Determination of Tetralogy of Fallot Characteristics in Iranian Patients

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Abstract

Background- Tetralogy of Fallot (TOF) is one of the most common forms of cyanotic congenital heart disease (CHD). The aim of this study is determination of demography, associated anomalies, peripheral pulmonary stenosis (PPS), blood group, Rh type, operation results and complications in TOF.

Methods- The records of 270 patients were reviewed. These patients had been admitted during 10 years (from 1993 to 2003).

Results- 60.37% of patients were male and 39.63% were female. 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 CA anomaly. The most common PPS was bifurcation stenosis. TOF was more common in blood group O patients. The mean age at the first palliative operation was 5.21 years and for total correction (TC), it was 7.19 years. Postsurgical mortality rate was about 3% and morbidity rate was 12.18% (excluding right bundle branch block, RBBB).

Conclusion- The trend toward earlier total correction and single stage early total correction of TOF should be recommended as the preferred management strategy ***(Iranian Heart Journal 2007; 8 (3): 21-26).***

Key words: Tetralogy of Fallot ■ congenital heart disease ■ cardiac surgery

Tetralogy of Fallot (TOF) comprises a constellation of cardiovascular findings that share the following common anatomic abnormalities: a large mal-aligned 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 forms of cyanotic congenital heart disease (CHD).[1-7]

The aim of the study is to determine the demography, associated anomalies, peripheral pulmonary stenosis (PPS), blood group and Rh type, operative results and surgical complications of TOF surgery in Iranian children. Because Shaheed Rajaie Cardiovascular Medical Center is the only major referral center for CHDs in Iran, collected data are universal, and related to all over Iran.

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Methods

This study was a descriptive and retrospective study based on patients' medical records. Sample size was 270 cases, hospitalized during a 10-year period (1993-2003) in Shaheed Rajaei Cardiovascular Medical Center in Tehran. Acquired data were analyzed and summarized in 7 tables. According to the records, physical examination, chest X-ray, ECG, echocardiography, catheterization, angiocardiography, pulse oxymetry and other diagnostic procedures were done for all patients.

Diagnosis and classification of TOF was done according to collected clinical and paraclinical data.

Results

Among 270 patients with TOF, 107 cases (39.63%) were female and 163 cases (60.37%) were male.

At the first admission, the mean age for females was 5.63 years (ranging from 6 months to 17 years), and for males was 6.56 years (ranging from 6 months to 17 years), and mean hemoglobin at first presentation was 16.69 g/dl (ranging from 10 to 24.3 g/dl) for all patients irrespective of sex and age (Table I).

With the exception of right aortic arch (RAA), coronary artery anomalies and patent foramen ovale (PFO), other associated anomalies were found in 98 cases (36.30%, Table II). Incidence of PFO, RAA and coronary artery anomalies were 44.81% (121 cases), 21.11% (57 cases) and 9.25% (25 cases), respectively (Table III).

Table I: TOF and demographic characteristic of patients.

Demography / Sex	Number	Percent	Diagnostic Procedures Echo & Cath		Patient's Age at the First Admission	Mean Hgb at First Admission
Female	107	39.63	107	100%	5.63 (4 mo-15y)	16.69 g/dl (10-24.30)
Male	163	60.37	163	100%	6.56 (6 mo-17y)	

Hgb=hemoglobin; mo=month; Echo=echocardiography; Cath=catheterization; y=year; TOF=Tetralogy of Fallot

Table II: TOF and associated anomalies.

Anomalies / No & Percent	ASD	PDA	LSVC	VSDs	AVSD	WPW	AI
Number	39	29	11	3	1	1	2
Percent (%)	14.44	10.74	4.07	1.11	0.37	0.37	0.74
Anomalies / No & Percent	SI Totalis	Interrupted-IVC		ABSENT			
		Azy-Co	HAzy-Co	LPA	RPA	PV	RSVC
Number	1	1	2	4	2	1	1
Percent (%)	0.37	0.37	0.74	1.48	0.74	0.37	0.37

AI=aortic insuffiency; ASD=atrial septal defect; AVSD= atrioventricular septal defect;Azy=azygos; H-Azy-Co= hemi-azygos-continuity; IVC=inferior vena cava; LSVC=left superior vena cava; LPA=left pulmonary artery; PDA=patient ductus arteriosus; PV=pulmonary valve; RPA=right pulmonary artery; RSVC=right SVC; SI=situs inversus; VSDs=multiple ventricular septal defects; WPW=Wolf-Parkinson-White syndrome; No=number

Table III: TOF and anomalies of coronary arteries.

Anomalies / No & Percent		D & L CB	SO CA	CB from LAD	LAD from RCA	LCX from RCA	Undetermined
Total	25	6	13	1	2	1	2
	9.26%	2.22%	4.81%	0.37%	0.74%	0.37%	0.74%

CB=conal branch; D&L CB=dilated and large CB; LAD=left anterior descending coronary artery; LCX=left circumflex coronary artery; RCA=right coronary artery; SOCA=single ostium coronary artery

Table IV: TOF and peripheral pulmonary stenosis (PPS).

PPS / No & Percent		Type I		Type II	Type III	Type IV
		LPA	RPA			
Total	38	8	6	22	0	2
	14.07%	2.26%	2.22%	8.15%	0%	0.74%

Among patients with coronary artery anomalies, single ostium coronary artery (SOCA) was the most common anomaly (4.81%).

It is mentionable that the left coronary artery (LCA) arose from the right coronary artery (RCA) in one case (0.37%), and RCA arose from left anterior descending artery (LAD) in 2 cases (0.74%).

There was peripheral pulmonary stenosis (PPS) in 38 cases (14.07%, Table IV).

Among patients with PPS, incidence of type I (single central stenosis) was 5.18%, type II (bifurcation stenosis) was 8.15% and type IV (central and peripheral stenosis) was 0.74%. There was no patient with type III PPS (multiple peripheral stenoses). With a view to blood grouping and Rh typing, 88.15% of cases were Rh^+ and 11.85% were Rh^-. The most common blood group among patients with TOF was type O (Table V).

Table V: TOF - blood groups and Rh types.

Patients		Groups Rh		A	B	AB	O
Number	270	+	238	65	60	24	89
		-	32	9	9	2	12
Percent	100%	+	88.15	24.08	22.23	8.88	32.96
		-	11.85	3.33	3.33	0.74	4.45
Total	Number		270	74	69	26	101
	Percent		100%	27.41	25.56	9.62	37.41

About 88 patients (32.60%) underwent palliative operation (modified Blalock-Taussig shunt) as the first operation (Table VI). The youngest case was 4 months old and the oldest was 17 years old, and the mean age of first palliative operation was 5.21 years. Corrective operation was done in 168 cases (62.22%) as the first operation. The youngest patient who underwent TC was 1.5 years old and the oldest was 17.5 years old (Table VI).

Transannular patch (TAP) was used in 136 cases (50.37%). With a view to post-surgical complications, mortality rate was 2.96%, postoperative RBBB (right bundle branch block) was present in more than 90% of patients. Complete heart block (CHB) was found in 7 cases (2.60%), in all of whom PPM (permanent pacemaker) was implanted. Other important complications such as hemorrhage and tamponade were seen in 11 cases (4.07), and all of them underwent re-operation.

Table VI: TOF - palliative and corrective operation.

Operation / Number, Age, Percent (%)	Shunt as First Operation								Total Correction (TC)		
	L-G	R-G	L+R	BT	Cent	R+Cent	L+Cent	R+L Cent	As the First Op.	As the Second Op.	TAP
Number	39	34	3	1	7	3	1	2			136=50.37%
Total	88 (32.60%)								168 (62.22%)	88 (32.6%)	
Age (y)	5.21 y (4 mo-17 y)								7.19 y (1.5-17.5)	6.43 y	

BT=Blalock-Taussig shunt; L-G=left Gore-tex shunt; Cent=central shunt; R-G=right Gore-tex shunt; L=left; R=right; Op=operation; TC=total correction; TAP=transannular patch; No=number; Y=year; ()=mean; mo=month

Table VII: TOF and postoperative complications.

Operation & Complications / Number, Percent (%)	MORTALITY		MORBIDITY					
			RBBB	CHB				Others
				Transient	Permanent+PPM			
	Shunt	T-C			Epicardial	Endocardial	Epi + Endocard	
No.	1	7	245	3	5	1	1	11
%	0.37	2.59	90.74%	1.11%	1.85	0.37	0.37	4.07%
Total	8 = 2.96%				7 = 2.60%			

Hemorrhage & Re-op=9; Tamponade =1; VSD patch dehiscence =1

CHB=complete heart block; PPM=permanent pacemaker; RBBB=right bundle branch block; Re-op=reoperation

Discussion

In our study, TOF was more common in males (male:female 3:2). With the exception of PFO, RAA and CA anomalies, other anomalies were found in 36.3% of patients; among them atrial septal defect (ASD) and patent ductus arteriosus (PDA) being the most common. PFO was found in 44.81%, RAA in 21.11% and CA anomalies in 9.25% of patients. Among CA anomalies, SOCA was the most common. PPS was found in 14.07% of cases. The most common type of PPS was bifurcation stenosis (Type II). TOF was more common in patients with type O blood group. Post-surgical mortality rate was about 1.13% (1 of 88) for palliative surgery and 2.73% (7 of 256) for TC. RBBB was very common. Interestingly, surgery- induced RBBB was less in one of our surgical teams. Other than RBBB, other complications were found in 21 patients (7.77%) after operation. The mean age at first palliative operation was about 5 years, and for TC without previous palliation was more than 7 years and for TC with previous palliative surgery was about 5.5 years.

Origin of LAD from RCA with anterior course across RVOT was found in 5% of TOF by 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 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 which focused on 938 patients from 12 institutions throughout the US 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). 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 another study by Mahle et al. primary complete repair (TC) of TOF was routinely performed in infancy.[17]

Many other lesions can coexist with TOF; PFO and ASD are common. A right aortic arch though not of functional importance, is common and when detected, it must alert us to the diagnosis of TOF.[18,19]

Stenosis within the pulmonary arteries themselves (PPS) is of major surgical significance and usually occur at branching sites from the bifurcation outwards.[20]

Absent pulmonary valve (APV) is a rare anomaly. This anomaly was seen in one of our patients (Table II). APV is usually associated with TOF, but has also been described with an intact ventricular septum (IVS), and is also commonly but not invariably associated with absence of ductus arteriosus.[21]

Because there is a trend toward earlier TC of TOF worldwide[1,2,12-14] due to improvement in the comprehensive surgical approach, technology and PICU care, single stage early TC of TOF 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 patients undergo surgery without invasive diagnostic procedures. Nonetheless, an invasive procedure is, on occasion necessary for determination of PA, CA and aortopulmoanry collateral arteries anatomy prior to deciding on the surgical or medical management strategies and for interventional treatment. We need more studies for definitive determination of relationships between TOF, blood grouping and HLA typing.

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