Original Article

THE CATHETERIZATION AND ANGIOGRAPHIC VARIATIONS IN IRAQI PATIENTS WITH TETRALOGY OF FALLOT

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Summary:

Background: *Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease and the catheterization and angiography still considered (in most centers) as essential preoperative diagnostic step. This retrospective aimed at evaluating the catheterization and angiographic finding in our Iraqi patients with diagnosis of TOF*

Patients and methods*: The catheterization and angiographic study of 200 patients with TOF referred to Ibn Al-Bitar Cardiac Center had been reviewed.*

Results: *There were 126 males and 74 females and their ages ranged from 11 months to 37 years. The catheterization data showed that 88% of the patients had equal LV and RV pressure. The evaluation of pulmonary artery pressure showed that all patients had normal pressure.The review of angiographic studies revealed that the perimembraneous VSDs were the most common type (91% of the patients). 13 patients (6.5%) had coronary anomalies, and the patent ductus arteriosus is the most common associated anomalies.*

Conclusion: *The frequency of doubly committed VSD and single coronary artery were higher than other studies and we showed there was no indication to measure the pulmonary artery pressure during catheterization.*

Keywords: *TOF, Catheterization, Angiography.*

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Introduction

At 1805, Louis Arthur Fallot made the precise anatomic diagnosis at bedside and proved by postmortem [1]. TOF is the most common form of cyanotic congenital heart disease. It represents about 8-10% of all congenital heart disease. The sex distribution is approximately equal or with slight male predominance. No specific etiologic agent has been identified and the Tetralogy is not concomitant of any genetically induced syndrome [2, 3]. TOF is characterized by 4 anatomical features :

(1) Ventricular septal defect (VSD).
(2) Pulmonary stenosis (PS).
(3) Overriding of aorta.
(4) Right ventricular hypertrophy (RVH).

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All agree that the VSD and pulmonary stenosis are essential to diagnosis, while the RVH is a consequence of elevated RV systolic pressure and not represent a congenital malformation. The four anatomical features of TOF are not coincidental but instead result from specific morphological abnormality which is the anterosuperior deviation of infundibular septum, which represent the anatomical hallmark or the "monology" of TOF with all the other components being secondary [4, 5].

PATIENTS AND METHOD

In this retrospective study, we have reviewed the catheterization and angiographic study of 200 patients with TOF, referred to Ibn-Albitar Cardiac Center, which is a tertiary center, from April 1993 to May 2001.

In this study, we included only those patients with classical TOF. We have excluded patient with TOF pulmonary atresia because of great clinical and anatomical differences between those two groups of TOF, we also excluded cases of large VSD (with large left to right shunt) and mild pulmonary stenosis and those with small VSD and pulmonary stenosis

because such situations are not true cases of cyanotic TOF.

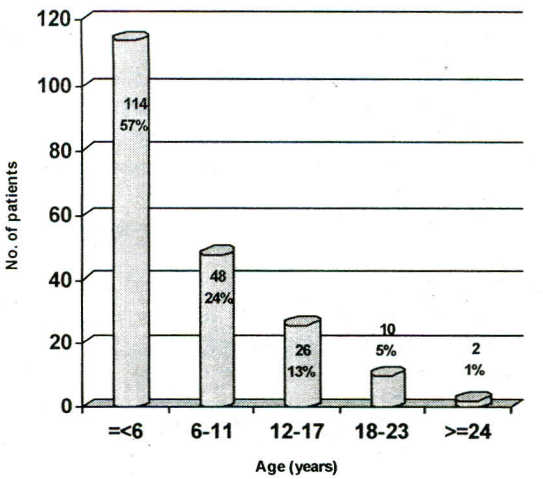
Catheterization data of all patients included in this study were reviewed. This procedure had been performed under general anesthesia in small and older children and local anesthesia with some sedation for adolescents and adults patients. For right heart catheterization, femoral vein access was used and from the right femoral vein, the catheter could be easily advanced into right atrium (RA), right ventricle (RV) and in some patients though the VSD into ascending aorta. Pulmonary artery (PA) engagement from RV done routinely and in some patients, despite repeated attempts the PA was not entered, and when there was defect in interatrial septum (ASD or PFO), catheter could be crossed to left atrium (LA) and left ventricle (LV). The pulmonary vein wedge pressure was sometimes recorded, as an indirect measurement of PA pressure, in those cases when catheterization of PA was unsuccessful. In most patients, left heart catheterization were done by catheter passed retrogradely from femoral artery to the aorta and then to the LV cavity. At catheterization, it is commonly possible to enter both great arteries and all four cardiac chambers. The pressure was measured by means of strain gauge manometer. Haemodynamic and saturation data were obtained from all patients before angiograms were done.

Angiograms were made for all patients. The RV angiography was performed with NIH catheter (side holes only) of different French size (according to patient age and size), and it was done in the two projections, anteroposterior and lateral view. The anteroposterior projection used with craniocaudal (up to 25o) for better demonstration of pulmonary trunk and proximal left and right pulmonary arteries and the addition of 5-10o of left anterior obliquity better profile the proximal left PA. The left heart catheterization was performed with pigtail catheter (5-6F). Left ventriculogram were selectively made in some patients when future definition of this chamber was desired, while the root aorotgrams done routinely and no selective coronary arteriograms were performed. Both the LV and aortic angiograms were performed in left anterior oblique projection, which defined as 60 to 70o left anterior oblique view combined with 20-30o craniocaudal angulation.

Both high osmolality ionic agents and low osmolality ionic agent (when available) contrast media were used in our hospital study in a dose of 1-1.5 ml/kg in each injection. Automatic injection was used routinely although hand injection occasionally used.

RESULTS

The studied group included 200 TOF patients. The patients' ages ranged from 11 months to 37 yeas. There were 126 males (63%) and 74 females (37%) patients. The patients distributed into different age groups, Figure 1&2.



Catheterization :

The catheterization data of all patients were reviewed for the following parameters :

Ventricular pressure:

the VSD is almost always large and nonrestrictive in TOF ensuring that the peak systolic pressure equals in both ventricles, in spite of this fact the differences between the systolic pressure of the two ventricles was not uncommon. In our series, the highest difference was 30 mm. Hg. These findings are shown in table (1).

Table (1): Ventricular pressure

Vent. pressure	No. of patients	Percentage
LV=RV	176	88
RV>LV	20	10
LV>RV	4	2
Total	200	100

Ventricular end diastolic pressure:

the left Ventricular end diastolic pressure (LVEDP), was elevated (>15mm.Hg) only in one patient while right Ventricular end diastolic pressure (RVEDP) was elevated >10mm.Hg in 5 patients. While both LVEDP and RVEDP were exceeding the upper limit of normal in 7 patients. None of these patients with abnormal intraventricular diastolic pressure presented any subjective or objective clinical manifestation of congestive cardiac failure.

Pulmonary artery pressure:

during cardiac catheterization the PA engagement and pressure measurement was done in 174 cases (87%) while in 26 patients (13%). PA could not be entered. From those with PA pressure data, pulmonary hypertension [mean PA pressure > 30 mmHg] discovered in 5 patients (2.8%), while in patients with no PA pressure measurement, the pulmonary vein wedge pressure were recorded in 7 patients and it was of normal value in all of them.

Angiography:

The angiography study was reviewed for the following findings.

Type of VSDs:

The type of VSDs in our patients summarized in table (2) as shown by angiographic views:

Table (2) : Type of VSDs

Type of VSD	No. of patients	Percentage
Perimembranous	182	91
Doubly committed	10	5
Inlet	3	1.5
AV canal	3	1.5
Muscular (single)	2	1
Total	200	100%

Pulmonary stenosis:

although stenosis at infundibular level is almost always presented in TOF patients and although the pulmonary valve is mostly abnormal, the basis of pulmonary stenosis was variable among the studies group, in 10 patients 5% the pulmonary valve was the only site of stenosis, while in 4 patients (2%) stenosis was present in the infundibulum only. In 2 patients with absent pulmonary valve the obstruction at the pulmonary annulus (pulmonary valve ring) and 184 patients (92%) both the valve and infundibulum were stenotic.

Table (3) : Level of pulmonary stenosis

Site of Pulmonary Stenosis	No. of patients	Percentage
Valvular	10	5
Infundibular	4	2
Pulmonary annulus	2	4
Valvular+ infundibular	184	92
Total	**200**	**100**

Pulmonary Trunk size:

the size of pulmonary trunk was evaluated both in echocardiography, angiography and in some patients at surgery. The caliber of pulmonary trunk was evaluated by comparing it's diameter to that of the aorta and classified as follows:

1- Normal: the diameter of pulmonary trunk was more than 80% of aorta.
2- Mild hypoplasia: diameter was 50-80% that aorta.
3- Moderate hypoplasia: diameter was between 30-50%.
4- Severe hypoplasia: the diameter less than 30% of that of aorta.
5- Dilated: the pulmonary trunk diameter larger than aortic diameter the distribution of our patients according to this classification shown in table (4).

Table (4): pulmonary trunk size

Pulmonary Trunk Size	No. of patients	Percentage
Normal	6	3
Mild hypoplasia	14	7
Moderate hypoplasia	126	63
Severe hypoplasia	50	25
Post stenotic dilatation	4	2
Total	**200**	**100**

Pulmonary branches:

in 195 patients the pulmonary arteries were confluent, while in 5 patients there were absent of left pulmonary artery branch. From the latter group there were 3 patients the left pulmonary artery could be demonstrated angiographically to (origin) from descending aorta through PDA. Five patients with confluent pulmonary arteries, there was stenosis at origin of the left (1 patient) and of the right (4 patients) pulmonary branch.

Collaterals:

In 5 patients (2.5%) significant broncho-pulmonary collaterals were detected.

Coronary arteries:

the coronary anomaly was recognized in 13 patients (6.5%) in 8 patients (4%) there was single coronary ostium. While left anterior descending coronary artery (LAD) arising from right coronary artery presented in 4 patients (2%) the one patient there was fistula between two coronary arteries and pulmonary.

Table (5) : Type of coronary anomalies

Type of anomalies	No. of patients	Percentage
Single coronary artery	8	4
LAD from RCA	4	2
Fistula between CA & PA	1	0.5
Total	13	6.5%

Associated anomalies:

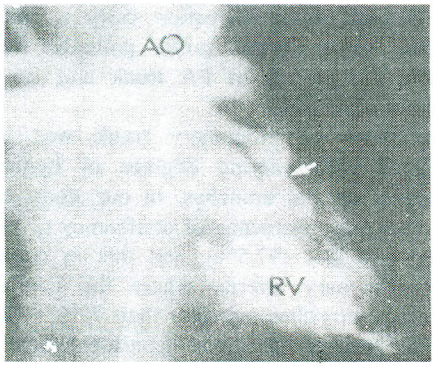
Patients had associated anomalies in addition to components of tetralogy.

Table (6) : Type of associated anomalies

Associated anomalies	No. of patients	Percentage
Patent Ductus Arteriosus	12	6
Left superior vena cava	7	3.5
Azygous in continuation of Inferior vena cava	2	1
Tricusped valve anomalies	1	0.5
Mitral valve anomalies	2	1

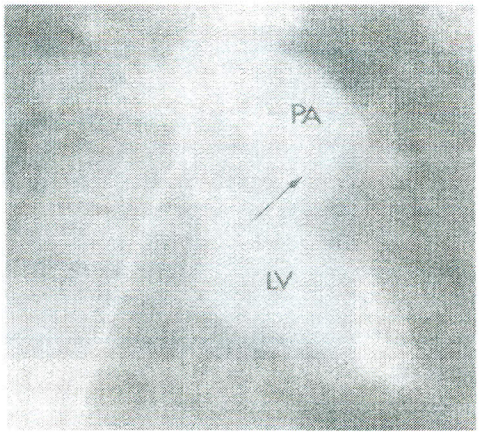
Table 7: Complications

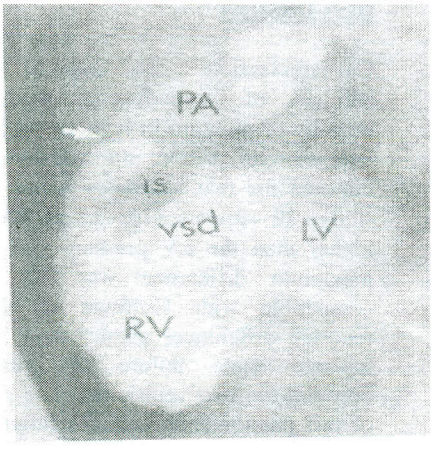
Complications	No. of patients	Percentage
Aortic regurgitation	11	5.5
Tricuspid regurgitation	10	5
Mitral regurgitation	5	2.5



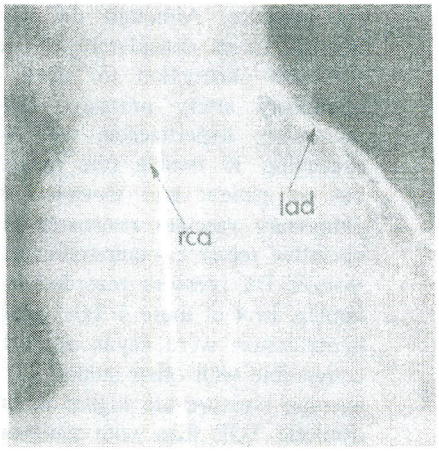
Frontal right ventriculogram

Right long axial oblique left ventriculogram



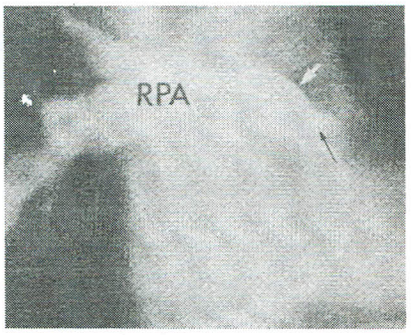


Left long axial oblique right ventriculogram



TOF with origin of left anterior descending coronary artery (lad) from right coronary artery

TOF with congental absence of left coronary artery (weight arrow)



DISCUSSION

Catheterization : In spite of the fact that the hallmark of haemodynamic study of TOF is the equality of pressure in both ventricles created by large unrestrictive VSD, the differences in pressure between the two ventricles is not uncommon. In this study, 20 patients (10%) had RV systolic pressure higher than the LV pressure and we found that the maximum difference was 30 mmHg, and this compatible with Hoffman et al. study (6) who reported differences greater than 30 mmHg probably indicate some failure of pressure equilibration between the two ventricles). The possible explanation of this phenomenon includes failure of rapid pressure equilibration across the VSD; occlusion of the defect by septal leaflet of TV; the defect is relatively small or the defect located in the infundibular chamber, but we could not specify the causes in this retrospective study (6).

The vast majority of our patients had normal PA pressure. Although the severity of pulmonic stenosis varies considerably, there is almost always sufficient obstruction to result in normal or low pulmonary artery pressure. Only 5 patients had pulmonary hypertension with mean PA pressure exceeding 30 mmHg (the maximum normal limit), but no patient had elevation of PA pressure of pulmonary vascular resistance to such a level that operative repair is contraindicated and the maximum systolic PA pressure recorded in this study was 45 mmHg. In 4 of these 5 TOF patients with pulmonary hypertension were acyanotic TOF and this finding compatible with other studies (7) who noted that the average pressure are higher in cases with mild or a cyanotic TOF than with patients with more classic TOF. The paradox of pulmonary hypertension with pulmonary stenosis explained by GOSUL and Coworkers (8) who reported that patient with unrestrictive large VSD with high pulmonary blood flow and as a natural history, may develop acquired infundibular pulmonary stenosis, and with the time the pulmonary arterial pressure remains at higher level (8).

Angiography: In spite that the clinical presentation of TOF patients, usually unrelated to the type of VSD associated with Tetralogy heart, the preoperative diagnosis of the type of VSD provide some basis for planning of appropriate surgical procedures. In our study (Table 2) we confirm that the large perimembranous sub-aortic VSD is the most common type (91.5%), we found that there was 10 cases (5%) with subarterial doubly committed VSD. This incidence is higher than that in series of Caucasian patients with TOF (3%) but our incidence still lower than that in Japanese TOF patients(10%) (9). The incidence of AV canal type VSD or the presence of additional muscular VSD (1.5%) is compatible with other series (10). No one of our patients with AV canal had Down's syndrome; the only patient with Down's syndrome included in this study had subaortic perimembranous VSD.

The level of pulmonary stenosis was determined in our study (table 3). Among most of our cases it was usual finding that stenosis at the infundibular and PV coexisted (92%). the high incidence of isolated valvular pulmonary stenosis in this study (5%), explained by the higher incidence of subarterial doubly committed VSD, in which the RV obstruction is mainly due pulmonic valvar stenosis. (11,12)

The pulmonary trunk caliber study in TOF patients in comparison to that of aorta are shown in table (4). Indicate that, certain degree of hypoplasia was almost always present, as a result of decrease of pulmonary blood flow. And normal pulmonary trunk size was exceptional. Two of those patients with post-stenotic dilatation, were diagnosed as TOF with congenital absent of PV and the other two patients had mild TOF (pink TOF). Fifty patients in our study have severe hypoplasia (PA less than 30% of Aorta) and about half of them had extreme hypoplasia with PA size less than 20% of the aortic caliber. And for such patients' total correction carry a considerable risk and some authors prefer palliative surgery to promote the growth of PA trunk and its branches before total correction.

Hypoplasia of pulmonary trunk was invariably associated with varying degrees of hypoplasia of pulmonary arterial branches. In our study, there had been very high incidence of confluency of pulmonary arterial branches (97.5%), and this in contrast with TOF pulmonary atresia, where the confluency of pulmonary branches was less than 70%, while in 20-30% of patients have non- confluent right and left pulmonary arteries (6). Among the patient with confluent pulmonary branches, few percentages

(2.5%) had stenosis at the origin of pulmonary artery branch (mostly the Right) and this incidence again less than the incidence with congenital pulmonary atresia (20%). In this study there were 5 patients with non confluent pulmonary arteries. In all of these patients there was discontinuous of left pulmonary artery with the pulmonary trunk (absence of the left PA). In 3 patients the left pulmonary branch demonstrate angiogrphically to be supplied by PDA, while in the other 2 patients it was absent, and not visualized angiographically. In such cases the left pulmonary artery, originally supplied by PDA, and with the closure of the ducts, the only flow to involved artery is via small collaterals, and overtime it becomes quit small in comparison with normally connected arteries. The higher incidence of involvement of left PA with such pathology related to the extension of the process of ductal closure to the left PA. Sometime the absent of one pulmonary branch due to thromosis of such branch.(12)

In TOF pulmonary stenosis the bronchial arteries tend to be enlarged but it rarely large enough to be detected angiographyically or to be considered as major collaterals, and this finding mimic those with TOF atresia. In TOF- pulmonary stenosis these collaterals was formed either in patient with congenital absence of PDA or the pulmonary stenosis was very severe, so the brachial arteries tend to exhibit extensive broncho-pulmonary anastemosis.(13,14)

In order to minimize complications resulting from anomalous coronary arteries, and to allow the planning of alternative surgical approaches, the course and distribution of the coronary arteries must be evaluated before operation. The reason for the high association of these anomalies with TOF is unknown. There was a wide variation in the incidence of coronary anomalies among different series (5 to 10%) depend on the type of the study. Low incidence usually reported by surgical team because of the difficulty to detect intraoperatively while postmortem examination indicates a higher incidence probably due to greater possibility of carefully evaluating of coronary arteries anomaly. Angiographic studies of coronary arteries in TOF represent the middle of these 3 methods (15).

In our series and by using aortic root injections "aortography" there was 6.5% of our patients had this anomaly (Table 5). Although this incidence still below the upper limit of the range it is higher than Kenneth study (16), who use selective coronary angiography routinely, and also below incidence of MC Manus et al study (14), and both recorded incidence of 3% . We found that the most common anomalies in our patients was the single origin coronary artery (4%), and the left anterior descending artery (LAD) from right coronary artery was the next common anomalies (2%).

The single coronary artery is relatively rare in other studies (15, 16, 17), this anomaly is the most common in our patients and consider as characteristic finding in our patients and should be taken in consideration. Because most of surgical repair in our center based on echocardiography (2D and colour flow Doppler) study, without routine preoperative angiography and because the surgeon often can define the coronary anatomy at the time of surgery, preoperative angiography to detect coronary anomalies routinely is not indicated, and should be limited only to cases with suspicion of anomalies on echo study, which in experts hand and good experience can suspect most of these anomalies.

Patency of the ducts was encountered in 16 patients(8%); 15 patients had left aortic arch and one patient only had Right aortic arch, and this confirm the old concept that PDA is almost always left sided structure.(12)

A left SVC draining into coronary sinus has no haemodynamic significant, but it may be important for the surgeon, because it requires special handling in extracorporeal circulation. LSVC draining directly to the left atrium is rare, and may result in persistent cyanosis after apparently successful repair of TOF. In this study the LSVC was present in 7 patients (3.5%) (6 detected by angiography and none at time of surgery) incidence is lower than other autopsy studies, where they reported incidence of 10.6% and 11% (12, 18).

In this study only one patient had bicuspid aortic valve, and two patients had mitral valve abnormality which confirm that the left sided lesion is rare in patient with TOF (19). Aortic incompetence coexists with TOF in 11 patients. The youngest patient was 5 years old and while more than half of these patient were above 12 years of age; one patient had history of infective endocarditis and only one had prolapse of right coronary cusp into the VSD. In all other patients, the incompetence results from massive dilatation of aortic root, which by itself progress with advancing age.

CONCLUSIONS AND RECOMMENDATIONS

Most TOF patients in our country were delayed in their presentation and referring for proper management.

The incidence of sub-arterial doubly committed VSD is higher in our patients than in cocusion patients but less than Japanese.

The most common coronary anomalies in our TOF patients are single origin coronary arteries.

Because the pulmonary artery pressure rarely, if ever, increased to a level that affect the decision of surgical repair, so the measurement of pulmonary artery pressure, which carries a risk, will add little, if any, to cath. study of TOF patients.

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