Original Article

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Determination of TOF Characteristics in Iranian Patients

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Abstract:

Objective: Tetralogy of fallot (TOF) is one of the most common forms of cyanotic congenital heart disease (CHD). The aim of this study was determination of demography, associated anomalies, peripheral pulmonary stenosis (PPS), blood grouping, Rh typing, operation results and complications in TOF.

Material & Methods: The records of 270 patients were reviewed. These patients were admitted from 1993 to 2003, in Shaheed Rajaei Hospital.

Findings: Male patients were 60.37% and females were 39.63%. Incidence of patent foramen ovale (PFO), Right Aortic Arch (RAA), coronary artery (CA) anomalies and other anomalies were 44.81%, 21.11%, 9.25% and 36.30% respectively. Single ostium coronary artery (SOCA) was the most common CA anomaly. The most common PPS was bifurcation stenosis. TOF was more common in O blood group patients. The mean age at the first palliative operation was 5.21 years and for TC was 7.19 years. Post surgical mortality rate was about 3% and morbidity rate, 12.18%.

Conclusion: Trend toward earlier total correction (TC), and single stage early TC of TOF should be recommended as the preferred management strategy.

Key Words: TOF, Congenital Heart Disease, Congenital anomaly, Characteristics, Iranian

Introduction

Tetralogy of Fallot (TOF) comprises a constellation of cardiovascular findings that share the following common anatomic abnormalities: a large malaligned ventricular septal defect (VSD), Overriding of the aorta over the interventricular septum (IVS), right ventricular outflow tract obstruction (RVOTO) and right ventricular hypertrophy (RVH). TOF is one of the most common form of cyanotic congenital heart disease (CHD)[1-7]. The aim of the study was the determination of demography, associated anomalies, peripheral pulmonary stenosis (PPS), blood grouping, Rh typing, operation results and surgical complications of TOF in Iranian children. Because Shaheed Rajaei hospital is the only major referral center for CHDs in Iran, the collected data is universal, and reflects the over all incidence of the anomaly in the country.

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Material & Methods

This study is a descriptive and retrospective study based on the patient's records. The sample size consisted of 270 cases, hospitalized during 10 years (1993-2003) in Shaheed Rajaei cardio-vascular center in Tehran, Iran. According to the records, for all patients, physical examination, chest x-ray, ECG, echocardiography, catheterization, angiocardiography, pulse oxymetry and other diagnostic procedures were done. Diagnosis and classification of TOF was done according to collected clinical and paraclinical data.

Findings

Among the 270 patients with TOF, 107 cases (39.6%) were female and 163 cases (60.4%) male. At the first admission, the mean age for females was 5.6 years (ranging from 6 months to 17 years), and for males 6.6 years (ranging from 6 months to 17 years). The mean hemoglobin at first presentation was 16.7 gr/dl (ranging from 10 to 24.3 gr/dl) for all patients irrespective of sex and age (table 1).

With the exception of right aortic arch (RAA), coronary artery anomalies and patent foramen ovale (PFO), other associated anomalies were found in 98 cases (36.3%) (table 2). Incidence of PFO, RAA and coronary artery anomalies were 44.8% (121 cases), 21.1% (57 cases) and 9.2% (25 cases) respectively (table 3).

Among patients with coronary artery anomalies, single ostium coronary artery (SOCA) was the most common anomaly (4.8%). In one case, the left coronary artery (LCA) arose from the right coronary artery (RCA) (0.4%), and in 2

Table 1- Tetralogy of Fallot and Demographic characteristic of patients

Sex	Number	Diagnostic Procedures Echo & Cath	Patient's Age at the First Admission	Mean Hgb at the First Admission
Female	107 (39.6%)	107 (100%)	5.63 (4 mo-15y)	**16.69 gr/dl (10-24.30)**
Male	**163 (60.4%)**	**163 (100%)**	**6.56 (6mo-17y)**	

Hgb= hemoglobin; gr/dl=gram per deciliter; mo=month; Echo=echocardiography; Cath= Catheterization; y=year; TOF=Tetralogy of Fallot

Table 2- Tetralogy of Fallot and associated anomalies

Anomalies	Frequency(%)	Anomalies	Frequency(%)
Atrial Septal Defect	**39 (14.4)**	**Interrupted-IVC**	
Patient Ductus Arteriosus	**29 (10.7)**	**Azygos-Continuity**	**1 (0.4)**
Left Superior Vena Cava	**11 (4.1)**	**Hemi-Azygos-Continuity**	**2 (0.7)**
Left Pulmonary Artery	**4 (1.5)**	**Pulmonary Valve**	**1 (0.4)**
Multiple Ventricular Septal Defects	**3 (1.1)**	**Right Superior Vena Cava**	**1 (0.4)**
Aortic Insuffiency	**2 (0.7)**	**Atrio-Ventricular Septal Defect**	**1 (0.4)**
Right Pulmonary Artery	**2 (0.7)**	**Wolf Parkinson White**	**1 (0.4)**
		Situs Inversus Totalis	**1 (0.4)**

Table 3- Tetralogy of Fallot and anomalies of coronary arteries

Anomalies	Frequency (%)
Dilated & Large Conal Branch	6 (2.2)
Single Ostium Coronary Artery	13 (4.8)
Conal Branch from Left Anterior Descending Coronary Artery	1 (0.4)
Left Anterior Descending Coronary Artery from Right Coronary Artery	2 (0.7)
Left Circumflex Coronary Artery from Right Coronary Artery	1 (0.4)
Undetermined	2 (0.7)
Total	25 (9.3)

cases the RCA arose from left anterior descending artery (LAD) (0.7%). There was peripheral pulmonary stenosis (PPS) in 38 cases (14.1%) (table 4).

Among patients, PPS of type I (single central stenosis) was 5.2%, type II (bifurcation stenosis) 8.1% and type IV (central and peripheral stenosis) 0.7%. There wasn't any patient with type III of PPS (multiple peripheral stenosis). In the determination of the blood grouping and Rh typing, 88.1% of cases were Rh^+ and 11.85% were Rh^-. The most common blood group among patients with TOF was O (table 5). About 88 patients (32.6%) underwent palliative operation (Gore–tex shunt) as the first operation (table 6). The youngest case was 4 months and the oldest 17 years old, and the mean age at the first palliative operation was 5.2 years.

Corrective operation TC (Total correction) was done in 168 cases (62.2%) as the first operation. The youngest patient undergoing TC was 1.5 years and the oldest one 17.5 years old (table 6). Transanular patch (TAP) was used in 136 cases (50.4%). After surgery, the mortality rate was

Table 4- Tetralogy of Fallot and peripheral pulmonary stenosis (PPS)

Anomalies	Frequency (%)
Type I	
Left Pulmonary Artery	8 (3.0)
Right Pulmonary Artery	6 (2.2)
Type II	22 (8.1)
Type III	0
Type IV	2 (0.7)
Total	38 (14.1)

3.0%, and post operative RBBB (right bundle branch block) was present in more than 90% of patients. Complete heart block (CHB) was found in 7 cases (2.6%), in all of them PPM (permanent pacemaker) was implanted. Other important complications, such as hemorrhage and temponad were seen in 11 cases (4.07%), and in all of them the patients underwent a re-operation.

Discussion

Due to specific characteristics, such as enough sample size, duration and reliability of the collected data, this study is unique until now in Iran. In our study, TOF was more common in males (male/ Female= 60%:4%). With the exception of PFO, RAA and CA anomalies, other anomalies that were found in 36.3% of the

Table 5- Tetralogy of Fallot and Blood groups & Rh types

Blood Group & RH		Frequency (%) Positive	Frequency (%) Negative	Total (%)
Rh		238 (88.1)	32 (11.9)	270 (100)
Blood Group	A	65 (24.1)	9 (3.3)	74 (27.4)
	B	60 (22.2)	9 (3.3)	69 (25.6)
	AB	24 (8.9)	2 (0.7)	26 (9.6)
	O	89 (33.0)	12 (4.4)	101 (37.4)

Table 6- Tetralogy of Fallot and paliative & corrective operation

	Operation	Age (year)	Frequency (%)
Shunt as the first operation	Left Gore-tex shunt	5.2 (0.3- 17)	39 (14.4)
	Right Gore-tex shunt		34 (12.6)
	Left & Right		3 (1.1)
	Blalock Taussig shunt		1 (0.4)
	Central shunt		7 (2.6)
	Right + Central shunt		3 (1.1)
	Left + Central shunt		1 (0.4)
	Right + Left Central shunt		2 (0.7)
Total correction	AS the First Operation	7.19 (1.5-17.5)	168 (62.2)
	As the Second Opration	6.43	88 (32.6)
	Trans anular Patch		136 (50.4)

patients, were atrial septal defect (ASD) and patent ductus arteriosus (PDA) which were the most common.PFO was found in 44.8% , RAA in 21.1% and CA anomalies in 9.2% of the patients. Among CA anomalies SOCA was the most common anomaly. PPS was found in 14.1% of cases. The most common PPS was bifurcation stenosis (type II), TOF was more common in patients with O blood group. Post surgical mortality rate was about 1.1% (1 of 88) for palliative surgery and 2.7% (7 of 256) for TC. RBBB was very common. Interestingly, surgery induced RBBB was observed less in one of our surgical teams. Other than RBBB, complications

Table 7- TOF and post operative complications

	Operation	Frequency (%)
Mortality	Shunt	1 (0.4)
	Total correction	2 (0.7)
Morbidity	Right Bundle Branch	245 (90.7)
	Complete Heart Block	
	Transient	3 (1.1)
	Permanent Pace Maker	
	Epicardial	5 (1.8)
	Endocardial	1 (0.4)
	Epicar+Endocar	1 (0.4)
Others	Hemorrhage & Reoperation	9 (3.3)
	Temponade	1 (0.4)
	VSD patch dehiscence	1 (0.4)

were found in 21 patients (7.7%) after operation. The mean age for palliative operation was about 5 years, and for TC without previous palliative surgery was more than 7 years and for TC with previous palliative surgery was about 5.5 years.

Origin of the LAD from RCA with anterior course across RVOT was found in 5% of TOF in other studies. A large conal branch (accessory LAD) was seen in up to 15% of cases in another study. SOCA may be present in approximately 4% of patients [9, 10]. The rate of associated cardiac anomalies in total is high. ASD is reported to be present in a majority of patients. In other studies a PFO or true ASD was found in 83% of TOF and the incidence of a LSVC was found to be 11% [8, 11].

The trend for early primary repair of CHD is increasing in developed countries. According to a multicenter analysis of the choice of initial surgical procedure in TOF, focused on 938 patients from 12 institutions throughout the USA, who underwent their initial operation during a 10-year period (1986-1995) the percentage of palliative surgery (aortopulmonary shunt) decreased from 35% (1986-1990) to 22% (1991-1995) and the percentage of primary complete repair increased accordingly [15].

According to another study in a developing country, hospital mortality rate of palliative surgery was about 6% [16]. In Mahle et al study, primary complete repair (TC) of TOF was routinely performed in infancy [17].

Many other lesions may co-exist with TOF. PFO and ASD are common. A right aortic arch, though not of functional importance, is common and when detected, should alert the physician for further investigations in the diagnosis of TOF [13, 18].

Stenosis within the pulmonary arteries (PPS) is of major surgical significance and usually occurs at branching sites from the bifurcation outwards [19]. Absent pulmonary valve (APV) is a rare anomaly. This anomaly was seen in one of our patients (table 2).

APV is usually associated with TOF, but it was also seen with an intact ventricular septum (IVS), and was commonly, but no invariably associated with the absence of the ductus arteriosus [20].

Early correction, as a single stage early TC of TOF worldwide [1 ,2 ,12, 13, 14] due to improvement in the comprehensive surgical approach, technology and PICU care, should be regarded as the preferred management strategy.

With the evolution of noninvasive technology such as echocardiography, the indications for diagnostic cardiac catheterization have diminished substantially. Because diagnostic catheterization is invasive and time-consuming, we recommend, that the patients undergo surgery without invasive diagnostic procedures. Nonetheless, invasive procedure is, on occasion necessary for determination of PA, CA and aortopulmoanry collateral arteries anatomy in order to decide on surgical or medical management strategies, for interventional treatment and for a definitive anatomical diagnosis. We need more studies for definitive determination of relationship between TOF, blood grouping and HLA typing.

Conclusion

Trend toward earlier total correction, and single stage early TC of TOF should be recommended as the preferred management strategy.

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