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Epidemiological Profile of Cyanotic Congenital Heart Disease in the "B" Surgery Department of Point G University Hospital, before the Advent of Extracorporeal Circulation

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Abstract

Aim: Evaluate the epidemiological profile of cyanotic congenital heart disease in Mali before the advent of extracorporeal circulation in the "B" Surgery Department at the Pont G University Hospital. **Patients and Methods:** This was a retrospective and descriptive study that took place from January 1, 2011 to December 31, 2017. The records of patients with cyanotic congenital heart disease in the "B" surgery department of Point G University Hospital were collected. Patients operated on for cyanotic congenital heart disease were included in this study. Non-operated patients were not included. **Results:** The records of 17 patients operated on for cyanotic congenital heart disease were retained. The average age of patients at the time of surgery was 5.18 years with extremes of 2 and 18 years. Boys were in the majority with 59%, *i.e.* a sex ratio of 1.42. Patients resided in Bamako in 82% of cases. Inbreeding was found in 35.3%. Eighty-eight percent of children were born at term and 94% had up-to-date vaccination status. The average duration of patient follow-up between diagnosis and surgical management was 5 years with extremes of 2 years and 12 years. Tetralogy of Fallot regular form was the most represented heart disease. **Conclusion:** Cyanogenic congenital heart disease remains the most frequent congenital pathologies in our country. They most often affect male children. Consanguinity is the most common etiological factor found. Tetralogy of Fallot regular form remains the most common.

Keywords

Epidemiology, Congenital Heart Disease, CHU Point G, Mali

1. Introduction

Cyanogenic congenital heart disease (CCC) is a large group of malformations with very different anatomopathology, treatment and prognosis. They have in common an oxygen desaturation of the arterial blood, responsible for cyanosis [1].

The incidence of congenital heart disease is estimated at 6 - 8 per 1000 live births [1]. They account for approximately 25% of all congenital malformations [1].

Cyanotic congenital heart disease accounts for one-third of all congenital heart disease [2].

Thanks to advances in pediatric cardiology, congenital heart disease is being diagnosed earlier and earlier. In developed countries, some forms are even diagnosed prenatally, and 40% to 50% during the first weeks of life [3].

They are serious and constitute one of the main causes of neonatal morbidity and mortality [2].

Their prognosis has been considerably improved in recent years by advances in cardiac surgery. However, while some malformations can be repaired very satisfactorily from an early age, others are only accessible to palliative treatment, for which many uncertainties persist on the medium and long-term evolution. [4].

Our service has had a heart surgery culture since 1978 and operates on certain cyanotic congenital heart diseases with a beating heart due to the absence of extracorporeal circulation.

Few studies have been carried out on the management of cyanotic congenital heart disease in Mali, which is why we have initiated this study to determine the epidemiological profile of patients operated on for cyanotic congenital heart disease [5] [6] [7].

2. Methodology

This was a retrospective and descriptive study that took place from January 1, 2011 to December 31, 2017. The records of patients with cyanotic congenital heart disease in the "B" surgery department of Point G University Hospital were collected. Patients operated on for cyanotic congenital heart disease were included in this study. Non-operated patients were not included in the study. The comparison test was the Chi2 test and the probability $p < 0.05$ was considered significant. The parameters studied were hospital frequency, sex, age, geographical origin, frequency of consanguinity, type of congenital heart disease, average duration of patient follow-up between diagnosis and surgical management.

3. Results

The records of 17 patients operated on for cyanotic congenital heart disease were collected. These cyanotic congenital heart diseases accounted for 21% of operated congenital heart diseases (82 cases), and 0.73% of all surgical interventions (n = 2322).

Fifty-nine percent of children operated on were boys and the sex ratio was 1.42 in favor of them.

The average age of patients at the time of surgery was 5.18 years with extremes of 2 and 18 years (Table 1).

Patients came from inside Bamako in 82.4% of cases (n = 14) and from the city of Ségou in 5.8% (n = 2) (Table 2).

The Fulani ethnic group was the most represented (Table 3).

Inbreeding was found in 35.3% of patients (6 cases) and was absent in 58.8% (10 cases). It was unknown in 1 case (5.9%).

Eighty-eight percent of patients were from term pregnancies (Table 4) and

Table 1. Patients according to age groups.

Age groups (year)	Numbers	Percentage
0 - 2 years	2	11.8
2 - 5 years	7	41.2
5 - 10 years	5	29.4
>10 years	3	17.6

Table 2. Distribution of patients by residence.

Residence	Number	Percentage
Bamako	14	**82.4**
Ségou	2	11.8
Koulikoro	1	5.8
Total	**17**	**100**

Table 3. Patients according to ethnicity.

Ethnic group	Number	Percentage
Peulh	**6**	**35.3**
Bambara	4	23.5
Soninké	3	17.6
Dogon	1	5.9
Malinké	1	5.9
Minianka	1	5.9
Sonrhaï	1	5.9
Total	**17**	**100**

Table 4. Patients by term of pregnancy.

Terme	Number	Pregnancy
Term birth	**15**	**88.2**
Premature	1	5.9
Exceeded term	1	5.9
Total	**17**	**100**

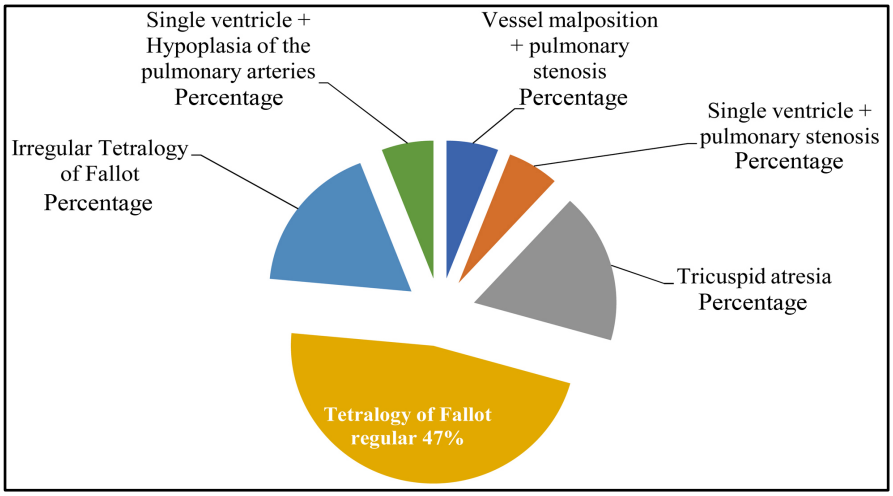

Figure 1. Breakdown by type of heart disease.

vaccination was up to date in 94% of patients.

Regular tetralogy of Fallot, irregular tetralogy of Fallot and tricuspid atresia were the most common heart diseases (**Figure 1**).

4. Discussion

The hospital prevalence of cyanotic congenital heart disease was 21% of operated congenital heart disease and 0.73% of all surgical procedures.

This result is superior to that reported by Ndongo-Amougou S *et al.* in Yaoudé, Diby Kouakou F *et al.* in Ivory Coast, Cloarec *et al.* in France who respectively reported a hospital prevalence of 8.7%, 2.3% and 9.8% [2] [8] [9].

The variations in prevalence observed in the literature would be the consequence of differences in means of diagnosis and methodology. In addition, the reported prevalences are estimates. Indeed, bicuspid aortic valve disease, by far the most common congenital heart disease with a prevalence of 1.5% in the general population, is usually not counted because it is most often diagnosed in adulthood [10].

In our study, 59% of operated were boys and 41% were girls. Thus, the sex ratio was 1.42 in favor of boys.

Moons P *et al.* in Belgium reported an identical distribution between girls (51%) and boys (49%) [11]. However, for congenital heart disease with recognized severity (persistent ductus arteriosus, Ebstein's disease, tricuspid atresia,

tetralogy of Fallot, pulmonary atresia, transposition of the great vessels, common arterial trunk, single ventricle, total pulmonary venous return), a male preponderance was noted with a male/female ratio of around 1.5 [11]. Robert-Gnansia E *et al*. reported in the Lyon registry a sex ratio of 1.45 [1]. There may therefore exist a sex ratio different from 1, with variable values, for certain congenital heart diseases (**Table 5**) [12]. These differences have no clear explanation to date. Somerville J insinuated several potential factors (**Table 6**) but not validated [13]. A preponderance is noted in girls for interatrial communication and the atrioventricular canal [13] [14]. The preponderance noted in boys mainly concerns abnormalities of the aortic valve including bicuspid valve, aortic coarctation,

Table 5. Preponderance by sex for the main congenital heart diseases [12].

Anomalies	Preponderance by sex	Estimated ratio
Female interatrial communication	Feminine	7/10
Female atrioventricular canal	Feminine	6/10
Persistent ductus arteriosus	Feminine	7/10
Left ventricular outflow tract obstructions		
At the aortic valve level	Male	7/10
Below the aortic valve	Male	7/10
aortic coarctation	Male	7/10
tricuspid atresia	Male	7/10
Tetralogy of Fallot	Male	6/10
Pulmonary atresia with ventricular septal defect	Male	6/10
Transposition of the great vessels	Male	7/10
Corrected Transposition of Large Vessels	Feminine	7/10
Common arterial trunk	Male	7/10
Single ventricle	Male	8/10
Pulmonary venous return abnormalities	Male	6/10
Male coronary abnormalities	Male	7/10

Table 6. Possible factors that may explain gender differences in congenital heart disease [13].

Possible factors
Biological differences
Smaller size of heart chambers and vascular vessels in women
Different endothelial vascular physiology
Pregnancy
Influence of gene expression by sex hormones
Different expression of genetic polymorphism according to gender

tetralogy of Fallot, transposition of the great vessels and hypoplastic left ventricle [13] [14] [15]. Ventricular communications and pulmonary valve stenosis are observed in both girls and boys. The male preponderance for pathologies concerning the aortic valve and the aorta is poorly understood. The high frequency of aortic abnormalities in patients with a malformation syndrome involving the absence of a second normal X chromosome suggests that a genetic factor modulating aortic development could be located on the X chromosome [16]. The sex ratio is highly variable among the obstacles to the left ventricular outflow tract: with a 4/1 ratio in favor of boys for valvular stenosis including bicuspid valve, a 2/1 ratio in favor of boys for infra stenosis-valvular, while the ratio is close to 1 for supra-valvular stenoses.

The average age at the time of the intervention in our study was 5.18 years. This delay in care could be explained by the lack of technical equipment and the lack of financial means of the parents of sick children. It is currently lower and lower with the improvement of surgical techniques and constitutes a factor of good results because PAH has little chance of being fixed before the age of 6 months [17] [18] [19].

Inbreeding is a risk factor for congenital heart disease. In our study, it was noted in 35.3% of patients. Nazari P [20] and Majeed-Saidanam [21] reported 48.7% and 49.6% respectively.

Premature babies are often at risk of developing various heart conditions including congestive heart failure and congenital heart defects.

The prematurity rate in our study was 5.9%.

5. Conclusion

Cyanogenic congenital heart disease remains the most frequent congenital pathologies in our country. They most often affect male children. Consanguinity is the most common etiological factor found. Tetralogy of Fallot regular form remains the most common.

Conflicts of Interest

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

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