Scientific Research Publishing

Open Journal of Pediatrics, 2022, 12, 857-866
https://www.scirp.org/journal/ojped
ISSN Online: 2160-8776
ISSN Print: 2160-8741

Congenital Heart Disease in Newborns: Epidemiological and Clinical Particularities in a Neonatology Department in Mali

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How to cite this paper: Doumbia, A.K., Koné, O., Dembélé, A., Sidibé, N., Banou, N., Diall, H.G., Togo, P., Coulibaly, O., Sacko, K., Konaté, D., Sidibé, T.S.L.N., Diakité, F.L., Diarra, B., Dembélé, G., Maïga, B., Cissé, M.E., Konaré, H., Doumbia, A., Ahamadou, I., Diakité, A.A., Traoré, F.D. and Tog, B. (2022) Congenital Heart Disease in Newborns: Epidemiological and Clinical Particularities in a Neonatology Department in Mali. *Open Journal of Pediatrics*, **12**, 857-866.
https://doi.org/10.4236/ojped.2022.125087

Received: October 21, 2022
Accepted: November 26, 2022
Published: November 29, 2022

Abstract

Introduction: Congenital heart disease is an important cause of mortality, chronic morbidity, and disability in children in poor countries. The objective of this study was to highlight the diversity of congenital heart defects in the neonatology department. **Methodology:** This work had taken place in a neonatology service over a period of six months from January 2019 to June 2019. It was a prospective descriptive study with information collected from the medical records of newborns with congenital heart disease. **Results:** Of 1478 neonates hospitalized during the study period, 41 had congenital heart disease, for a hospitalization rate of 2.77%. CHD accounted for 2.77% of neonatal hospitalizations. The sex ratio was 1.73 (26 boys/15 girls). Consanguinity was found in 19% of cases. The average time to consultation was 11 days. The main clinical signs were heart murmur (58%), respiratory distress (56%) and extracardiac malformations (54%). The main cardiac malformations found were atrial septal defect (46%), patent ductus arteriosus (44%), ventricular septal defect (17%), complete atrioventricular canal (15%), and transposition of the great vessels (5%). The case fatality rate was 29%. **Conclusion:** Delayed diagnosis, generally related to the long delay in consultation, and lack of surgical management partly explain this high case fatality.

Keywords

Congenital Heart Disease, Neonates, Management

1. Introduction

Congenital heart disease (CHD) is the most common fetal malformation [1] [2]. They affect approximately 6 to 8 per 1000 live births and are the most common cause of infant death from congenital malformations [3] [4]. The prenatal incidence of CHD is higher (2.4% - 52%) than its postnatal incidence (0.3% - 1.2%) [1] [3]. This huge variability is due to the different screening policies adopted in different countries [1] [5]. The accuracy of these incidence rates is debated and it is concluded that they are probably somewhat low. CHD is much more frequent in regions where fetal echocardiography is performed as a rule during pregnancy [5]. In Tunisia, the incidence of neonatal heart defects varies between 1 and 1.9‰ [6]. In sub-Saharan Africa (excluding South Africa), data on CHD are scarce; their prevalence is estimated to be approximately 8 per thousand live births [7]. In these developing countries, cardiac malformations pose essentially two challenges: diagnosis and treatment [5]. The incidence is probably underestimated and a large number of cases escape diagnosis and management [6].

The exact etiology of congenital heart defects remains largely unknown; ap-proximately 80% - 90% of these cases are thought to have a genetic and envi-ronmental interaction [8] [9]. Environmental factors may be viral (congenital rubella, cytomegalovirus and coxsackie) or toxic (anti-epileptics, trimethadione, isotretinoin, lithium, alcohol, etc.). The diagnosis can only be made in 50 to 75% of cases by clinical examination, as the first symptoms are usually delayed [6] [10].

In Mali, data on neonatal heart disease are scarce. A better knowledge of congenital heart disease in the newborn in the pediatric hospital setting should make it possible to propose appropriate management. The objective of this study was to highlight the diversity of congenital cardiac malformations of the newborn in the neonatology department of the CHU Gabriel Touré in order to propose appropriate interventions to minimize the morbidity and mortality associated with this anomaly.

2. Methodology

This work took place in the neonatology service of the pediatrics department of the Gabriel Touré University Hospital. This hospital is a 3ème level reference structure located in the center of the district of Bamako. The Gabriel Touré University Hospital has an administration, 7 departments grouping together 26 medical-technical services. The pediatrics department is composed of three services:

- The neonatology department;
- The pediatric emergency department (22 hospital beds);
- The general pediatrics department: (58 beds).

The Neonatology Department has several cubicles and units.

- Box 1: reserved for newborns at stable term (23 cribs).
- Box 2: reserved for unstable term newborns (23 cribs).

- Box 3: reserved for premature babies and stable hypotrophs (15 cribs).
- Box 4: reserved for unstable premature and hypotrophic babies (16 cribs).
- Box 5: reserved for very premature babies (4 incubators).
- A Kangaroo Unit (1 consultation room and 1 hospitalization room).
- A sorting room.
- A meeting room.

It receives an average of 3492 patients per year, 85% of whom are referred by other health facilities. The service mainly provides curative care to newborns from the city of Bamako and its surroundings, often even from neighboring countries.

The staff is composed of:

- Two (02) full professors;
- Nine (9) Neonatology Physicians;
- Two (02) Senior Health Technicians;
- Sixteen (16) Health Technicians;
- A secretary;
- One (01) Surveillance Officer.

This was a prospective descriptive study.

The study was conducted from January 1, 2019, to June 30, 2019 (6 months).

The subjects included were neonates with congenital heart disease (CHD) diagnosed by cardiac Doppler echocardiography and hospitalized in the neonatal department during the study period. The diagnosis of CHD was suspected on the basis of clinical arguments (respiratory or feeding difficulty, murmur, dysmorphia, and malformative syndromes) and confirmed by trans-thoracic Doppler echocardiography performed by a cardiologist.

Neonates followed at another center and those whose records did not contain information were excluded from the study.

The data was collected on a pre-established form containing following information:

- Patient's identity: age, gender;
- Maternal history (family);
- Antenatal history (course of pregnancy);
- Prenatal history (delivery: place, mode, complications of pregnancy, condition of birth, birth status, weight, height, CP);
- Neonatal history;
- Feeding habits from birth to date of examination;
- Socioeconomic living conditions;
- Indication of the examination (reason for consultation or hospitalization);
- Data from the clinical examination;
- Date examination performed (age of discovery of heart disease);
- Chest X-ray result;
- Cardiac Doppler ultrasound;
- Biological examinations;

- Management;
- Immediate outcome of newborns.

The data were processed and analyzed on Microsoft Word 2010 and SPSS version 21 software.

Parental or guardian consent was required prior to patient inclusion. Confidentiality and anonymity were respected. The study was approved by the local health and academic authorities.

3. Results

Of 1478 neonates hospitalized during the study period, 41 had congenital heart disease, for a hospitalization rate of 2.77%. At admission, 51% of the newborns were less than 1 week old. The mean age of the newborns was 15 days, with extremes of 1 and 28 days. In 61% of cases, the diagnosis was confirmed during the first 15 days of life. Table 1 shows the distribution of newborns with congenital heart disease according to sex, ages of the patient and mothers, place of birth, and time to consultation. The average age of the mothers was 25 years, with extremes of 18 and 42 years. They were multiparous in 31% of cases and were over 30 years of age in one third of cases. They were unemployed or housewives in 66% of cases and city dwellers in 76% of cases. They had given birth in a referral health center (CSREF) in 49% of cases and at home in 7% of cases. Prematurity was observed in 17% of the newborns and 44% of the patients had low birth weight. Consanguinity was found in 19% of cases.

CHU (university hospital center), CSCOM (community health center), CSREF (reference health center)

The main clinical signs observed were heart murmur (58%), respiratory distress (56%), feeding difficulty (41%) and facial dysmorphia (24%). These signs are listed in Table 2. The main diagnosis at admission was malformative syndrome (34%). Down syndrome was associated with congenital heart disease in 12% of cases. Cardiomegaly was observed in 60% of cases.

Anemia was observed in 12% of patients and C-reactive protein was positive in 20% of cases.

Heart disease was of the left-to-right shunt type in 90% of cases. An atrial septal defect was found in 32% of patients [Table 3 & Table 4]. Cyanogenic heart disease was observed in 10% of cases.

The treatment received was oxygen therapy (71%) in case of desaturation and antibiotic therapy in 98% of cases. Diuretics were administered in 15% of patients and ACE inhibitors in 7%. No neonates were operated on. The case fatality rate was 29% after a 1-month follow-up. Death occurred in 75% of cases before 1 month of life. Death occurred in 83% of the cases with cardiorespiratory failure. The average length of hospitalization was 6 days with extremes of 2 and 60 days.

4. Comments and Discussions

The aim of this prospective study was to contribute to the improvement of early

Table 1. Demographic profile of patients with congenital heart disease.

Variables		Workforce	%
Gender	Male	26	63
	Female	15	37
Place of birth	CHU	3	7
	CSREF	20	49
	Medical practice	9	22
	CSCOM	6	15
	At home	3	7
Age at admission	1ère week	21	51
	2ème week	6	15
	3ème week	6	15
	4ème week	8	19
Age at diagnosis	1 to 15 days	25	61
	16 to 21 days	9	22
	22 to 28 days	7	17
Consultation period	1 to 7 days	25	61
	8 to 14 days	5	12
	15 to 21 days	8	20
	22 to 28 days	3	7
Age of mothers	18 and 19 years old	3	7
	20 to 24 years old	11	27
	25 to 29 years old	15	36
	30 to 34 years old	8	20
	Over 35 years old	4	10

Table 2. Clinical aspects of congenital heart disease cases.

Clinical diagnosis		Workforce	%
Clinical signs	Normal weight	22	54
	Low birth weight (premature and hypotrophic)	18	44
	Macrosomia	1	2
	Heart murmur	24	58
	Respiratory distress	23	56
	Difficulty in eating	17	41
	Low saturation	11	27
	Facial dysmorphia	10	24

Continued

	Cyanosis	1	2
Associated malformations	Down syndrome	5	12
	Choanal atresia	2	5
	Wrist and foot agenesis	2	5
	Duodenal atresia	2	5
	Polydactyly with syndactyly	2	5
	Cleft lip to palate	1	2
	Hydrocephalus	1	2
	Bladder exstrophy	1	2
	Craniosis and microcrania	1	2
	Plum belly syndrome	1	2
	Anorectal malformation	1	2
Initial diagnosis	Malformative syndrome	14	34
	Neonatal infection	7	16
	Prematurity	6	15
	Perinatal anoxia	4	10
	Respiratory distress	3	7
	Neonatal occlusion	2	5
	Congenital heart disease	2	5
	Bronchiolitis	1	2
	Severe acute dehydration	1	2
	Neonatal jaundice	1	2

Table 3. Prevalence of congenital heart disease.

Types of congenital heart disease		Numbers (n = 41)	%
Ultrasound diagnosis	Inter-auricular communication	13	32
	Persistence of the ductus arteriosus	10	24
	Ventricular septal defect + Persistence of the ductus arteriosus	4	10
	Complete atrio-ventricular canal	6	15
	Atrial septal defect + Persistence of the ductus arteriosus	3	7
	Transposition of the large vessels + ventricular septal defect + atrial septal defect	2	5
	Tricuspid atresia + Atrial septal defect and persistence of the ductus arteriosus	1	2
	Ventricular septal defect	1	2
	Common artery	1	2

Table 4. Distribution of patients by outcome.

Variables		Workforce	%
Become	Exeat (Normal output)	27	66
	Deaths	12	29
	Exit against medical advice	2	5
Age at death (n = 12)	2 days	1	8
	9 days	1	8
	13 days	3	25
	25 days	1	8
	26 days	3	25
	33 days	1	8
	37 days	1	8
	54 days	1	8
Circumstances of death (n = 12)	Decompensated anemia	2	17
	cardio-respiratory failure	10	83

detection by providing information on newborns with CHD hospitalized in a neonatal unit and to highlight the shortcomings observed in the management of cases.

For the interpretation of our results, the absence of antenatal diagnosis, the short study period and the small sample size should be taken into account. A short observation period could be the reason for an underestimation of the frequency [9] [11]. The incidence of congenital heart disease in newborns remains high. In this study, a higher incidence (2.77%) is reported because it was performed in a tertiary care unit, which is a referral hospital and all neonates admitted to the unit were included in the study. The incidence of CHD varies between countries, racial and ethnic groups [12]. There are several factors explaining this variation, including lack of technical facilities and necessary skills. As a result, many cardiac malformations go undetected [12] [13]. These difficulties in identifying CHD had been described in detail by several authors [7] [13]. In addition, some congenital heart defects, such as aortic bicuspidism, are usually not accounted for because they are most often diagnosed in adulthood [14] [15].

Data from Africa are scarce. It seems very likely that poor access to health resources leads to a low detection rate [16] [17]. This difference in prevalence could also be genetic, environmental, socioeconomic, cultural or ethnic in origin [18] [19] [20]. Certain specific maternal contexts (consanguinity, diabetes, toxicity, epilepsy, advanced age, infections) would increase the risk of congenital malformations in general and congenital heart disease in particular [21].

The majority of newborns included in this study were referred by peripheral health centers with a longer consultation time (on average 11 days). We observed a male predominance. A female preponderance would be observed in

AIC and atrioventricular canal and a male preponderance would be noticed in aortic valve anomalies including bicuspidism, aortic coarctation, tetralogy of Fallot, transposition of the great vessels and hypoplastic left ventricle [15].

The main presenting symptoms of CHD were respiratory distress (56%), low birth weight (44%), feeding difficulty (41%) and facial dysmorphia (24%). The clinical examination of the newborn can orient the diagnosis in 50% to 75% of cases [22]. A heart murmur (58%) and desaturation (27%) were observed. Ac-cording to the literature, the patient is most often short of breath in case of left-right shunt (CIA, IVC, PCA), cyanotic in case of right-left shunt (tetralogy of Fallot, TGV) [20] [22]. It is breathless and cyanotic in case of mixed venous returns (complex heart disease) [22].

Certain genetic abnormalities or extracardiac congenital malformations should alert health care providers and lead to routine cardiac ultrasound [18].

Two-dimensional echocardiography with color and pulse doppler is the gold standard for the detection of cardiac malformations in the neonatal period. Re-cent studies have shown that the measurement of transcutaneous saturation on the first day of life is a novel, reliable, and inexpensive method for the early de-tection of cardiac defects [22].

Heart disease with a generally non-cyanogenic left-to-right shunt was the most common finding in this study. Similar observations had been reported in the literature [6] [18] [22]. AIC was the most common cardiac malformation in this study, whereas ventricular septal defect (VSD) is considered by many authors to be the most frequent [23] [24] [25]. The high rate of persistent ductus arteriosus is thought to be related to a high prevalence of prematurity [26]. CHD are most often associated with other extracardiac malformations, as well as chromosomal abnormalities [18] [23]. Down's syndrome was observed in 5 patients, *i.e.* 12% of the population. According to the literature, CHD is frequently described in patients with Down syndrome and is the main cause of death in this population during the first two years of life [23].

The organization of CHD management should start from the antenatal period, if not as soon as clinical symptoms are discovered. It should be multidisciplinary, involving pediatricians, gynecologists, obstetricians, cardiologists, radiologists, geneticists and intensive care anesthetists. In Mali, the management of these patients is mainly hampered by the poverty of the patients and the technical facilities. Major efforts are therefore needed to improve screening and case management. The high frequency of congenital malformations should encourage us to move towards antenatal diagnosis.

5. Conclusion

Congenital heart disease is a major health concern in the neonatal department. The most common cardiac malformations are atrial septal defect, patent ductus arteriosus and ventricular septal defect. Emphasis should be placed on early prenatal screening and prompt intervention. Pediatric cardiology should be of

primary importance in training programs.

Conflicts of Interest

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

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