VARIANT ANATOMY OF TRANSPOSITION OF THE GREAT ARTRIES WITH ITS DIFFERENT OPERATIVE TECHNIQUES

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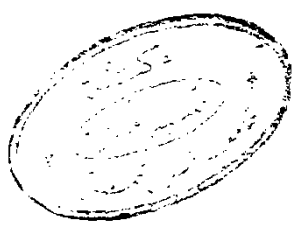

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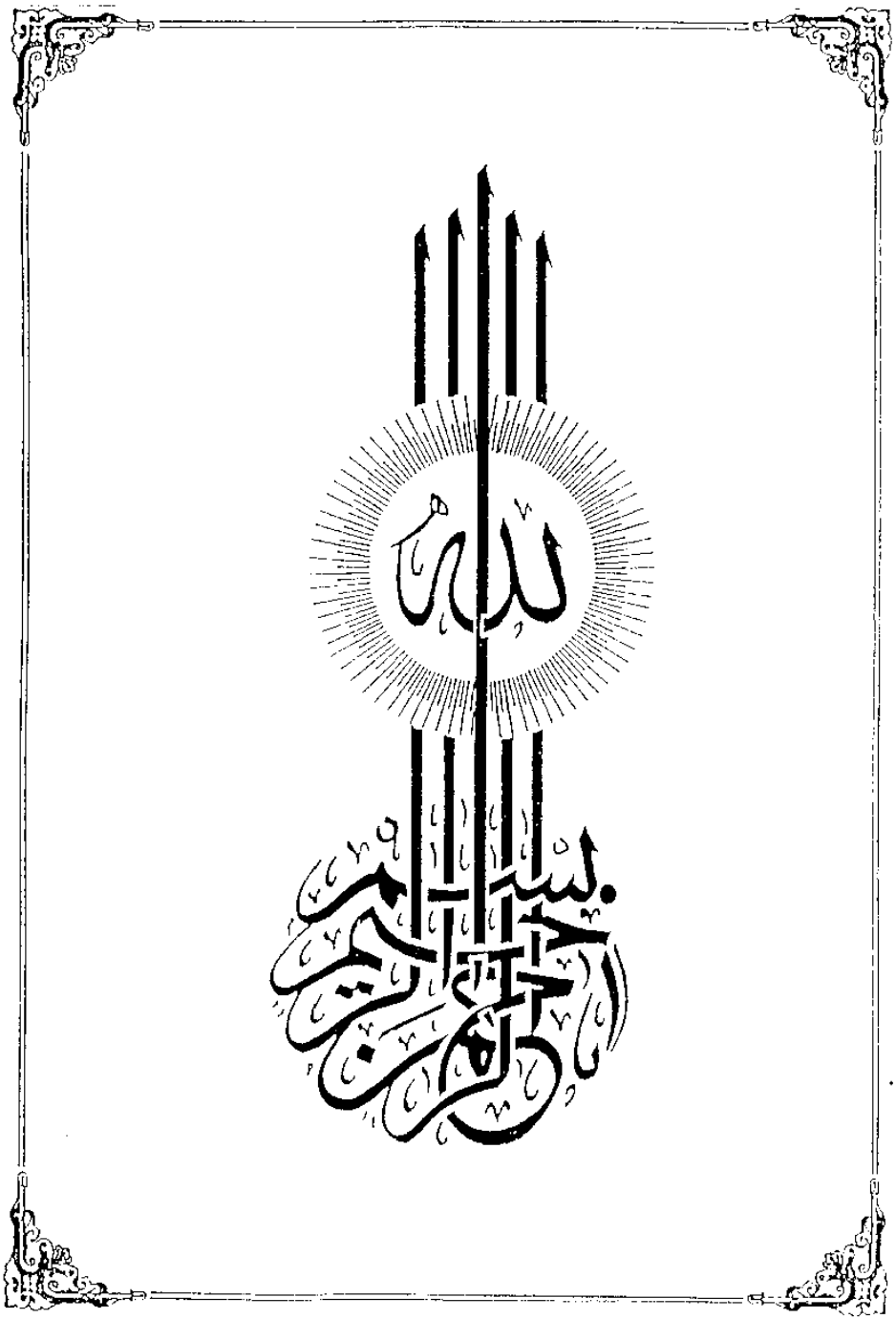

CONTENTS

CHAPTER I

INTRODUCTION

The most common abnormal anatomic relationship between the great arteries and the ventricles is complete transposition of the great arteries (TGA). Transposition of the great arteries literally means that the aorta and the pulmonary artery are placed across (trans = across, ponere = to place), rather misplaced across the ventricular septum so that each great artery arises completely above the wrong ventricle : aorta from the right ventricle, and pulmonary artery above the left ventricle.

The most common clinical representation of the transposition abnormality is understood to represent complete, physiologically uncorrected, transposition of the great arteries (TGA). Functionally this type of transposition is characterized by poorly oxygenated systemic venous blood being misdirected to the systemic circulation and highly oxygenated pulmonary venous blood similarly misdirected to the pulmonary circulation.

NATURAL HISTORY

INCIDENCE:

Transposition of the great arteries (TGA) represents approximately ten percent of all congenital cardiac malformations (Turley et al., 1978). The minimal incidence of transposition of the great arteries is 1 : 4,500 births but it may be as high as 1 : 2,130 births (Liebman et al., 1969). A more recent incidence is reported to occur in between 1 : 2,300 and 1 : 5,100 live births (Gutgesell et al., 1979).

Epidemiological surveys have suggested an increased prevalence in infants with prenatal exposure to sex hormone therapy & contraceptive pills (Nora et al., 1973). Maternal age was not a factor in the incidence, but there was some evidence that the incidence increases when the mother has had three previous children (Liebman et al., 1969). Extracardiac congenital anomalies are infrequent (9%) in infants with TGA compared to incidence with ventricular septal defect (33%) or tetralogy of Fallot (31%) (Fyler D., 1980).

A strong male preponderance of 60-70% with a male to female ratio of 2 : 1 is always present. There are, however, striking differences among the anatomic subgroups so that the simple group has a male to female ratio of 3.3 : 1 while the complex group is 1 : 1. Of interest is that of the children with VSD, the ratio is the expected 2 : 1 in favor of males, except for the group with pulmonary vascular obstruction, where the male-to-female ratio is 1 : 1. Females, therefore, may be prone to the development of pulmonary vascular disease (Liebman et al., 1969).

SURVIVAL:

Transposition of great arteries (TGA) is a common lethal malformation where only 10% survive one year (Gutgesell et al., 1979). For the whole group of TGA without management, the age of death was as follows : by 1 week, 28.7%; by 1 month, 51.6% and by 1 year, 89.3%. The average life expectancy at birth was 0.65 years; at 1 week of age, 0.87 years; at 1 month, 1.12 years; and at 1 year, 3.92 years. (Liebman et al., 1969).

Associated lesions had a marked effect on the prognosis:

- Simple TGA (no VSD, no ASD) 0.11 years.
- TGA + VSD without increased pulmonary vascular resistance: 0.28 years.
- Complex cases: 0.49 years.
- TGA + ASD: 0.81 years.
- TGA + VSD + Pulmonary vascular obstruction: 2.0 years.
- TGA + VSD + Pulmonic stenosis: 4.85 years.
- Single ventricle: 6.79 years.

Thus for the group of TGA + VSD, the average life expectancy is 1.87 years; but for a child at 1 year the life expectancy is 5.06 years & for a child of 7 years, the life expectancy is 9.07 years (Liebman et al., 1969).

MODE OF DEATH:

The poor survival in patients with TGA and essentially intact ventricular septum is related primarily to anoxia. Inter- current pulmonary infections may develop and are particularly lethal because they reduce effective pulmonary blood flow and lead rapidly to increasing hypoxia, acidemia and death. Death in this group may also result from cerebrovascular events. These are usually due to the polycythemia and increased blood viscosity secondary to severe cyanosis, particularly in association with dehydration. However, hypoxia plus hypochromic microcytic anemia have also been implicated in the etiology of these events. Nonfatal cerebrovascular events also occur in about 6% of patients treated by balloon atrial septostomy and include cerebral abscess. (Phornphutkul et al., 1973).

Patients with TGA and important ventricular septal defect (VSD) usually die with congestive heart failure. The modes of death described for patients with simple

transposition sometimes pertain in this group as well and include frequent intercurrent pulmonary infections.

HISTORICAL NOTE:

The first morphological description of transposition of the great arteries (TGA) is attributed to Baillie in 1797. The term "transposition of the aorta and pulmonary artery" was coined by Farre when he described the first known case of this anomaly in 1814. Transposition (trans = cross, ponere = to place) meaning that the aorta and pulmonary artery are displaced across the ventricular septum. (Van Praagh R., 1971).

In subsequent pathological description that included attempts to explain its embryological basis, the word transposition was used to describe an anterior position of the aorta relative to the pulmonary artery, and by the early 1900, it had become accepted practice to include any abnormal position of the aorta, regardless of its ventricular origin, under this heading. This broad confusing definition was clarified by Van Praagh and colleagues as recently as 1971, when they strongly advocated a return to Farre's original definition of transposition, and introduced the useful term "MALPOSITION" for those abnormal positions of the aorta in which the spatial relationship between the great arteries at the semilunar valve level had been distributed or both great arteries failed to be displaced across the ventricular septum. This literal meaning of transposition is now accepted by most pathologists and surgeons. (Van Praagh R., 1971).

The surgery of TGA commenced in 1950 when Blalock and Hanlon at the Johns Hopkins Hospital described a closed method of arterial septectomy designed to provide mixing of pulmonary

and systemic venous return at atrial level. Edwards and Bargeron modified the Blalock-Hanlon procedure in 1964 by resuturing the septum so as to connect the right pulmonary veins to the right atrium.

The palliation of TGA was revolutionized when Rashkind and Miller in Philadelphia introduced balloon atrial septostomy (BAS) in 1966 (Rashkind & Miller, 1966). However, in 1971 at Great Ormand Street Hospital in London Tynan showed that BAS did not allow all babies with TGA to survive until repair (Tynan et al., 1971). A modification of this procedure was introduced in 1975 by Park and colleagues with their substitution of a blade rather than a balloon at one end of the catheter (Park et al., 1982).

Throughout the 1950 there were attempts to correct TGA either at the atrial or great artery levels. The concept of a physiologic correction at the atrial level by switching the atrial septum so that caval return was directed to the left ventricle and pulmonary venous return to the right was first proposed by Albert at a meeting of the American College of Surgeons in 1954. The first successful operation of this type was accomplished by Senning in 1959 (Senning A., 1959) by refashioning the walls of the right atrium and the atrial septum. Mustard's procedure, in which the atrial septum is excised and a pericardial baffle is used to redirect caval flow, was devised in an attempt to create larger atria then when were produced the Senning procedure and was successfully introduced at the Toronto Sick Children Hospital in 1963 and reported in 1964 (Mustard W., 1964). The Mustard technique soon was adopted in nearly all cardiac surgical centers. The somewhat disappointing results of the atrial switch operation for TGA and large VSD continued to be a stimulus for the development of an arterial switch operation, particularly the

right (systemic) ventricle sometimes failed late postoperatively in these patients. Very early (1954) Mustard had described unsuccessful attempts to perform an arterial switch operation in seven patients with transfer of the left coronary ostium to the pulmonary artery and use of a monkey lung as the oxygenator. Jatene and colleagues in Brazil achieved a major breakthrough in 1975, with the first successful use of the arterial switch procedure, applying it in infants with TGA & VSD (Jatene et al., 1976). Soon after, Yacoub reported successful cases (Yacoub et al., 1976 & 1977). An important technical modification of the original Jatene procedure has been the demonstration by Lecompte and colleagues that direct anastomosis of both great arteries without the interposition of a tube graft is possible when the pulmonary artery bifurcation is transferred in front of the distal ascending aortic arch (Lecompte et al., 1981).

As the arterial switch operation was evolving, the use of a valved extracardiac conduit to effect the anatomic repair was also developed. This was described simultaneously by Kaye and Stansel and earlier by Damus and colleagues and consists of division of the pulmonary trunk and anastomosis of its proximal end with the posterior aspect of the ascending aorta. A valved conduit is placed between the right ventricle and distal pulmonary artery. The left ventricle thus becomes the systemic ventricle, the aortic valve remaining closed because of the higher upstream pressure. If a VSD is present, it is closed (Damus et al., 1982).

CHAPTER II

ANATOMY OF THE HEART
WITH
TRANSPOSITION OF GREAT ARTERIES
MORPHOLOGICAL VARIATIONS

DEFINITION

Complete Transposition of Great Arteries is a congenital cardiac anomaly in which the aorta arises entirely or in large part from the morphological right ventricle and the pulmonary artery arises entirely or in large part from the morphological left ventricle i.e. ventriculo-arterial discordant connection.

TERMINOLOGY

The modifying term "complete" transposition has come to indicate that the transposed great arteries are physiologically uncorrected; systemic venous blood flows predominantly to the aorta and pulmonary venous blood to the pulmonary artery. This contrasts the term "corrected" in corrected transposition i.e. physiologically corrected. It should, however, be recognized that both the typically physiologically uncorrected and physiologically corrected transposition are "complete transpositions" i.e. both great arteries are completely misplaced across the ventricular septum and arising from morphologically inappropriate ventricles. (Paul M.H., 1983).

An additional modifying term, "simple", has been applied to exclude from consideration hearts with complete transposition having certain complex associated malformations such as: tricuspid atresia, mitral atresia, common AV orifice,

and univentricular hearts with or without small outlet chamber "Complex TGA". This simple type of complete transposition includes transposition with intact ventricular septum, ventricular septal defect (VSD) patent ductus arterious (PDA), straddling tricuspid orifice- valve, or left ventricular outflow tract obstruction (LVOTO) as designated by most people. However, surgical papers often use the term simple or isolated transposition of the great arteries to designate a subgroup of TGA patients with intact ventricular septum (IVS) but without any other significant associated lesion. Hence excluding large VSD, large PDA and significant left ventricular outflow tract obstruction (LVOTO). (Trusler et al., 1980).

MORPHOLOGY

RIGHT VENTRICLE

The right ventricle (RV) is normally positioned, hypertrophied and large. Its inflow and sinus portions are essentially normal in their architecture. In about 90 % of cases, there is a subaortic conus (infundibulum), and the aorta is rightward and anterior and ascends parallel to the posterior and leftward pulmonary trunk. In such hearts there is also an infundibular (conal) septum, which in the absence of a ventricular septal defect (VSD) joins normally with the ventricular septum between the limbs of the trabecula septomarginalis (TSM). The infundibulum does not deviate to the left as in the normal hearts but projects directly superiorly from the sinus portion of the ventricle (Kirklin. et al., 1986).

There is less wedging of the pulmonary artery between

mitral and tricuspid valves in TGA than of the aorta in normal hearts. As a result, there is a larger area of contiguity between mitral and tricuspid valves than normally. These atrio-ventricular valves may be at virtually the same level, and the AV septum and the membranous interventricular septum are then smaller than usual or rarely, even absent (Anderson et al., 1983).

In about 10 % of hearts with TGA and intact ventricular septum (IVS) the subaortic conus in the RV is absent or very hypoplastic. The the aorta is either directly anterior or anterior and to the left of the pulmonary artery origin, or rarely, posterior. In a few instances, however, a posteriorly placed aorta is associated with a subaortic conus. This combination may make the arterial switch operation difficult (Van Praagh et al., 1971).

LEFT VENTRICLE

The left ventricle (LV) rarely contains an infundibulum (conus), there usually being pulmonary - mitral fibrous continuity comparable to the aortic - mitral continuity present in the normal heart. In about 8 % of hearts with TGA and most commonly in those with a VSD, a subpulmonary conus is present in the left ventricle. The subpulmonary conus is frequently stenotic. In the majority of these cases, the aorta still lies anteriorly and to the right but it may be leftward or posterior (Van Doesburg et al., 1983).

ATRIA

The atria are normally formed. Right atrial size is usually increased above normal, particularly when the ventricular septum is intact in which case a patent foramen ovale also exists.

SPATIAL RELATIONSHIP AT SEMILUNAR VALVE LEVEL

The possible positions of the origin of the great arteries are nearly infinite around the 360 of a circle but may be simplified as:

(1) Normal, with aorta to the right (in situs solitus) and to the left (in inversus) and somewhat posterior to the pulmonary artery.
(2) Aorta anterior to the pulmonary artery, either directly or somewhat to the right (D- malposition).
(3) Aorta to the left of the pulmonary artery (L-malposition).
(4) Aorta posterior to the pulmonary artery. Normally, the great arteries tend to cross rather to run parallel. In contrast, when the great arteries are malposed, their first portions are usually parallel (De la Cruz et al., 1976).

Almost any spatial relationship between the great arteries at the semilunar valve level can be observed in the patient with TGA and atrial situs solitus. The most common relationship is the aorta right- sided and anterior. The great arteries can also be side- by- side or the aorta may be to the left and anterior.

PULMONARY ARTERIES

The left ventricular outflow tract is usually directed toward the right pulmonary artery. Thus, beyond infancy, there is usually asymmetric pulmonary blood flow, with the axis directed more toward the right lung than the left. This observation has been confirmed by pulmonary perfusion scans. Aneurysmal dilation of the pulmonary arteries may be present,

but absence of the pulmonary valve is rare. (Muster et al., 1976).

CORONARY ARTERIES

The coronary arteries in TGA arise from the aortic sinuses that face the pulmonary artery, irrespective of the inter- relationships of the great arteries. Thus the noncoronary sinus is usually the anterior one. The right coronary usually arises from the rightward posterior sinus and the left coronary from the leftward posterior sinus. Both usually have a normal branching pattern, although not infrequently the right coronary gives origin to circumflex artery that passes to the left behind the pulmonary artery. (Rowlatt et al., 1962).

The variations of coronary artery anatomy in hearts with TGA are considerable and important to the surgeon when doing the arterial switch operation. The right coronary arises from the right sinus in 90 % and from the left sinus in 10 % of cases. The usual pattern of origin and distribution is present in over 60 % of cases. Both coronary arteries arise from the right sinus (from a single or double ostium) in 10 % of cases. (Goor et al., 1975). Yacoub et al., classified the different modes of origin and early branching of the coronary arteries in TGA into 5 types based on his observations during anatomic correction. He suggested three techniques for the transfer of these different types of coronary arteries during arterial switch operation in order to avoid undue tension, torsion, or kinking of the proximal coronary arteries or their early branches (Yacoub et al., 1978). Of considerable potential significance in the atrial switch (Mustard or Senning) operation is the course of the sinus node artery in hearts with TGA. This artery usually arises from the right