Clinical Features Of Iraqi Patients With Tetralogy Of Fallot

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Summary:

Background: *Tetralogy of Fallot is the most common cyanotic congenital heart disease which represents about 8-10% of all congenital heart diseases. It is characterized by four morphological features, large malalignment ventricular septal defect, Pulmonary stenosis, Overriding of aorta, and Right ventricular hypertrophy. The infant with TOF does well for the first few months of life with minimal or no cyanosis. The cyanosis begins to increase with secondary slow increase in polycythemia as well. Hypercyanotic spells are the most common complicating features of TOF*

Patients and methods: *This is a retrospective study of 200 patients with Tetralogy of Fallot (TOF) referred to Ibn Al-Bitar Center for Cardiac surgery from April 1993 to May 1999. The diagnosis was established by echocardiography, catheterization and angiographic study. For each patient, clinical history, physical examination, O2 saturation and hematocrit level had been reviewed.*

Results: *The patients' ages ranged from 11 months to 37 years. The weight of 42 patients (22%) and the height of 38 patients (20.5%) were below 3rd centile. There were 60 patients (30%) were polycythemic (hematocrit above 65%). Only 7 patients (14%) had low cyanosis (pink TOF) and one third of the patients had history of hypercyanotic spells. The CNS complications were the most common complications.*

Conclusions: *The study revealed that most of our patients had been delayed in their presentation for proper medical and surgical management. The study showed that most of TOF patients had normal growth pattern and the most important factor affecting the growth was the level of oxygen desaturation and we found high incidence of both infective endocarditis and CNS complications.*

Introduction:

At 1805, Louis Arthur Fallot made the precise anatomic diagnosis of the most common cyanotic congenital heart disease which represents about 8-10% of all congenital heart diseases. No specific etiologic agent has been identified and it is not concomitant feature of any genetically induced syndrome. Tetralogy of Fallot (TOF) is characterized by four morphological features [1, 2]:

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

Although the cyanosis may be present at birth, but usually the infant with TOF does well for the first few months of life with minimal or no cyanosis "as the PS is mild", and at 4-6 months of age "as the severity of the pulmonary stenosis increase", the cyanosis begin to increase with secondary slow increase in polycythemia as well. Evident clubbing usually takes a number of months to develop.As the child begins to walk, he will exhibit obvious exercise intolerance and it is common for children with TOF to squat or assume a position equivalent to squatting which is considered as a hallmark of TOF [2, 3]. Hypercyanotic spells are the most common complicating features of TOF and can occur both in patients with mild and deep cyanosis [4]. TOF also may be complicated by infective endocarditis and thromboembolic complications and it is considered as the most common cardiac malformation associated with brain abscess[5]. Most investigators indicate that about three quarters of patients born with TOF reach their first birthday and about 50% of patients with untreated TOF die by the age of 5 years and only 28% of them complete their first decade of life and 10% are alive at the age of 20 years, 5% at the age of 30 years and only 3% at the age of 40 years [6].

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Patients And Methods:

In this retrospective study, we reviewed the clinical data of 200 patients with TOF referred to Ibn Al-Bitar Center for Cardiac surgery from April, 1993 to May, 1999. Clinical data (including growth parameters), oxygen saturation and hematocrit level had been reviewed. The diagnosis of patients was established by echocardiography, catheterization and angiographic study, and we included only patients with classical TOF. We had excluded patients with TOF and pulmonary atresia because of

greta clinical differences between these two groups of TOF. We also excluded cases of large VSD (with large right-to-left shunt) and mild pulmonary stenosis and those with small VSD and pulmonary stenosis because such situations are not true cases of acyanotic TOF.

Results are expressed as median and range and the data were evaluated by paired t-test for individual paired comparison. The level of statistical significant was setup at $p<0.05$.

Results:

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

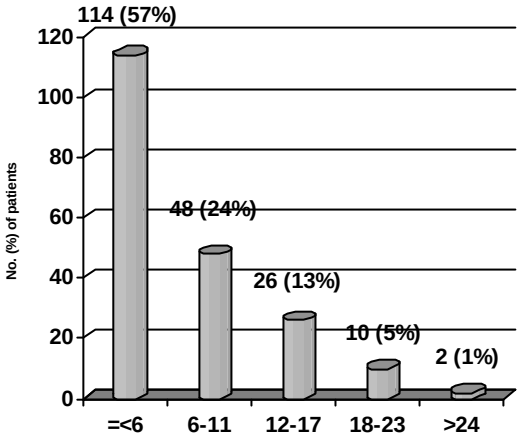


Figure (1) : Age group of the patients

Growth: The evaluation of general growth status of TOF patients included in this study done by measurement of weight and height of 188 patients preoperatively, mostly at time of catheterization.

Among 188 patients for whom the weight was reviewed there were 42 patients (22.5%) their weight below 3rd centile, while the weight of 146 patients (77.5%) above 3rd centile (table 1).

Table (1): Percentile of the patients' weight.

Percentile	No. of patients	Percentage
$\leq$ 3rd	42	22.5
3-97	139	73.8
> 97	7	3.7
Total	**188**	**100**

The height of 38 patients (20.3%) were below 3rd centile, while the height 150 patients (79.7%) above this centile (table 2).

Table (2): Percentile of the patients' height.

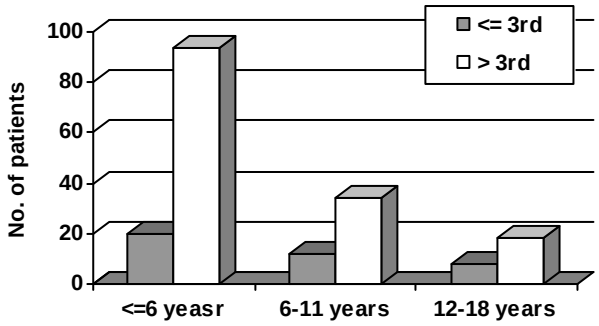
Percentile	No. of patients	Percentage
$\leq$ 3rd	38	20.3
3-97	143	76
> 97	7	3.7
Total	**188**	**100**

It is clear from tables (1 & 2) that the majority of patients (about 3/4th of patients) had normal growth record (above 3rd centile), and only relatively small group [(22.5% and 20.3%) for weight and height respectively] fall in the failure category (below 3rd centile). It is also clear from these two tables that weight and height are equally affected and the differences are statistically not significant.

The affection of growth (weight and height) increased with advancing age (table 3) and (figure 2).

Table (3): Growth percentile in age groups of the patients.

Age	$\leq$ 3rd	> 3rd	Total
$\leq$ 6 years	20 (17.5%)	94 (82.5%)	114 (100%)
6-11 years	14 (29%)	34 (71%)	48
12-18 years	8 (34.2%)	18 (65.8%)	26
Total	42	146	188



Fig(2): Growth percentile in age groups of the patients.

Oxygen saturation of arterial blood: The oxygen saturation of arterial blood were studied from catheterization data and we found that the majority of patients 87 patients (43.5%) had oxygen saturation between 65% to 85% and 52 patients (27.5%) below 65% (table 4).

Table (4): O_2 saturation of the patients.

Percentage of O_2 saturation	No. of patients	Percentage
≤ 65	52	26
65-85	87	43.5
≥ 85	61	32.5
Total	200	100%

In addition to the age, the other variable could affect the growth was the degree of oxygen desaturation of systemic arterial blood. (table 5)

Table (5): Weight percentile according to the O_2 saturation.

Percentile	Sat (≤ 65)	Sat. (> 65)	Total
≤ 3rd centile	20 (47%)	22 (53%)	42
> 3rd centile	32 (22%)	114 (78%)	146
Total	52	136	188

The p-value = 0.007 which is highly significant.

Table (6): Height percentile according to the O_2 saturation.

Percentile	Sat (≤ 65)	Sat. (> 65)	Total
≤ 3rd centile	18 (47.3%)	20 (52.7%)	42
> 3rd centile	34 (22%)	116 (78%)	146
Total	52	136	188

The p-value = 0.003 which is highly significant.

So the most important factors influencing the growth are the age and the O_2 saturation.

Hematocrit (PCV): Sixty patients had hematocrit level (PCV) above 65%; 24 patients had PCV level less than 45% while the majority (116 patients) had PCV range from 45 to 65%. The highest PCV recorded was 85% (table 7).

Table (7): Hematocrit level of the patients

PCV	≤ 45%	45-65%	≥ 65%	Total
No. (%) of patients	24 (12%)	116 (58%)	60 (30%)	200 (100%)

Cyanosis: Physical examination of the studied group revealed that among the 200 patients, 14 patients (7%) had no cyanosis and classified as acyanotic or pink TOF, while all other patients had variable degree of cyanosis with clubbing of fingers and toe nails (table 8).

Table (8): percentage of Cyanosed patients.

Cyanosis	Positive	Negative	Total
No. (%) of patients	186 (93%)	14 (7%)	200 (100%)

Spells: Among the 200 patients included in this series, 76 patients (38%) had history of hypercyanotic spells and in most patients, the spells were severe enough to search medical treatment. We did not found relation between the history of spells and the degree of oxygen desaturation of systemic arterial blood. The p-vale=0.074 which is not significant**.**

Complications: the most common complication in studied patients were infective endocarditis which developed in 12 patients (6%) and CNS complications in 10 patients (5%) (table 9).

Table (9): Complications in TOF patients

Complications	No. of patients	Percentage
Infective endocarditis	12	6
Brain abcess	5	2.5
Cerbrovascular accident (CVA)	5	2.5

Discussion

The sex distribution of patients included in this study confirm the previous fact that there was male predominance and the age distribution consistent with natural history of TOF, and indicate the late presentation of most of our patients in seeking for proper management [(6)].

The evaluation of growth of our patients, which was done preoperatively revealed that majority had good growth status, and only minority of these patients had poor growth, also this study showed that both weight and height had the same degree of affection. The growth of TOF patients (height and weight) are directly related to the degree of systemic arterial oxygen desaturation and the p-value was 0.007 and 0.003 for weight and height respectively. These results are compatible with Thomas P.G.[(7)] review who reported that "poor weight gain is seldom a problem in TOF patients" and with George W Land study[(8)] who wrote (this is comparison with non cyanotic congenital heart disease with significant left to right shunt where most patients had very poor weight gain). The progressive growth adverse affection with advancing age can be explained on the basis of increasing degree of obstruction to pulmonary blood flow with increasing the degree of systemic arterial oxygen desaturation. So the most important factor influencing the growth and the decision of

surgical repair is the degree of O_2 saturation of systemic arterial blood.

The physical examination of our patients confirm that the cyanosis was the most frequent finding (93%) and the remaining cases (7%) had no cyanosis (pink TOF). The percentage of patients with pink TOF in our series is lower than that reported by Wood et al.[9], who recorded incidence of 10% and this may indicate the tendency for delay seeking of medical advice and for referring the patients with mild symptoms. In our series, one third of patients had hypercynantic spells, and the absence of correlation with the degree of systemic arterial oxygen desaturation indicate that spells occurred both in mild and in severe form of TOF.[4, 10]

We found a wide range of oxygen saturation (50%-98%) which indicates the wide variation in the severity of Tetralogy of Fallot and there was a direct relation between the degree of oxygen saturation of systemic arterial blood and the level of hematocrit which range from 40 to 85%. The high percentage of patients with hematocrit level of more than 65%, indicate the delay of surgical treatment of most of our patients.

There is a high incidence of infective endocarditis as a comparison incidence of other studies 2.3% and this indicates loss of orientation about the importance of antibiotic prophylaxis against infective endocarditis both by the physician, dentist and patients. The central nervous system complications also considered higher in our patients in relation to other studies and this explained of the bases of the delayed in surgical correction lack of proper medical care of non operated patients and the high percentage of patients with polycythemia in which there is high risk of thrombosis and bleeding [5,10].

Conclusions And Recommendations:

1. Most of TOF patients in our country were delayed in their presentation and referring for proper medical and surgical management.
2. In addition to the age of the patients the level of arterial O_2 desaturation is the most important predicting factor for the possibility of growth failure and the decision of proper time of surgery.
3. There was high incidence of CNS complications due to the large number of patients with polycythemia and delayed surgical management of the patient.

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