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Research Article

Haemophilia Knowledge among Health Care Providers in a Tertiary Hospital Lagos, Nigeria

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Abstract

Background: Lack of adequate knowledge about Haemophilia and the management of this rare bleeding disorder among health care providers is of a serious public health concern and likely to increase morbidity and mortality.

Objectives: This study was undertaken to assess the level of knowledge about Haemophilia among health care providers in a tertiary health care centre in Lagos, Nigeria.

Methods: This was a cross sectional hospital based survey. A self-administered questionnaire was filled by respondents concerning their knowledge and management of Haemophilia. Completed questionnaires were collated and analyzed with statistical package for social sciences software (SPSS) version 16.

Results: The response rate to this survey was 78.2%. The respondents comprised of 65(41.1%) males and 93(58.9%) females with an age range of 20-59 years and a mean of 38±9.5years. The knowledge of the pathogenesis of Haemophilia among the respondents was good 153(96.8%) while about half 106(66.5%) were aware of the male predominance in this bleeding disorder. Less than half 61(38.6%) of respondents were aware that pain is a symptom in Haemophilia bleed. Fifty eight (36.7%) of the respondents knew that soft tissue and prolonged cut bleeds occur while 110 (69.6%) were aware that joints and muscles are mainly affected by bleeding in Haemophilia. Ninety three (58.9%) of the respondents were aware that HIV infection could complicate the treatment of Haemophilia while 97(61.4%) agreed that hepatitis B and C infections also could.

Conclusion: The knowledge of the clinical features and management of hemophilia needs to be improved on among surveyed health care providers in Lagos, Nigeria.

Keywords: Haemophilia Knowledge Nigeria care Treatment Providers

Introduction

Hemophilia refers to an X-chromosome- linked and autosomal recessive rare group of hereditary bleeding disorders which affects males primarily [1-3]. The underlying molecular defect is a reduced or dysfunctional synthesis of coagulation factor VIII (Hemophilia A) or coagulation factor IX (Hemophilia B), or coagulation factor XI (Hemophilia C) [2-6]. While hemophilia A and B have an X-linked inheritance, hemophilia C is autosomal recessive. In these disorders, there is abnormality in haemostasis which can lead to spontaneous or post-traumatic prolonged bleeding. As a result of this deficiency or dysfunction, the hemophiliac has an impaired ability to stop bleeding when a blood vessel is broken. The hemophiliac bleeds for a much longer time than the normal person. Prolonged bleeding and re-bleeding are the diagnostics symptoms of hemophilia. This bleeding disorder can also be caused by a spontaneous genetic mutation; therefore some people with hemophilia (PWH) do not have a family history [1,4]. The development of inhibitory allo antibodies to coagulation factor VIII (FVIII) can severely complicate the treatment of genetic cases [1,7]. Rarely, the spontaneous development of inhibitory auto antibodies directed against plasma coagulation FVIII results in acquired hemophilia A [1,8] .Acquired hemophilia A is a life threatening disorder and is associated with autoimmune or lymph proliferative diseases, malignancies, dermatological disorders , drugs and interactions, and postpartum period [9-12].The hallmark of hemophilia is hemorrhage into the joints and muscles [2,13].

Hemophila A is the most common form with an incidence of 1: 10,000 male births worldwide [3,14] Hemophilia B has an incidence of 1:20,000-30,000 male births

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worldwide [13]. Hemophilia remains a lifelong non-curative chronic condition and is associated with significant morbidity [15]. Hemopilia A is characterized by prolonged bleeding into various organs of the body. However, the clinical features of the disease vary depending on the level of the coagulation factor present in the blood [5] Based on the severity of the clinical manifestations, the disease has been classified as mild, moderate and severe, although overlap occurs between categories [5]. A previous study has documented a strong association between residual coagulant activity and the clinical bleeding severity in factor VIII deficiency [16].The normal value for coagulation factor VIII (FVIII) assays is 50-150% [1]. In severely affected patients with hemophilia A, there is less than 1% of FVIII normal blood level and activity [5]. Bleeding into joints accounts for approximately 75% of episodes in the hemophiliac [5]. The affected individual frequently experiences spontaneous and prolonged bleeding from cuts or injuries or post-surgery or dental extraction [5]. Other clinical presentations include haemathrosis, haematoma, pseudotumours, haematuria, unusual bleeding after vaccinations, pain, bloody stool, idiopathic epistaxis, unexplained irritability in infants [13]. In moderate and severe hemophilia, prolonged bleeding post circumcision may be the first indication of a bleeding disorder in the neonate [13]. Severe hemophilia is usually diagnosed in childhood. In the absence of effective treatment, there is recurrent haemathrosis resulting in chronic hemophilic arthropathy which occur by young adulthood and are highly characteristic of the severe form of the disorder. Moderately affected patients have greater than 1-5% FVIII normal blood level and activity. In this category, haemathrosis is less disabling than in the severely affected patients. The mildly affected persons with hemophilia A have 6-30% of normal FVIII level and activity with infrequent bleeding episodes [5]. Mild disease may go undiagnosed and discovered only following postoperative prolonged haemorrhage or post trauma [5]. Laboratory studies for a suspected hemophilia A patient include the following; Complete blood cell count, Screening coagulation studies (prothrombin time [PT], activated partial thromboplastin time [aPTT]), FVIII assay and FVIII inhibitor assay (Bethesda assay) [1]. Ideally, the management of hemophilia A should be provided through a comprehensive care center while FVIII concentrate could be given as prophylaxis at home [1].The treatment of hemophilia may involve the following; management of haemostasis, management of bleeding episodes ,use of factor replacement products and adjuvant medications, treatment of patients with factor inhibitors ,treatment and rehabilitation of patients with hemophilia synovitis [1] Early diagnosis of hemophilia ,communication with the patient and their parents or guardians and implementation of the best treatment are essential [16] It has been found and documented that high level of knowledge and practical skills are required for effective management of this rare blood disorder [17]. Lack of information about bleeding disorders has been described as a serious public health concern [18]. Previous studies have shown that complications of hemophilia can be decreased or prevented and quality of life would be improved by early diagnosis and appropriate management [19-21]. A previous research also concluded that health instructions have an impact on improving knowledge and practices of hemophilia A [22].

This study was carried out to assess the level of basic knowledge, clinical presentation, laboratory diagnosis, care and treatment as well as complications and prevention of hemophilia among health care providers in Lagos, Nigeria. Based on obtained results, appropriate recommendations for the introduction of advocacy and awareness programmes on hemophilia would be made. Optimal knowledge about the diagnosis, care and treatment of this genetic disorder would reduce morbidity and mortality among PWH.

Methods

This study was hospital based cross sectional survey among health care providers and was carried out from March to April 2016 in a tertiary care (740 beds) hospital in Lagos, Nigeria. This state owned tertiary health care facility and referral centre is situated at Ikeja, the capital city of Lagos which is a commercial nerve centre, located South west Nigeria. As a tertiary hospital in Lagos, which is the largest urban area in Nigeria, this hospital serves an estimated population of about 22 million residents. A self-administered 29 item structured questionnaire was used to collect data. Also captured was the acquired skill of care and treatment, complications of and prevention of hemophilia. Obtained information was imputed into computer and statistical analysis was done using Statistical Package for Social Sciences (SPSS) software version 16.0. Results are presented in simple tables of frequencies and percentages (Tables 1-4).

The structured questionnaire was in the following parts:

i. Socio demographic characteristics such as age, gender, designation and the Department of the respondents

ii. The respondents' knowledge of the pathogenesis and clinical features of hemophilia.

Iii. The respondents' knowledge of the laboratory diagnosis, care and treatment, complications and prevention of hemophilia.

Results

The response rate was 78.22%. There were 158 respondents comprising 65 (41.1%) males and 93 (58.9%) females with a mean age of 38 ±9.50 years. Most 54 (34.2%) of the respondents were in the age group 30-39years. One hundred and fifty three (96.8%) of the respondents were aware that hemophilia is hereditary while 105 (66.5%) agreed that males are predominantly affected.

Table 1: Demographic Characteristics of Respondents.

Characteristics	Frequency	%
Age group (years)		
20 - 29	33	20.9
30-39	54	34.2
40-49	44	27.8
50-59	27	17.1
Gender		
Male	65	41.1
Female	93	58.9
Occupational Groups		
Physician	73	46.2
Dentist	27	17.1
Nurse	45	28.5
Physiotherapist	13	8.2
Total	158	100

Table 2: Respondents 'Knowledge of the Pathogenesis and Clinical Features of Haemophilia.

Knowledge Variables	Response		
	Yes		
n (%)	No		
n(%)	Unknown		
n(%)			
Haemophilia is an inherited rare blood disorder	153(96.8)	1(0.6)	4(2.5)
Haemophilia is an infectious disease	3 (1.9)	154 (97.5)	1 (0.6)
Prolonged bleeding in haemophilia is due to deficient or dysfunctional coagulation factor			
Haemophilia predominantly affects the male	105(66.5)	31(19.6)	22(13,9%)
Predominantly affects the females	26 (16.5)	104(65.8)	28(17.7)
Haemophilia is curable	37 (23.4)	96(60.7)	25 (15.8)
Prolonged post circumcision bleeding is common	150(94.9)	3 (1.9)	5 (3.2)
Pain is a symptom	61(38.6)	70 (44.3)	27(17.1)
Soft tissue and prolonged cut bleeds occur	58 (36.7)	85(53.8)	15 (9.5)
Bleeding spontaneously into joints is a feature	128(81.0)	15(9.5)	15(9.5)
Rebleeds are common	130 (82.3)	11(7.0)	17(10.8)
Bleeding does not affect weight bearing joints	17 (10.8)	110 (69.6)	31 (19.6)
Prolonged bleeding after tooth extraction may occur	147(93.0)	5(3.2)	6(3.8)
Knee joint bleed is most frequent	85(53.8)	30(19)	43(27.2)
Brain bleed may occur	109(69)	18(11.4)	31(19.6)
Affected females may have heavy menstrual flow	92 (58.2)	40(25.3)	26 (16.5)

Table3: Respondents' Knowledge of Laboratory Diagnosis, Care, Treatment, Complications and Prevention of Haemophilia.

Knowledge Variables	Response		
	Yes		
n(%)	No		
n(%)	Unknown		
n(%)			
Coagulation factor assay is most important for diagnosis	138(87.3)	3(1.9)	17(10.8)
Deficient coagulation factor replacement is the mainstay in management	129 (81.6)	6(3.8)	23(14.6)
Prophylaxis with factor concentrate is proactive and beneficial	126(79.7)	8(5.1)	24(15.2)
Care and Treatment is lifelong	124(78.5)	14(8.9)	20 (12.6)
Life expectancy is close to normal with appropriate Care and Treatment	103(65.2)	6(3.8)	49 (31.0)
Genetic Counseling is relevant in the prevention of Haemophilia	138 (93.2)	5 (3.4%)	5(3.4)
Female relatives of the Haemophiliac need genetic testing	127(85.8)	14(9.5)	7(4.7)
Prenatal diagnosis is important in prevention	128(88.2)	4(2.8)	13(9.0)
HIV infection may complicate treatment	93(58.9)	47(29.7)	18(11.4)
Hepatitis B and C may complicate treatment	97 (61.4)	27(17)	34(21.5)
A multidisciplinary comprehensive approach of Care and Treatment is critical	150 (94.9)	0(0)	8 (50.1)
Have you ever seen a Haemophiliac?	80 (50.6)	72 (45.6)	6 (3.8)
Have you managed a Haemophiliac	48(30.4)	104 (65.8)	6 (3.8)

One hundred and fifty (94.9%) of the respondents knew that prolonged post circumcision bleeding is a common presentation of hemophilia. Sixty one (38.6%) of respondents was aware that pain is a symptom in hemophiliac bleed. Fifty eight (36.7%) of the respondents knew that soft tissue and prolonged cut bleeds occur while 110 (69.6%) were aware that weight bearing joints are mainly affected by bleeding in hemophilia. One hundred and forty seven (93.0%) of the respondents were aware that prolonged bleeding may occur post tooth extraction in hemophilia. One hundred and thirty eight (87.3%) of the respondents knew that the diagnosis of hemophilia is mainly by deficient coagulation factor assay while 129(81.6%) agreed that replacement therapy is the mainstay of treatment. One hundred and twenty six (79.7%) of the respondents agreed that prophylaxis with factor concentrate in hemophilia management is proactive and beneficial. Ninety three (58.9%) of the respondents were aware that HIV infection could complicate the treatment of hemophilia while 97(61.4%) agreed that hepatitis B and C infections also could .One hundred and fifty (94.9%) of the respondents agreed that a comprehensive multidisciplinary approach to the management of hemophilia is the ideal. About half 80(50.6%) of the respondents had ever seen while 48(30.4%) had managed a hemophiliac. One hundred and thirty eight 138(93.2%) and128 (88.2%) of the respondents knew that genetic counseling and prenatal diagnosis were respectively relevant in the prevention of hemophilia. Thirty seven (23.4%) of the respondents had the impression that hemophilia is curable. One hundred and three (65.2%) of the respondents were aware that IIfe expectancy is close to normal with appropriate care and treatment for the hemophiliac.

Table 4: Self-Administered Questionnaire.

Hemophilia knowledge among health workers			
Date……			
Serial No……..			
Biodata			
Age (years) =			
Gender M F			
Designation……			
Telephone Number……			
Department……			
*** Please circle the applicable			
Knowledge of Hemophilia			
Hemophilia is			
(i) Inherited bleeding disorder usually	Yes	No	Unknown
(ii) may arise from mutation	Yes	No	Unknown
(iii) There is dysfunctional or deficient clotting factor	Yes	No	Unknown
(iv) a common disorder	Yes	No	Unknown
(v) mostly affects males	Yes	No	Unknown
(vi) affects females mostly	Yes	No	Unknown
(vii) curable disease	Yes	No	Unknown
Signs and symptoms of Hemophilia include:			
Acute or chronic pain	Yes	No	Unknown
Prolonged post circumcision bleeding	Yes	No	Unknown
Prolonged internal and external bleeding	Yes	No	Unknown
Bleeding into joints spaces spontaneously	Yes	No	Unknown
Re-bleeds are a common feature	Yes	No	Unknown
Bleeding does not affect weight bearing joints	Yes	No	Unknown
Prolonged bleeding after tooth extraction	Yes	No	Unknown
Knee joint is mostly frequently affected	Yes	No	Unknown
Bleeding into the brain may occur	Yes	No	Unknown
Bleeding may be spontaneous	Yes	No	Unknown
Laboratory Diagnosis	Yes	No	Unknown
Plasma clotting factor level assay is most important	Yes	No	Unknown
Prenatal diagnosis is possible	Yes	No	Unknown
Care and Treatment	Yes	No	Unknown
Primary deficient factor replacement is the mainstay	Yes	No	Unknown
Prophylaxis with factor concentrate is proactive and beneficial	Yes	No	Unknown
On demand factor replacement is as effective as prophylaxis	Yes	No	Unknown
Treatment is lifelong	Yes	No	Unknown
Life expectancy has improved to close to normal	Yes	No	Unknown
with appropriate care and treatment	Yes	No	Unknown
A specialist team for comprehensive care of hemophilia is ideal practice	Yes	No	Unknown
Complications of treatment include:			
HIV infections	Yes	No	Unknown
Hepatitis B and C Infection	Yes	No	Unknown
Prevention of Hemophilia	Yes	No	Unknown
Genetic counseling is relevant	Yes	No	Unknown
Prenatal diagnosis is important	Yes	No	Unknown
Have you ever seen hemophilia?	Yes	No	Unknown
Have you ever managed a case of hemophilia?	Yes	No	Unknown

Discussions

Our survey assessed the basic knowledge of the pathogenesis, clinical features, laboratory diagnosis as well as care and treatment of hemophilia among health care providers in Lagos, Nigeria. We observed that 153 (96.8%) of the respondents were aware that hemophilia is hereditary while 105 (66.5%) agreed that the males are predominantly affected. A previous study reported more than 80% of the respondents as having high medium knowledge levels about the inheritance pattern of hemophilia A [23].The pattern of inheritance of hemophilia is reportedly X-linked recessive, thus, with rare exceptions, males are affected while females are carriers of the trait [2]. Majority of respondents 150 (94.9%) agreed that prolonged post circumcision bleeding is a common clinical presentation of hemophilia in the neonatal age. It is documented that prolonged post circumcision bleeding in the neonate may be the first sign of severe hemophilia A in a baby boy [13]. We also observed in our survey, that 58(36.7%) respondents agreed that prolonged soft tissue and cut bleeds occur while 110 (69.6%) were aware that weight bearing joints are mainly affected by spontaneous bleeding in this disorder. We found that 61(38.6%) of the respondents were aware that pain is a symptom in hemophiliac bleed. The bleeding in hemophilia has been documented to be extremely painful in the acute stage and leads to long-term inflammation and deterioration of the involved joint or muscle [13,24]. A previous research concluded that continued research in the area of hemophilia pain is needed [25]. Hemophilia A and B have been reported as clinically indistinguishable and are characterized by prolonged and repeated bleeding episodes particularly into muscles and joints [2,13]. Also, we found out that 147(93.0%) of the respondents were aware that prolonged bleeding may occur post tooth extraction. It has been reported that dental extraction is the most frequent surgical procedure performed and the resultant bleeding could be prolonged in Hemophilia A [1,5]. We found in our survey that 138 (87.3%) of the respondents knew that the diagnosis of hemophilia is mainly by coagulation factor assay. It has been documented that the diagnosis of hemophilia is established by specific quantitative coagulation factor assay [2,5,7,26,27]. In the less developed countries, the greatest challenges of hemophilia care have been identified to include recognition and diagnosis with a lot of under diagnosed cases [28,29]. Previous studies have shown that complications of hemophilia can be decreased or prevented with improved quality of life by early diagnosis and appropriate treatment which is essential [14,18- 21]. We also found in this survey that most of the respondents 129(81.6%) agreed that replacement of deficient coagulation factor is the mainstay of management of the hemophiliac. This finding is similar to the result of a previous study on Treatment and related knowledge where most interviewees had good basic knowledge of treatment [30]. One hundred and twenty six (79.7%) agreed that prophylaxis with coagulation factor concentrate in hemophilia management is proactive and beneficial. Specific coagulation factor replacement which could be on demand after every bleeding episode or as a prophylaxis regularly two or three times a week is reportedly the choice of therapy in hemophilia A [31]. Coagulation factor replacement therapy in hemophilia A plays an important role in the effective management of this rare bleeding disorder. It has been reported that in the developing nations, there is little or no access to coagulation factor concentrates as a result of which there is greater morbidity and mortality among the hemophiliacs [32]. However, recombinant products are increasingly regarded as the preferred choice due to a low risk of viral infection [31]. Reportedly, developments in gene therapy may provide an attractive model for the treatment of hemophilia by eliminating

the need for regular injections of coagulation factor VIII [14]. We observed that 93 (58.9%) of respondents were aware that HIV infection could complicate the treatment of hemophilia while 97(61.4%) agreed that hepatitis B and C infections also could .In the choice of coagulation factor replacement therapy for hemophilia care, the prevention of blood borne infectious agent transmission is primary. It has been reported that virus inactivated concentrates or detergent treated plasma derivates are safe where available while untreated concentrates and plasma products should be avoided [7]. Recombinant technology and viral inactivation methods have virtually eliminated the risk of blood borne infections [7]. The USA hemophilia foundation has instructed her PWH to refuse the use of all non-heat treated blood products, while the UK hemophilia Society has also threatened to do same [25].

We found out in our survey, that 80(50.6%) of respondents had ever seen while 48(30.4%) had managed a hemophiliac. This finding may be due to inadequate knowledge about this bleeding disorder and therefore under diagnosis. Previous studies reported that a majority of hemophiliac patients live in the developing countries like ours where the blood disorder is under diagnosed and undertreated [28,30]. One hundred and fifty (94.9%) of the respondents in our survey agreed that a comprehensive multidisciplinary approach to treatment is the ideal. Previous studies have documented that optimal care of patients with hemophilia requires a comprehensive approach delivered by a multidisciplinary team of specialist's [27,33]. Comprehensive programmes have been documented to make a major improvement in the treatment of hemophilia and a positive impact on the quality of life. One hundred and thirty eight (93.2%) and 128(88.2%) of the respondents were aware that genetic counseling and prenatal diagnosis were relevant in the prevention of hemophilia respectively. Previous studies concluded that genetic counseling services could be integrated with the antenatal care service as it will provide information that would enable a woman make an informed decision in relation to family planning [3,34]. We also found out that 103(65.2%) of the respondents were aware that life expectancy may be close to normal with appropriate care and treatment for the hemophilia. In Ireland, previous research documented that the life expectancy of a child born with hemophilia is essentially normal with a reportedly excellent quality of life [3].

In conclusion, the knowledge of the clinical features and management of hemophilia needs to be improved on among surveyed health care providers. We recommend that frequent medical education about this rare bleeding disorder be arranged to improve the knowledge and management skills of healthcare providers in our setting as this would reduce mortality and morbidity related to delayed diagnosis and treatment. Also, in developing nations such as Nigeria, there is a need to set up hemophilia treatment centres with specialty care providers who would be trained to administer factor products properly and safely.

Addendum

T M Balogun: Conceptualization and design of the study, literature review, data analysis and draft report writing

B M Agboola: implementation and data collection

O O Onigbinde: implementation, data collection and critical review

A Ajayi: Data analysis

I Iredu: Data analysis

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