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Primary Total Repair for Cyanotic Congenital Heart Diseases in Saudi Adults: A 9-Year Retrospective Analysis

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Suudi Erişkinlerde Siyanozlu Doğuştan Kalp Hastalıklarında Primer Tam Tamir

Doğumsal kalp hastalıkları nedeniyle 1981 ile 1990 yılları arasında ameliyata tabi tutulan 1375 erişkinin 58'inde, siyonuzlu doğuştan kalp anomalisi bulunuyordu. Anılan hastalara dair eldeki bu retrospektif incelemede Eisenmenger sendromu dahil edilmedi. Bu çalışmada başvuru sırasındaki klinik forma, incelemeler, patoloji yelpazesi, yapılan primer cerrahi tamirin türleri, cerrahi sonuçları ve izleme irdelenmektedir.

Cyanotic complex congenital heart disease in adults is rare in the developed countries nowadays, as early recognition and proper medical and surgical treatment is carried out in infancy or childhood. Such anomalies are, however, not infrequently seen in adults in the developing countries [1], due to either lack of medical and surgical facilities 20-30 years ago, negligence in early referral, the indolence of such patients, socioeconomic conditions, or illiteracy. Cyanotic congenital heart disease is rarely seen today in the West, and only a few data are available in the literature on primary total repair of congenital cyanotic heart disease in adults and their surgical results [2-4].

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MATERIAL and METHODS

Fifty-eight adult patients with complex cyanotic congenital anomalies were studied between 1981 and 1990. Their ages ranged from 14 to 38 years, with a mean age of 19.1 years. There were 37 males and 21 females. Fifty-one patients were in N.Y.H.A. functional class III, and 7 patients in N.Y.H.A. class IV. All patients with Eisenmenger syndrome were excluded from the study. None of the 58 patients had earlier medical or surgical type palliative intervention. Fifty-one patients presented with obvious central cyanosis, while this was not easily clinically detectable in 7 patients due to the presence of associated anemia due to iron deficiency and malnutrition in 5 patients, and parasitic infestation in 2 patients. The anemia had therefore masked the cyanosis. The main complaint was exercise intolerance in 49 patients (84.5 %), squatting in 7 patients (12.1 %), hemoptysis in 3 patients (5.2 %), repeated chest infections in 8 patients (13.3 %), congestive heart failure in 2 patients (3.4 %), low cardiac output and moribund state in 1 patient (1.7 %), and infective endocarditis in 1 patient (1.7 %). Clubbing was present in only 23 patients (39.7 %), 3 patients presented with cerebral abscess (5.2 %). Fifty-three patients had lesions with

Table 1. Associated lesions seen in 58 adult cyanotic patients

Lesion	No. of patients
Cerebral abscess	3
Coronary artery anomalies	
- Anomaly of course	4
- Anomaly of origin	2
- Coronary artery fistula	1
Persistent left superior vena cava draining into right atrium	2
Double chambered right ventricle	1
Infective endocarditis	2

Table 2. Types of total repair performed for 58 cyanotic adult patients

Lesion	No. of patients
Tetralogy of Fallot	31
- V.S.D. patch closure	31
- Infundibular resection	25
- Pulmonary valvotomy	6
- R.V.O.T. patch	3
- Angioplasty of a hypoplastic main pulmonary artery	2
Triglogy of Fallot	8
- Pulmonary valvotomy	8
- A.S.D. pericardial patch closure	8
- Transannular patch	2
D- T.G.A. with V.S.D. patch closure	4
L- T.G.A. with P.S. and V.S.D.	3
- V.S.D. patch closure and insertion of a valved conduit from RV to MPA	3
Tricuspid Atresia	3
- Fontan	2
- Bjork	1
Ebstein Anomaly	1
- T.V.R. and A.S.D. patch closure	
- T.V. repair and A.S.D. closure, in both cases, obliteration of atrialized portion of right ventricle was added	
Single Ventricle	2
- Fontan right atrio-pulmonary anastomosis using non-valved conduit	1
- and valved conduit	1
Total Anomalous Pulmonary Venous Return	1
- Supracardiac type, ligation of persistent left superior vena cava, left atrial common pulmonary vein trunk anastomosis and ASD pericardial patch closure	
Congenital T.S. and P.S. with Sinus Venosus A.S.D.	1
- Tricuspid valvotomy, pulmonary valvotomy, re-routing of right superior pulmonary vein into left atrium using autologous pericardial patch	
Total A.V. Canal	1
- Total repair of tricuspid and mitral valve and patch closure of VSD & ASD	
Common Atrium with P.V. Stenosis	1
- Pulmonary valvotomy and septation of common atrium using a pericardial patch	

reduced pulmonary blood flow (91.4 %), while only 5 had lesions with increased pulmonary blood flow (8.6 %). The hemoglobin ranged from 8 to 23 g %, with a mean of 18 g %. The hematocrit ranged from 29 % to 83 %, with a mean of 56 %. The systemic oxygen saturation ranged from 55 % to 82 %, with a mean of 75 %. The cardiothoracic ratio was increased in more than 75 % of the patients, with a mean of 66 %. Fifty-three patients had oligemic lungs and 5 had plethoric lungs seen on the chest X-ray. Cardiac catheterization was performed on all of our patients, with detailed study of oximetry, pressure data and cineangiography. The mean left ventricular ejection fraction was 65.0 % (range 40-80 %), the mean left ventricular enddiastolic pressure was 5.0 mmHg (range 1-15 mmHg); the mean right ventricular systolic pressure was 102 mmHg (range 32-232 mmHg), and the mean right ventricular enddiastolic pressure was 4.1 mmHg (range 2-20 mmHg). The mean pulmonary artery pressure was 22.5 mmHg (range 8-112 mmHg). We did not have any established Eisenmenger syndrome in this series, even though pulmonary artery pressure at systemic level was encountered. None of these 58 patients had undergone any type of medical or surgical palliavite treatment or intervention in infancy or childhood. The distribution of the encountered complex anomalies is shown in Table 2.

Surgical Techniques

During induction and maintenance of anesthesia, all efforts were made to avoid sudden hypotension which could eventually lead to more central cyanosis, with consequent myocardial hypoxia and low cardiac output. In all 58 patients, access was gained through a median sternotomy incision. Cardiopulmonary bypass was instituted in a speedy, but adequate manner, using bi-caval cannulation for venous drainage and cannulation of the ascending aorta for arterial return. If hypotension occurs during these manipulations, a vasoconstictor such as Aramine is given intravenously, or the ascending aorta is pinched temporarily digitally, as to increase systemic vascular resistance and thus reduce the right to left shunt. In patients with persistent left superior vena cava, the vein was temporarily occluded, as long as it drained into a present left innominate vein.

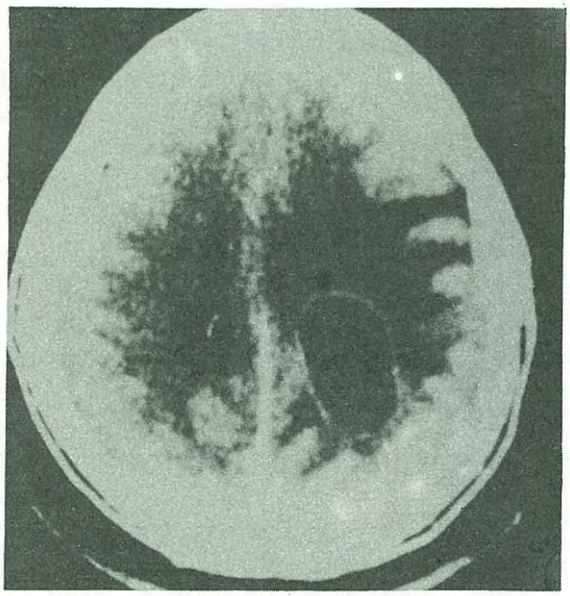

Fig. 1. CT-scan showing cerebral abscess in a 32-year-old adult male patient with D-transposition of the great arteries and a large subaortic ventricular septal defect.

Only membrane oxygenators were used (CML Cobe and Maxima, Johnson and Johnson), to minimize damage to platelets, erythrocytes and clotting factors, thus reducing the known tendency towards bleeding associated with cyanotic heart diseases. Fresh blood, platelet concentrates and fibrinogen were made available whenever possible, to counteract the bleeding tendency. Myocardial protection was always achieved by even central cooling on total cardiopulmonary bypass to 28°C before arresting the heart with cold hyperkalemic cardioplegic solution (Plegisol, Abbott). Additional topical cooling was achieved by pouring iced saline into the intra and extracardiac cavities. Much emphasis was placed on proper myocardial preservation in these adults, as chronic myocardial hypoxia could have led to certain myocardial fibrosis and loss of contractile sarcolemmal elements. In patients with severe cyanosis and torrential collateral circulation, deep hypothermia to 20°C was practiced in association with keeping the bypass flow at 0.5 liters/m^2, as practiced by Wheatleley, to reduce the non-coronary collateral flow and enable us to perform the surgical procedure with better vision and precision.

Total correction of the cyanotic cmoplex congenital cardiac anomalies was possible in all 58 patients. Pulmonary wedge biopsies from the upper and lower lobes of both lungs were done in 3 patients with advanced pulmonary hypertension, to be evaluated according to Edward and Heath grading. Myocardial fibrosis was noted in 2 patients, due to the chronic long-standing hypoxia, although this did not affect the outcome of the surgery in those patients.

RESULTS

Three patients died after the total correction, as they could not be weaned from cardiopulmonary bypass, and severe RV failure occurred. One had a total correction for tetralogy of Fallot, and the other for triology of Fallot; the third patient died following Fontan procedure for single ventricle with post-operative continuous bleeding, leading to cardiac arrest. He was re-opened in the intensive care unit with unsuccessful cardiopulmonary resuscitation (CPR). Death in these adults suffering from cyanotic cardiac anomalies is mainly related to the chronic and advanced myocardial hypoxia which leads to myocardial fibrosis and right ventricular dysfunction or failure occurring post cardiopulmonary perfusion.

The perioperative surgical mortality rate was therefore around 5.2 %. All patients had strict and regular follow-up with clinical, hematological, electrocardiographic, radiographic and echocardiographic serial examination. No late death has been reported so far. The postoperative functional status revealed that 48 patients (82.8 %) moved to N.Y.H.A functional class II, which was a remarkable improvement from their preoperative functional status. In all patients, the central cyanosis has disappeared totally. The encountered postoperative complications were congestive heart failure due to right ventricular dysfunction in 2 patients, who required a prolonged ventilatory and pharmacologic cardiotonic support (more than 3 days). Seven patients required transitional cardiotonic support (1-3 days). Three patients were re-opened for continuous bleeding due to well developed collateral circulation. One of these 3 patients developed cardiac tamponade and required urgent surgical evacuation. One presented with persistent supraventricular arrhythmias for 3 weeks which responded to Verapamil.

Total correction in this subset of patients is encouraged as it has satisfactory subjective and objective improvement and an acceptable surgical mortality rate. However, the importance of early diagnosis and treatment in infancy or early childhood cannot be over-emphasized to prevent development of advanced pulmonary hypertension, congestive heart failure, progression into Eisenmenger state, development of arrhythmias or occurrence of sudden death.

DISCUSSION

Congenital cyanotic heart disease in adults is uncommon in the developed countries, where well-established primary health care facilities exist, and detection of these patients, proper investigation and treatment is carried out early in life. In most developing countries, it is not unusual to find adults going about with an undiagnosed congenital cyanotic heart disease, compounded by anemia, which tends to mask the central cyanosis in the patients. Tetralogy of Fallot is the most commonly encountered disease entity in this subset of patients, as seen in our series. The other cyanotic heart diseases may be so severe that they do not survive to adulthood. These include severe pulmonary atresia, transposition of the great atreries, single ventricle and endocardial cushion defect. Patients with Ebstein's anomaly who have mild tricuspid valve disease and a right to left shunt via an atrial septal defect (ASD) can survive to adulthood. Eisenmenger's syndrome can complicate the pathology in these patients, in which case surgical correction is contraindicated. Remarkably, none of the patients in our series had a preoperative complication of increased pulmonary vascular resistance. In some patients who had elevated pulmonary artery pressure and marginal increase in pulmonary vascular resistance, we performed open lung biopsy prior to the planned cardiac surgery, to determine the degree of obliterative lung disease and hence, determine operability. Our operative mortality of 5.2 % is an indication that operation for congenital cyanotic heart disease in the adult is a feasible venture. Therefore, these patients should not be denied surgery. Some authors have reported a mortality in these patients similar to repair in childhood from their experience with repair of tetralogy of Fallot in adults.

Adult patients with congenital cyanotic heart disease have been subjected to long periods of cyanosis and polycythemia. Hypoxia is also common. All these account for the findings of myocardial fibrosis due to long-standing hypoxia, which could contribute to postoperative right ventricular failure. There is a high incidence of operative and postoperative bleeding due to well developed numerous collateral vessels to the lungs [5], and the mediastinum, in addition to disturbances of the clotting factors, thrombocytopenia and thromboasthenia. Late postoperative arrhythmias have been reported in adults undergoing correction of congenital cyanotic heart diseases [6,7]. The maximum follow-up so far in our series is 98 months, but we have not yet encountered this complication. The actuarial survival after 5 and 9 years is 85 % and 78 % respectively. Early recognition, investigation, referral and surgery is mandatory to avoid development of the known associated complications, such as congestive heart failure, pulmonary hypertension, infective endocarditis, brain abscess and hemoptysis, coronary artery disease and cardiomyopathy. Pregnant women with cyanotic heart diseases may encounter a 50 % fetal mortality or birth of infants with low birth weight for the gestational age. Spontaneous termination of pregnancy is also an ever persisting hazard. In conclusion, like most other outhors, we advocate that adults with congenital cyanotic heart disease can undergo a primary total corrective operation safely with low operative mortality, [1-3] provided that adequate myocardial protection and preservation is practiced during the open heart surgery, and a significant residual gradient in diseases with right ventricular outflow tract obstruction is either avoided, or reduced as much as possible.

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