Study Of The Right Ventricular Function After Total Surgical Correction Of Tetralogy Of Fallot

Thesis

Submitted For Partial Fulfillment Of
The Doctorate Degree In cardio-thoracic Surgery

By

Mohamed Shaffik Hassan Abd Allah

M. B. B. Ch., M.S. (Surg.)

Supervised by

Prof. Dr. Mohamed S. El-Fiky
Professor of Cardio-thoracic surgery
Faculty of Medicine, Ain Shams University

Prof. Dr. M. Magdi Mostafa
Professor of Cardio-thoracic surgery
Faculty of Medicine, Ain Shams University

Prof. Dr. Samir M. El-Mahmoudy
Associate Professor of Cardio-thoracic surgery
Faculty of Medicine, Al-Azhar University

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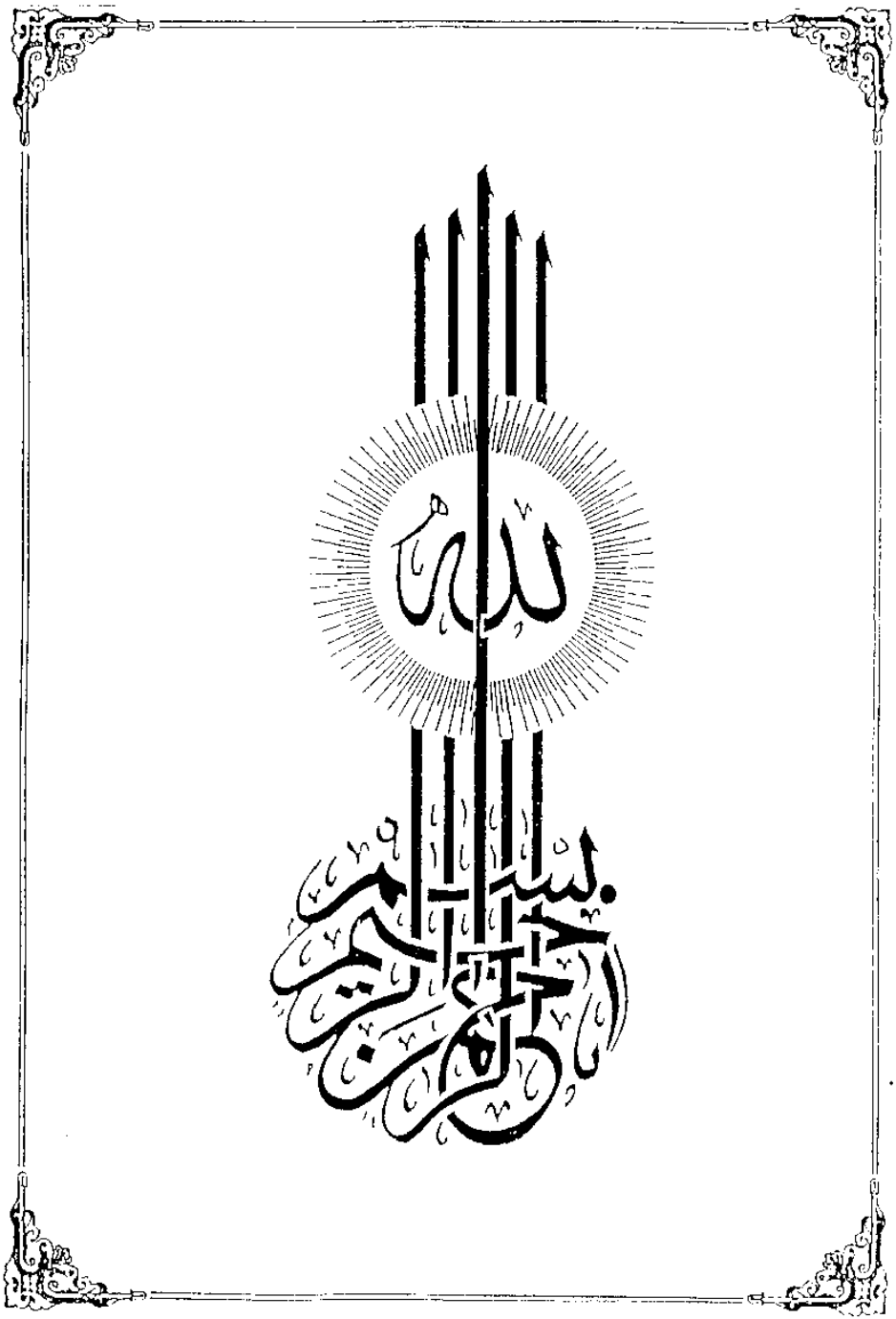

This Humble Work To
My Parents Who Are The
First Teacher To Me

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M. S. Abd Allah

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LIST OF ABBREVIATIONS

Ao	Aorta
AV	Arterioventricular
BP	Blood pressure
BSA	Body surface area
BUN	Blood urea nitrogen
CPB	Cardiopulmonary bypass
CTR	Cardio-thoracic ratio
DC	Defibrillating shock
ECG	Electrocardiogram
FFP	Fresh frozen plasma
HB	Hemoglobin
ICU	Intensive Care Unit
IVC	Inferior vena cava
LAO	Left anterior oblique
LL	Lower limbs
LV	Left ventricle
No. pts.	Number of patients
NYHA	New York Heart Association
P RV/LV	Pressure ratio
PCV	Packed cell volume
PDA	Patent ductus arteriosus
PI	Pulmonary incompetence
PS	Pulmonary stenosis
pt. No.	Patient number
PTFE	Poly tetra fluoroethylene
QP	Pulmonary blood flow
QS	Systemic blood flow
RAO	Right anterior oblique
RBBB	Right bundle branch block
RR	Respiratory rate
RV	Right ventricle
RVOT	Right ventricular outflow tract
SVC	Superior vena cava
TC	Total correction
TOF	Tetralogy of Fallot
TR	Tricuspid regurgitation
TSM	Trabecula-septomarginalis
VSD	Ventricular septal defect

CONTENTS

INTRODUCTION & AIM OF THE WORK

Congenital Cardiovascular disease is defined as an abnormality at birth in cardiocirculatory structure or function ***(Friedman, 1988).***

Tetralogy of Fallot (TOF) represents 10 percent of all forms of congenital heart diseases, and it is the most common cardiac malformation responsible for cyanosis ***(Kirklin and Karp, 1970; Rowe, 1978)***.

Many physicians described the malformation starting by ***Stensen (1672), Sandifort (1777), John Hunter (1784), William Hunter (1784), Farre (1814), Gintrac (1824), Hope (1839), and Peacock (1866). Fallot (1888)*** was the first physician who described accurately the clinical and the pathologic manifestations [Interventricular septal defect (VSD), pulmonary valve stenosis (PS), overriding of the aorta and concentric hypertrophy of the right ventricle]. He reported 55 patients with congenital heart disease, most of whom had the tetralogy malformation. Retrospectively, it is remarkable that such a large number of patients could have been reported by a single author at that time ***(Ross and Sabiston, 1990)***.

Despite the fact that accurate clinical diagnosis could often be established after these contributions by Fallot, many years have passed before definitive treatment of the condition became available. In ***1944 Blalock*** had done a systemic pulmonary anastomosis by joining the

subclavian artery to the pulmonary artery and the child showed marked improvement. Several months later, ***Blalock and Taussig (1945)*** reported this patient and two others. ***Potts et al., (1946)*** introduced the descending aorta to left pulmonary artery anastomosis, ***Brock and Campbell (1950)***, expanded the scope of palliative operations by adding closed infundebulectomy and pulmonary valvotomy.

After that a new era in surgical treatment was established when Lillehei and Varco had done the first successful open repair at the university of Minnesota in 1954 by using controlled cross circulation. In the next year, Lillehei replaced this technique with the use of cardiopulmonary bypass ***(Lillehei, et al., 1955).***

Since then, several advances in diagnosis and surgical intervention had clued that the degree of obstruction to the pulmonary blood flow is the principal determinant of the clinical presentation and future reconstruction. It is presented as infundibular stenosis of the right ventricle in approximately 50% of cases which may be combined with valvular obstruction in another 20 to 25% ***(Child, et al., 1984)***. Also, it may be presented as supravalvular and peripheral pulmonary arterial narrowing ***(Marady, et al., 1984)***.

In many infants and children the obstruction to the right ventricular outflow tract (RVOT) is mild but progressive ***(Friedman, 1988).***

Castaneda and Norwood in ***1983*** recommended elective repair in infancy within the first or second year of life primarily to avoid the secondary hypertrophic changes of the parietal bands and the free right ventricular wall and also the development of heavy trabeculations binding the parietal and septal bands to the right ventricular wall. So, repair at a younger age with good follow-up is recommended as 25% of the surgically untreated infants born with tetralogy will die in their first year of life and 40% are dead by 3 years of age ***(Kirklin and Barratt-Boyes, 1986).***

The results of reconstruction are related to the presence of residual RVOT obstruction, residual VSD, ventricular arrhythmias and late development of conduction disturbance ***(Zhao, et al., 1985)***. The mean cardio-thoracic ratio tends to be somewhat higher in patients with a transannular patch than in those treated by infundebulectomy alone ***(Fuster, et al., 1980)***. Furthermore, hemodynamic evaluation by invasive and non invasive techniques have demonstrated abnormalities of right ventricular volume and function ***(Rocchini, 1977)***, as well as reduced cardiac performance during exercise ***(Wessel, et al., 1980A).***

The aim of this work is to study the right ventricular function before and after total surgical correction of tetralogy of Fallot, relating different types of right ventricular reconstruction to different anatomic finding and trying to find out the best way of reconstruction in ralation to the surgical outcome.

EMBRYOGENESIS

Many views were published to clarify the developmental abnormalities in TOF. The older concept that TOF is due to truncal malseptation at the expence of the pulmonary artery, have been disputed, since it does not readily account for the malalignment VSD and the infundibular abnormality ***(Zuberbuhler, 1989).***

Van Praagh, (1970) considered TOF to be due to underdevelopment of the distal portion of the pulmonary conus. According to this concept, the right ventricular outflow tract (RVOT) obstruction and secondary right ventricular hypertrophy result from hypoplasia of the parietal band of the crista supraventricularis. The VSD is caused by "cristal malalignment" and the aortic dextroposition is due to underdevelopment of the subpulmonary conus.

Becker and Anderson, (1978) considered the anomaly to result from lack of normal rotation and unequal partitioning of the distal bulbus . The lack of normal rotation produces the aortic dextroposition and the malalignment VSD. Anterior displacement of the bulbar ridges causes unequal division of the pulmonary and aortic components of the distal bulbus and narrowing of the pulmonary outflow tract.

In all, there are many factors in the process of the development sharing in the malformation of TOF.

PATHOLOGIC ANATOMY

In normal heart, the infundibular or outlet septum inserts between the limbs of the trabecula septomarginalis, and separates the subaortic and subpulmonary outflow tracts.

The trabecula septomarginalis extends from the ventricular apex toward the RVOT area and terminates in anterior and posterior limbs, fused with the ventriculo-infundibular fold; The ventriculo-infundibular fold is a portion of ventricular free wall which is a part of the inner curvature of the heart, normally it lies between the tricuspid and pulmonary valves ***(Zuberbuhler, 1989)***. In TOF there is divorce of these structures, the outflow septum being inserted anterior to or fusing with the anterior limb of the trabecula septomarginalis. This anomalous insertion of the outflow septum leaves a malalignment gap in the ventricular septum (VSD) which permits the aorta to override the septum and at the same time produces narrowing of the pulmonary infundibulum. The right ventricular hypertrophy can be considered a hemodynamic consequence of these abnormalities ***(Becker and Anderson, 1983)***.

So, the anatomical features of TOF are based on the presence of a large unrestrictive VSD that ensures equal ventricular pressures, along with existence of sufficient pulmonary stenosis (PS) to cause pulmonary arterial pressure to be normal or subnormal. Although a perimembranous VSD and infundibular stenosis will be found in most

patients (classic TOF), Others may exhibit different types of VSD and other sites of RVOT obstruction ***(Zuberbuhler, 1989)***.

PULMONARY STENOSIS

The pulmonary stenosis or RVOT obstruction is variable in degree and in location.

Subvalvar PS:

which is present in almost all patients of TOF, results from anterior deviation of the outflow septum. This narrowing of the subpulmonary components is proportional to the degree of anterior deviation. The infundibular (outflow) septum is often hypertrophied, adding to the obstruction. The hypertrophied abnormal positioning parietal and septal bands are secondary to distal infundibular narrowing. In some instances the apical components of the trabecula septomarginalis, together with apical trabeculations, forms a hypertrophied apical shelf "double chamber right ventricle" ***(Becker and Anderson, 1978)***. The severity of obstruction increases with time due to progressive Hypertrophy and may lead to an acquired infundibular atresia ***(Becker and Anderson, 1983 & Zuberbuhler, 1989)***.

Annular Stenosis:

The pulmonary annulus is normally a muscular structure, and like the infundibulum varies in diameter during the cardiac cycle. In TOF, it is almost always smaller in diameter than the aortic ring, in a way that adequate relief of obstruction cann't be accomplished without insertion

of a transannular patch, the annulus may become thick from fibrosis which is usually an extension of endocardial thickening (fibrosis) ***(Lev and Eckner, 1964 & Rao, et al., 1971).***

Valvar Stenosis:

The pulmonary valve is abnormal in most cases, stenotic in 75% of cases, and approximately two thirds of the stenotic valves are bicuspid. The stenosis at the pulmonary valve is usually the result of leaflet tethering rather than commissural fusion. The length of the free edge of the tethered thickened leaflets is considerably shorter than the diameter of the pulmonary artery, so that the valve can not open adequately and the pulmonary artery is pulled inward at the point of commissural attachement producing a localized narrowing or waisting of the artery at distal valve level ***(Kirklin and Barratt-Boyes, 1986).***

Stenosis may also be present, due to tethering of the pulmonary artery at the site of insertion of the deformed valve leaflets ***(Arciniegas, 1985).***

Supravalvar Pulmonary Stenosis:

The pulmonary trunk is nearly always smaller in size than the aorta in TOF, which is most marked when there is diffuse right ventricular hypoplasia ***(Becker and Anderson, 1978).***

Stenosis at the origin of either one or both pulmonary arteries is present in 15-20% of cases. Some degree of diffuse hypoplasia of the

distal pulmonary artery branches is frequently present and was observed in 82% of the patients. Although absence of the right pulmonary artery has not been described, the left pulmonary artery branch is absent in about 3% of patients with TOF ***(Arciniegas et al., 1980A).***

VENTRICULAR SEPTAL DEFECT

The potential area of communication between the ventricles is always large, except in very rare cases where its right ventricular margin is shielded by insertion of accessory leaflet tissue of the tricuspid valve to its rim ***(Faggian, et al., 1983)***. The VSD is perimembranous in the majority of cases, results from malalignment of the infundibular septum. It is difficult to specify one single plane as being the locus of the VSD in TOF. The most obvious plane would intersect the orifice of the aortic valve, which seems an inappropriate margin for a defect. It is however, possible to imagine the defect as a cone of space, with the base being the overriding aortic valve (superiorly), bordered posteroinferiorly by an area of fibrous continuity between the tricuspid, mitral, and aortic valves. The border of the defect is muscular anteriorly and anteroinferiorly and is made up of the anterior and posterior limbs of the trabecula septomarginalis (TSM) respectively. The posterior limb of the TSM fused with the ventriculo-infundibular fold forming a muscular rim between the tricuspid valve and the defect inferiorly ***(Anderson, et al., 1981).***

Much less commonly, the VSD is associated with absence or deficiency of the infundibular septum 3%, termed "supracristal or