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Tetralogy of Fallot: Epidemiological, Clinical and Management Aspects of 56 Cases at the Ignace DEEN National Hospital in Conakry

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Abstract

Introduction: Tetralogy of Fallot (TF) is a cyanogenic congenital heart defect. It comprises 4 distinct anatomical elements. **Objective:** The aim of this study was to describe the population of patients operated on for tetralogy of Fallot at the Ignace Deen National Hospital (IDNH), **Méthodologie:** A retrospective study was carried out. It included 56 patients with tetralogy of Fallot (TF) collected over a period of 3 years in the cardiology department of IDNH. **Résultats:** At the end of our study, we found that FT preferentially affects males, with a male/female sex ratio of 2:1. The mean age of onset was 5.5 years. The 1 - 5 year age group was the most represented, with extremes of 03 months and 16 years. The mean age of onset was 9 months. Consanguinity was the most common risk factor. The majority of patients (32) diagnosed with FT had undergone surgery within a year of diagnosis, with delays ranging from 1 to 6 months. The mean age at the time of surgery was 4.8 years. Moderate pulmonary insufficiency was the most frequent postoperative complication. Immediate postoperative mortality was low (3.1% or one patient). Conclusions: Tetralogy of Fallot is the most common cyanogenic congenital heart defect. A multidisciplinary approach involving obstetricians, pediatricians and cardiopediatricians is required for early detection and management, with the aim of improving vital prognosis.

Keywords

Congenital Heart Disease, Cyanogenic Heart Disease, Tetralogy of Fallot, Surgery

1. Introduction

First described in 1888 by Etienne-Arthur Louis Fallot, Tetralogy of Fallot (TF) is the most common cyanogenic congenital heart disease [1].

It comprises 4 distinct anatomical elements: Pulmonary tract stenosis (which may be infundibular, valvular or commonly both), so-called outlet or cono-ventricular septal defect (VSD) due to septal misalignment, aortic dextroposition <50% and right ventricular hypertrophy secondary to right ejection tract obstruction [2].

It affects approximately 1 newborn per 2,700 - 3,200 live births and accounts for 4-10% of congenital heart defects worldwide [3].

In Cameroon, Ndongo-Amougou *et al*. in Yaoundé in 2022 predominance of cases of tetralogy of Fallot in their study conducted in Yaoundé in 2022, accounting for 13.3% of cases [4].

In the Ivory Coast in 2019, in a study describing the epidemiological, diagnostic, therapeutic and evolutionary aspects of congenital heart disease in children, the prevalence of tetralogy of Fallot was 11.5% [5].

In Guinea, Bah MB found a prevalence of congenital heart disease of 58% and tetralogy of Fallot, a cyanogenic heart defect, was the most common, with a frequency of 27.4% [6].

Echocardiography is currently the reference test for diagnosing FT, regardless of the patient's age. The development of this non-invasive test means that this heart disease can also be diagnosed antenatally and postnatally [7].

Complete surgical repair of tetralogy of Fallot improves the survival rate in 90% of cases [7], with an operative mortal [8] [9].

In Guinea, with the support of humanitarian non-governmental organizations (NGOs) in collaboration with referring cardiologists, several advances were made between 2018 and 2024, not only in the early diagnosis of tetralogy of Fallot, but also in its surgical management.

There is little data on the results of these procedures and on patient follow-up after surgical correction, hence the initiation of this study.

2. Methodologies

2.1. Type and Duration of Study

We conducted a retrospective, descriptive, longitudinal study of the records of 56 patients aged 0 - 17 years followed in the cardiology department of the Hôpital national Ignace Deen over a 3-year period from 1 January 2020 to 31 December 2023.

2.2. Inclusion Criteria

Children aged 0 - 17 followed up for tetralogy of Fallot confirmed by cardiac ultrasound, whether or not they have undergone surgery.

2.3. Non-Inclusion Criteria

Patients whose records are not properly completed.

2.4. Sampling

We RECRUITED all the children who were followed during the study period and who met our selection criteria.

2.5. Study Variables

The parameters studied were divided into the following variables: sociodemographic, clinical, paraclinical and therapeutic aspects. We have reported on the medical and/or surgical treatment of patients, their post-operative follow-up and their natural history.

We classified the patients into 03 groups: those who had undergone surgery, those awaiting surgery and those who had been refused surgery. In the group of patients who underwent surgery, we reported the age at which the procedure was performed, the surgical technique used and the postoperative complications.

2.6. Data Management and Analysis

Data analysis and processing were carried out using EPI info software version 7.2.3 and Microsoft 365 for data entry, table and figure design. References are managed by Mendeley reference manager.

2.7. Consent and Ethics

We have obtained authorization from the ethics committee of the Ignace DEEN National Hospital to access patient records.

3. Results

Tetralogy of Fallot accounts for 17.5% of all congenital heart diseases.

The average age of discovery was 5.5 years. The 1 - 5 year age group was the most represented, with extremes of 03 months and 16 years. There was a male predominance, with 38 boys and 18 girls (**Table 1**). The most frequently reported aetiological factors were consanguinity (17.8%). Clinical signs were dominated by cyanosis (92.9% of cases) and we note that 23.2% of our patients had already had anoxic crises at the time their heart disease was diagnosed (**Table 2**). Polycythemia was present in 32 patients (58.2%) (**Table 3**). On chest X-ray, 96.4% of patients had a cloven-hoofed appearance and 91.1% presented with pulmonary hypovascularisation (**Table 4**).

We noted 69.6% of cases of regular Fallot and 30.4% of irregular Fallot.

Cardiovascular malformations associated with TF were patent foramen ovale (14.3%), followed by coronary sinus or coronary artery anomaly (12.5%), atrial septal defects (5.4%) and patent ductus arteriosus (5.4%) as shown in **Table 5**. The spontaneous evolution of our patients was marked by one case of death before transfer for surgery due to anoxic malaise. There was also one case of ischaemic stroke.

The majority of patients, *i.e.*, 32 patients diagnosed with tetralogy of Fallot, had undergone surgery within a year of diagnosis, with delays ranging from 1 month to

6 months. Three patients were excluded from surgery: one with ischaemic stroke, one with trisomy 21 and one with Allagile's syndrome as shown in Table 6.

Table 1. Socio-demographic characteristics.

Age groups		Number of cases (N = 56)	Percentage %
-1 year		1	1.8
1 - 5 years		30	53.6
6 - 10 year		17	30.3
11 - 15 years		6	10.7
+ 15 years		2	3.6
Gender	Men	38	67.9
	woman	18	32.10

Table 2. Clinical signs.

Reasons for consultation	Number of cases (N= 56)	Percentage %
Cyanose	52	92.9
Repeated infection	37	66.1
Shortness of breath when feeding or exercising	52	92.9
Difficulty eating	44	78.6
Retard staturo-pondéral	11	19.6
Squatting	38	67.9
Palpitation	6	10.7
Syncope	10	17.9
Anoxic symptoms	13	23.2

Table 3. Chest X-ray.

Chest X-ray	Number of cases (N = 56)	Percentage %
Clog heart	54	96.4
Hypo pulmonary vascularization.	51	91.1
Normal vascularization	5	8.9

Table 4. Distribution of patients according to biological results (based on hemoglobin level)

Biology (depending on hemoglobin)	Number of cases (N = 55)	Percentage %
Polycythemia	32	58.2
Anemia	8	14.5
Normal hemoglobin level	15	27.3
TOTAL	55	100

Table 5. Distribution of patients according to frequency of association with other congenital heart defects.

Associated congenital anomalies		Number of cases (N = 56)	Percentage %
Aortic Arch	Left	44	78.6
	Right	12	21.4
Patent foramen ovale		8	14.3

Continued

Coronary or coronary sinus anomaly	7	12.5
Communication inter atrial	3	5.4
Persistence of the atrial channel	3	5.4
Left superior vena cava anomaly	1	1.8

Table 6. Surgical management.

Surgical management	Number of cases (N = 56)	Percentage %
Patients operated on	32	57.1
Patients awaiting surgery	21	37.5
Patients refused surgery	3	5.4
TOTAL	56	100

All our patients underwent a complete cure, 96.9% of them as a first indication. Only one patient (3.1%) underwent palliative treatment with a right Blalock-type systemic-pulmonary anastomosis as a first-line treatment as soon as he arrived with severe anoxic symptoms. After stabilisation of his clinical condition, a complete cure was performed.

Enlargement using a transannular patch with a split pulmonary ring was performed in 13 patients (40.6%) (**Figure 1**). All the children were reassessed in the month following surgery before their return to Guinea, one month after their return and then at 3 months.

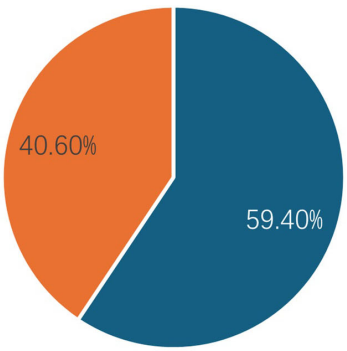

Figure 1. Lung enlargement technique used.

Follow-up was straightforward in the majority of patients (71.9%). Early post-operative complications were dominated by pulmonary insufficiency. They regressed before their return home. Two patients had small pleural effusions on their return with a good clinical evolution without symptoms. We noted one case of post-operative death due to septic shock.

4. Discussion

In a context where data is extracted directly from medical records retrospectively,

we are faced with information bias, selection bias and misclassification bias.

This study was conducted over a 3-year period from 1 January 2020 to 31 December 2023 and involved 56 patients with tetralogy of Fallot, with a prevalence of 33.7%. This result is double that found by BAH MB *et al* in their study in 2022 with 27 cases of tetralogy of Fallot [6]. This increase can be explained by the longer duration of our study, but also by the improvement in diagnostic management and the almost systematic referral of children with congenital heart disease to our department.

In our cohort, the mean age of diagnosis was 5.5 years, with extremes of 03 months and 16 years; this advanced age is significantly higher than that found in Western series [10] [11]. This may be explained by the low number of paediatric cardio units in Guinea, but also by socio-economic constraints preventing rapid consultation in specialist centres. Consanguinity is the aetiological factor most often found in our patients. It was reported in 17.8% of cases in our series.

Recent genetic studies have revealed more specific aetiologies, with the discovery of either a chromosomal abnormality or a genotypic mutation [12].

Functional signs are constant in our series. These signs only lead parents to consult specialised centres when they become obvious. This explains the average age of consultation (5.5 years).

In Western series, it has become rare for the diagnosis to be made in the presence of these clinical signs, given that the diagnosis is made either antenatally, thanks to the development of ultrasound, or in the neonatal period, thanks to systematic screening by simple auscultation of all newborns.

Echocardiography coupled with Doppler confirmed the diagnosis of Tetralogy of Fallot in all our patients and in all foreign series.

The VIC was conotruncal due to malalignment in all patients, in accordance with the literature [12]. Cardiac Doppler ultrasound can also be used to identify cardiovascular abnormalities associated with Tetralogy of Fallot.

Cardiac Doppler ultrasound can also be used to determine the shape of Tetralogy of Fallot, which is essential for choosing the surgical technique. In this study, the regular shape was predominant with 69.6% of cases. Over the years, developments in surgical methods and knowledge have improved the quality of treatment [13]. Survival rates are now reported to easily exceed 90% [13]-[15]. However, despite improving conditions, patients with FT are at risk of numerous complications that can lead to surgery, hospitalisation and mortality in the most severe cases [16]. In our cohort, the spontaneous evolution was fatal for a patient who died at home following a severe anoxic episode. The definitive treatment is open-heart surgical repair, which is indicated in all cases. The age at which it is performed electively has decreased over the years. Currently, the ideal age for elective surgery is between 3 and 11 months of age [17]. Some even perform it on newborn babies [18] [19].

Medical treatment and palliative surgery, which postpone definitive repair, are therefore of marginal importance today, except in cases of pulmonary atresia or

hypoplasia of the pulmonary arteries [12]. According to the therapeutic project, we have classified our patients into three groups:

The commitment of humanitarian NGOs and referral doctors in the management of this pathology made it possible to operate on 32 cases, *i.e.*, 57.1%.

Our result is superior to that of BAH MB *et al.* in 2022 who had 12 cases (32%) of tetralogy of Fallot operated on [6]. This can be explained by the longer duration of our study, but also by our larger cohort. Only one patient underwent palliative treatment with a right Blalock-type systemic-pulmonary anastomosis as a first-line treatment on arrival for severe anoxic malaise complicated by cardio respiratory arrest. One month after his condition had stabilised, a complete cure by sectioning the ring and widening the pulmonary tract was performed. The vast majority of our patients (96.9%) underwent surgical correction as first-line treatment, which is corroborated by the literature showing a trend towards greater use of this type of approach [17] [18]. The surgical technique for complete correction may involve either placement of a transannular patch (TAP) from the infundibulum of the VD to the pulmonary bifurcation to widen the stenosis of the VD outflow tract, or a technique to preserve the annulus and pulmonary valve without TAP [18] [19].

The use of a TAP currently depends on the degree of narrowing of the pulmonary annulus [18] [19].

Our analysis shows that the majority of patients benefited from preservation of the annulus. This choice could be explained by the fact that placement of a TAP has a major impact on the VD, which goes from a state of hypertrophy due to stenosis of the pulmonary tract to a state of significant volume overload with the removal of obstruction of the right outlet tract via this patch, thus creating significant pulmonary insufficiency. In our study, we found only two cases (6.25%) of junctional arrhythmia, which resolved rapidly after administration of antiarrhythmic drugs, with no recurrence reported.

Immediate postoperative mortality was 3.1% (one case) in our cohort. This result highlights the advances made in the surgical management of our patients with tetralogy of Fallot, with a positive impact on improving vital prognosis.

However, in order to be able to identify the variables that are important for post-operative results, it would be interesting to assess the outcome of these patients through a long-term study, analysing their progress, late complications and their various causes.

5. Conclusions

Thanks to the active involvement of medical NGOs and referring cardiologists, we can see that progress has been made in the surgical management of patients with tetralogy of Fallot, significantly reducing the short- and medium-term mortality of this condition, which has a poor prognosis in the absence of surgical treatment. However, despite the excellent results of paediatric cardiac surgery in the correction of Tetralogy of Fallot, these patients need regular long-term follow-

ups, in order to detect any complications at an early stage and organise their management.

The establishment of a cardiac surgery centre in our country and the creation of social security cover for all would contribute to reducing the morbidity and mortality associated with this condition and would help to limit dependence on healthcare.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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