



Knowledge, attitudes and nursing practices on hemophilia in the university clinic of pediatrics and genetics

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Abstract

Hemophilia is a disease caused by a deficiency of the coagulation factors VIII and IV. It is a rare bleeding disorder characterized by abnormal blood coagulation. It manifests itself in deep bleeding, notably hemarthrosis and hematoma, as well as external hemorrhage. The aim of this study is to evaluate the management of hemophilia among the nursing staff of the University Clinic of Pediatrics and Medical Genetics of this center. This disease was the subject of a semi-directed survey in the University Clinic of Pediatrics and Medical Genetics of the CNHU-HKM, where 36 nurses on duty were surveyed. The majority of respondents were between 40 and 50 years old. 55.26% were female, with the majority (68.42) having been on duty for 5 years. Half (54.11%) of those surveyed were unaware of the different types of haemophilia. Finally, 21.05% of staff surveyed had little knowledge of the consequences of hemophilia. This study confirms the need to educate nurses about this rare disease. There is the need to formulate suggestions with a view to strengthening nursing skills in this area.

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Introduction

Hemophilia is a bleeding disorder characterized by deficiency of factor VIII (FVIII) (in the case of hemophilia A) or factor IX (FIX) (in the case of hemophilia B) (de Raucourt *et al.*, 2022; Masson, 2017). It can be congenital or acquired. Congenital hemophilia is a sex-linked recessive disease (Arbour *et al.*, 2022; Romaric, 2021). It is a rare but serious disease. It manifests itself as deep bleeding, notably hemarthrosis and hematoma, but also external hemorrhage (Natacha *et al.*, 2022; Ramé and Thérond, 2023). According to the World Federation of Hemophilia, prevalence is estimated at around one case in 10,000 births.

Hemophilia affects 1 in 10,000 people. Depending on the type of hemophiliac, it is estimated that 1 in 5000 boys will have hemophilia A and 1 in 30,000 boys will have hemophilia B (Goudemand, 2009). Annual global surveys conducted estimate the number of hemophiliacs worldwide at 400,000 (Thabti, 2017). Few cases of hemophilia have been studied in Black Africa. The first cases were described in Southern Africa in 1940, followed by Central Africa in 1950 and West Africa in 1967 (Diop *et al.*, 2003). In Morocco, the study involved 16 haemophilia patients collected at the regional haemophilia reference center in the orient paediatrics department from 2011 to 2013. These included 15 type A hemophiliacs including 3 cases of severe hemophilia (20%), 5 moderate hemophiliacs (33%) and 7 minor hemophiliacs (47%) and one case of hemophilia B. The mean age of the hemophiliacs was 6 years and 2months +/- 3.7 years with a peak between 5 and 10 years (Benajiba *et al.*, 2014).

In Benin, a study published in 1997 on 29 haemophiliacs revealed 27 haemophiliacs A and 02 haemophiliacs B. Patients ranged in age from 16 months to 35 years, with an average age of 19. Anti-haemophilic factor assays revealed

76% severe forms, 18.5% moderate forms and 7.4% minor forms (Boco *et al.*, 1997). Since then, Benin has had no new statistical data on hemophilia, and considering WHO statistics, which estimate that one child in 10,000 suffers from hemophilia, 1,200 children in Benin suffer from this anomaly in a population estimated at 12 million. At present, 125 cases are followed up in the Hematology Clinic of the Centre National Hospitalier Universitaire Hubert K. MAGA (CNHU-HKM), and belong to the Association Béninoise de l'Hémophilie (ABH). The diagnosis of hemophilia is based on the assay of factor VIII by coagulant method associated with the detection of a coagulation inhibitor on a mixture test on a TCA and then the titration of this inhibitor whose threshold of positivity is 0.6 Bethesda unit (Le Cam-Duchez, 2015). The occurrence of this pathology also requires the search for an associated pathology in 50% of cases acquired haemophilia occurs in a context free of any underlying pathology, several types can be found associated pathologies: autoimmune diseases, cancers, dermatological diseases, infections. Of course, there is also the association with postpartum (Le Cam-Duchez, 2015). We are convinced, however, that lack of awareness of this condition among the general population, and among healthcare staff in particular, is a cause of early mortality among hemophiliacs, and moreover explains the rarity of their presence in the various health centers. Nurses are at the heart of patient care, and in order to contribute to improving the quality of their actions in the face of this disease in the Clinique Universitaire de Pédiatrie et de Génétique Médicale of the Centre National Hospitalier Universitaire Hubert K. MAGA (CNHU-HKM), it was opportune to direct our study towards their knowledge, attitudes and practices in this care. To this end, the aim of this study is to evaluate the management of hemophilia among the nursing staff of the University Clinic of Pediatrics and Medical Genetics of this center.

Materials and methods

Study design

In this study, the equipment used consisted mainly of a survey form for collecting information, a digital camera, an audio device (self-recording) for recording interviews, and a position marker (GPRS). This is a descriptive cross-sectional study that took place from June 12 to June 23, 2023. Our sample consisted of nurses from the Emergency and Hospitalization Units of the University Clinic of Pediatrics and Medical Genetics of the CNHU-HKM, using a non-probabilistic method.

In our study, we included all nurses working in the emergency and hospitalization units of the University Clinic of Pediatrics and Medical Genetics, who had given their consent to take part in the study and had at least two (2) months' seniority. Nurses who did not provide clinical care were not included. As the study method is non-probabilistic, the sample size is made up of all nursing staff meeting the inclusion criteria and having agreed to take part in the study.

Survey

The data collection technique consisted of a questionnaire survey. We conducted individual semi-structured interviews with the nurses concerned. The survey form was based on the literature review more precisely through the study carried out by Salih in 2015 (Salih, 2015). The variables to be considered in the study are listed below: knowledge, attitudes and practices Nurses in the management of hemophilia, socio-demographic characteristics of respondents: age of respondent, gender, marital status, seniority.

Data analysis

The data recorded on the survey forms were then entered into Microsoft Excel 2016 and subjected to statistical analysis using SPSS.26.0 software.

Results

Socio-demographic characteristics

Of the 36 nurses questioned, 55.26 of the nurses were female (Fig. 1). About the breakdown of study population by age group presented by the Fig. 2, 41.67% were between 40 and 50 years old, with 68.42% having been with the company for 5 years.

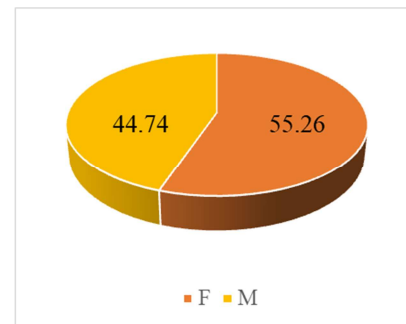


Fig. 1. Population breakdown by gender

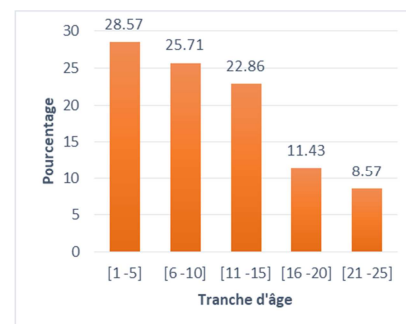


Fig. 2. Breakdown of study population by age group

The results from the breakdown by socio-professional category, we noted that 44.44% having a BEPC (Brevet d'Etude du Premier Cycle) and the others have respectively the bachelor (27,78) and baccalaureate (27,77) (Fig. 3).

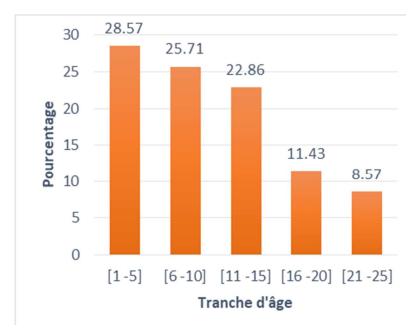


Fig. 3. Breakdown by socio-professional category

According to the distribution of the study population according to place of knowledge of the disease, 35,14% have a training in this (Fig. 4).

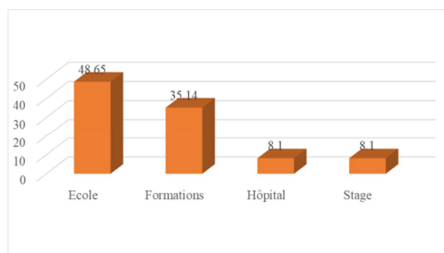


Fig. 4. Distribution of the study population according to place of knowledge of the disease

The nurses questioned all had a good knowledge of what hemophilia is, but only 72.53% knew the symptoms of this disease (Table 1). Although 48.65% had learned about the disease at school, and 35.14% through training, 54.11% had not mastered the different types of hemophilia. All nurses know the cause of the disease, but only 78.95% have a good knowledge of its consequences. The rarity of hemophilia cases means that nurses have no preliminary knowledge of what to do when faced with a case.

Table 1. Distribution of study population by level of disease knowledge

Questions	Good knowledge (%)	Poor knowledge (%)
What causes hemophilia?	100	00
What are the consequences of hemophilia?	78,95	21,05
What are the clinical signs of hemophilia?	72,53	27,47
What are the different types of hemophilia?	45,89	54,11

Discussion

This is a descriptive cross-sectional study of nurses working in the University Clinic of Pediatrics and Medical Genetics at the CNHU-HKM in Cotonou, with the aim of assessing their knowledge, attitudes and practices regarding their role in relation to a child with hemophilia.

The nurses were chosen because of the clinic in which they work. Indeed, this is the referral clinic for children admitted to the center. Hemophilia is an autosomal recessive disease carried by the X gonosome (Purello *et al.*, 1985). The circumstances surrounding the discovery of this disease are difficult, despite the fact that it affects more and more people worldwide, in Africa and particularly in Benin (Baglo *et al.*, 2023). It's a disease that's little-known both to the general public and to many healthcare professionals. As a result, it is most often discovered in the complication stage, where the prognosis is already poor before patients are admitted to the Clinique Universitaire des Maladies du Sang (CUMAS) at the CNHU-HKM, Benin's referral clinic (Morel *et al.*, 2013).

Thirty-six (36) nurses were surveyed during the data collection phase. The majority of respondents were between 40 and 50 years of age (41.67%), with extremes ranging from 30 to 60 years. This indicates a moderately young professional population. This may indicate sufficient experience in the knowledge of common pathologies such as hemophilia. Indeed, if it is known that the age limit for recruitment to the civil service is generally 35, we have a fringe part of the study population with a minimum of 5 years' experience in the practice of care. This can easily be seen from the high proportion of agents with more than 5 years' seniority (68.42%). 55.26% of agents are female. It is well known that medical professions are highly prized by women. We can also explain this fact by the change in behavior in Beninese society, which is increasingly giving women the chance to blossom. Nurses with a Bachelor's degree was the most represented, with 44.44%. This is a positive factor for the quality of care provided to children. The government's policy of training health workers to the minimum level of Baccalaureate at all medical training courses, as at INMeS, also helps to explain this fact.

Generally speaking, all the nurses surveyed had a good knowledge of the disease definition. They also all knew the cause of the disease, stating that it is a deficiency in coagulation factors VIII and IX, linked to a genetic mutation with the possibility of genetic transmission (*Massimo et al.*, 2014). This knowledge, although theoretical, shows that hemophilia is easily diagnosed by these health professionals. The role of education is also highly appreciated in this high level of knowledge among professionals, as only 35.14% have received training in hemophilia, apart from the 48.65% who received instruction during their nursing studies. This also testifies to the good performance of training curricula focusing on the following pathologies in Benin.

Nevertheless, training time for in-depth study of these issues is sometimes insufficient. As a result, more than half (54.11%) of those surveyed are unfamiliar with the different types of hemophilia. It is therefore important that practical training courses fill these gaps. Lastly, 21.05% of staff surveyed had poor knowledge of the consequences of hemophilia. These figures show a good level of knowledge and attitude towards the disease. This could be explained by the high quality of training received. These results concur with those found by Salih in 2015, who worked on pediatric nurses' views on hemophilia in Kirkuk City University Hospital (Salih, 2015).

In terms of practice, all respondents confirmed a rarity of hemophilia cases on the ward. These results differ from those found by Isidore, who counted around 150 cases in the same hospital (Isidore, 2021). This would be due to the modernization of society and health promotion which offer good practices, consultations and premarital tests aimed at reducing genetic diseases such as hemophilia. Awareness-raising in recent years has also had its share of impact on the gradual disappearance of this disease. The rarity of this disease can also make people forget

what to do about it. This is what we found in our study, and this lack of preliminary knowledge of how to deal with hemophilia can be fatal in rare cases. It is therefore important to make suggestions for improving the level of knowledge and practices of caregivers.

Conclusion

Hemophilia is a serious bleeding disorder, which usually reveals itself only by chance. It is an inherited disorder caused by a deficiency in the gene responsible for coagulation factors VIII and IX. The aim of this study was to acquire knowledge, attitudes and practices in the management of hemophilia among nursing staff. As nurses are an irreplaceable link in the care team, mastery of the disease's semiology and practical conduct will be of paramount importance in detecting the disease as quickly as possible, providing emergency care and helping to organize referrals to referral structures. The present qualitative descriptive study was carried out in one of the clinics of the Centre National Hospitalier et Universitaire Hubert Koutoucou Maga in Cotonou. This was the Clinique Universitaire de Pédiatrie et de Génétique Médicale, where 36 nurses on duty were surveyed. According to the results of this survey, the nurses in this department have a good level of theoretical knowledge, but practice is far from being a reality. There is the need to formulate suggestions with a view to strengthening nursing skills in this area.

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