery setups, but the intensive care stay must be transitioned between the two facilities.

Unlike the experience in North America, late presenters to our center characterize the main bulk of all adult congenital heart disease patients.[6] The perioperative care is provided to adult congenital heart disease patients in an integrated fashion in the same facility. Medical personnel involved in the care of these patients are maintaining their skills and experience through daily handling of both

adult and pediatric cardiothoracic surgical cases. From an academic point of view, constant education is provided to our staff through weekly meetings of the adult congenital heart disease forum, monthly journal club, and the annual organization of an international adult congenital heart disease conference. Moreover, international locum/visiting physicians to our institution and our personnel attending international meetings outside the country have facilitated the exchange of a great deal of knowledge and information.

The delivery of appropriate perioperative care is a largely unmet challenge everywhere. If the experience in adult and pediatric cardiothoracic diseases is retained, an ongoing clinical interaction by the medical staff is required.[7] In our institution, operative strategy, cardiopulmonary support, and postoperative care are of no additional burden on the operating room staff or intensive care unit staff. Clinical management is conducted with comparable quality for elective or emergency situations. Vasoactive medications, different modes of ventilation, monitoring lines, and targeted hemodynamic parameters for adult congenital heart disease patients are part of routine intensive care management. In this series, reperfusion injury of the lung and pulmonary hypertension represented distinguished postoperative issues that needed a longer hospital course and specific treatment. We believe that reperfusion lung injury seen post-Rastelli's procedure is an exceptional postoperative morbidity and is best dealt with by using invasive and noninvasive positive pressure ventilations, provided that additional pulmonary blood flow through aortopulmonary collaterals is ruled out.[8]

In summary, the perioperative care to adult congenital heart disease patients can be arranged in (1) an adult hospital with the assistance of a pediatric cardiac team; (2) a children's hospital with the assistance of an adult cardiac team and the potential transfer to an adult hospital postoperatively; or (3) an integrated setup, such as our example in this report, where both adult and pediatric cardiothoracic surgery cases are cared for in the same facility. Nevertheless, here are the advantages of our perioperative care setup for adult congenital heart disease patients: (1) decreasing patient anxiety and mounting patient trust in the staff through continuity of care; (2) cost-effectiveness; (3) enhancing familiarity of the surgeon with the congenital anomalies in a larger client; (4) enriching the knowledge and experience of the anesthesiologists and perfusionists regarding organ protection, hemostasis, and intraoperative support; (5) exercising comprehensive nursing care for adult patients with congenital heart disease; (6) lessening apprehension of the medical staff; (7) providing an ideal design for a potential adult congenital heart disease multidisciplinary training center.

Therefore the ongoing clinical interaction and frequent exposure of the staff to adult and pediatric cardiothoracic procedures has helped in facilitating the provision of optimum care to the patients of adult congenital heart disease. This distinctive environment allows our group to provide appropriate care to our adult congenital heart disease patients in a well-integrated discipline. Such a design can be used as a cliché for establishing a new surgical adult congenital heart disease unit and for training staff members who will be involved in providing care for this group of patients.

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