Evaluation Of The Severity Of Fallot's Tetralogy By A Scoring System Using The Clinical Data & Investigations

Thesis
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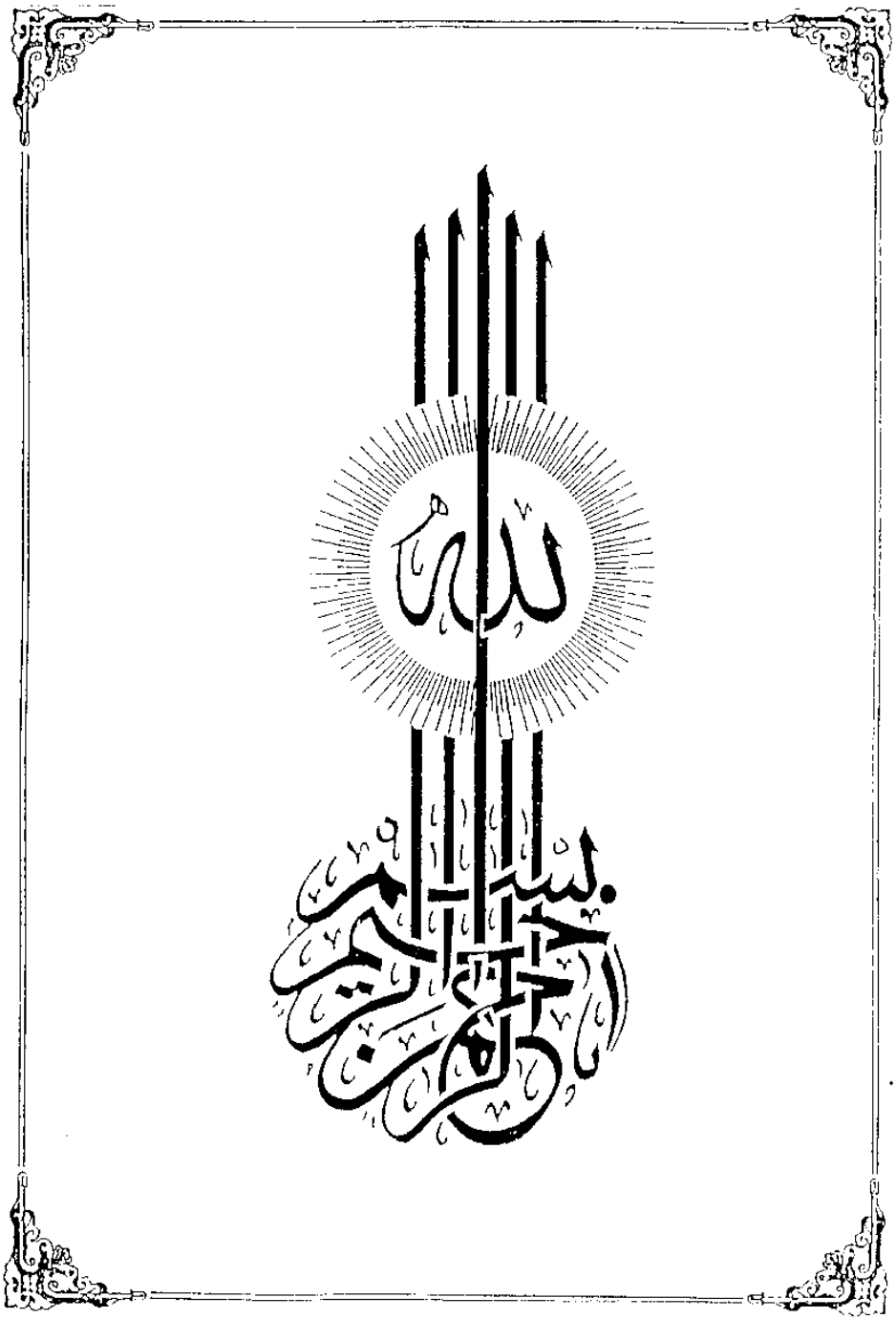

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Introduction
&
Aim Of Work

Introduction & Aim Of The Work

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

The most common serious cyanotic malformation of the heart is the tetralogy of Fallot (TOF) ***(Gaffrey et al., 1983)***.

Tetralogy of Fallot represents 10% of all forms of congenital heart disease, and it is the most common cardiac malformation responsible for cyanosis ***(Kirklin and Karp, 1970)***.

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 to describe accurately the clinical and the pathologic manifestations (interventricular septal defect, pulmonary valve stenosis, 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).***

The degree of obstruction of the pulmonary blood flow is the principal determinant of the clinical presentation and future reconstruction. It 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)***.

It may be with a supravalvular and peripheral pulmonary arterial narrowing ***(Morady et al., 1984)***.

In many infants and children the obstruction to the right ventricular outflow 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 band and the free right ventricular wall and also the development of heavy trabeculations binding the parietal and septal bands to right ventricular wall.

So, repair at a younger age with good follow-up is recommended as 25% of the surgically untreated infants born with severe tetralogy will die in the first year of life, 40% are dead by 3 years of age ***(Kirklin and Barratt boyes, 1986)***. Furthermore, haemodynamic evaluation by non invasive technique (echo cardiography) and invasive technique (Cineangiography) have demonstrated the anatomical finding and progress of volume and function with different age and other risk factors ***(Roochini et al., 1981)***.

Aim Of The Work

The aim of this work is to determine a scoring system for patients with Fallot's tetralogy to evaluate the severity of the condition.

Review Of Literautre

Etiology Of Tetralogy Of Fallot

In most cases, no actual cause can be found for the TOF. However, the condition has occurred in association with rubella in the first trimester of pregnancy, with exposure to thalidomide ***(Vickers, 1967)***, or in association with diabetic embryopathy ***(Kucera et al., 1965)***.

Genetic studies suggest a multifactorial causative mechanism which occurs in Down, XXX, Turner, Klippel-feil, and Noonan syndromes, but is often less common than other cardiac lesions ***(Laks and Breda, 1991)***.

Recently, ultrastructural degeneration is dependent of secondary hypertrophic or hypoxic changes has been observed in both right and left ventricles of patients with tetralogy of Fallot, suggesting the intriguing possibility of an underlying cardiovascular fetopathy as the cause of TOF ***(Toussaint et al., 1987)***.

Embryogenesis

Van Praagh and associates (1970) believed that tetralogy of Fallot is due to underdevelopment of the distal portion of the

pulmonary conus. According to this concept the right ventricular outflow tract obstruction and secondary right ventricular hypertrophy resulted from hypoplasia of the parietal band of the crista supraventricularis.

The ventricular septal defect (VSD is caused by "Cristal malalignment" and 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 ventricular septal defect.

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.

"Pathologic Anatomy"

The cardinal anatomical feature of TOF is anterior deviation of the septal insertion of the infundibular septum.

In the normal heart, the infundibular septum inserts between the limbs of the trabecula septomarginalis and separates the

outflow of the right ventricle from that of the left ventricle, it is fused with the tricuspid and pulmonary valves to form the crista supra ventricularis. In TOF, there is Divorce of the structures, the infundibular septum being inserted anterior to or fusing with the anterior limb of the trabecula septomarginalis. This anomalous insertion of the infundibular septum leaves a malalignment gap in the ventricular septum "the septal defect" 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 haemodynamic consequence of these abnormalities ***(Becker and Anderson, 1983)***.

Since right ventricular outflow tract obstruction and the large "VSD" are the two essential anatomic features, the essence of the problem is closely related to the variations in their pathologic anatomy ***(Rowe, 1978)***.

Right Ventricular Outflow Tract Obstruction

In "IF" the right ventricular outflow tract obstruction is variable in degree and in location. Infundibular obstruction of varying degree is present in almost all cases, the hypertrophical parietal and septal bands and the crista supra-ventricularis

contribute to the infundibular obstruction. In 39% of patients the obstruction is localized to the parietal and septal components of the crista supraventricular without including the distal pulmonary conus ***(Hawe et al., 1972)***.

The obstruction may be close to the pulmonary valve resulting in a small infundibular chamber, or it may be proximally situated leaving a large infundibular chamber with a well demarcated as infundibular and third ventricle ***(Satyanarayana et al., 1971)***.

The pulmonary valve is stenotic to a varying degree in 75% of cases and approximately two thirds of the stenotic valves are bicuspid, the leaflets of a stenotic valve are usually thickened, frequently severely so a feature that increases the amount of obstruction at valve level ***(Iev and Eckner, 1964)***.

The valve cannot open adequately and the pulmonary artery is pulled inward at the point of commissural attachment producing a localized narrowing or wasting of the artery at distal valve level ***(Kirklin and Barrett-Boyes, 1986)***. The pulmonary ring is normally a muscular structure and like the infundibulum varies in diameter during the cardiac cycle.