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Assessment of Knowledge of Health Workers on Haemophilia at the University of Medical Sciences Teaching Hospital, Ondo State, Nigeria

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Abstract: Haemophilia is a rare hereditary bleeding disorder that occurs primarily among males. Adequate knowledge of its clinical presentation and management of this disorder by health workers will improve the clinical course of patients having this disorder. The study was done to assess the knowledge of clinical presentation and management of Haemophillia among health workers at the University of Medical Sciences Teaching Hospital Complex, (UNIMEDTHC) Ondo State, Nigeria. A cross sectional hospital based survey was done using questionnaires that were self-administered by the recruited respondents. Completed questionnaires were collated and analyzed with statistical package for Social Sciences Software (SPSS) version 25. A total of 194 health workers participated in the study with a response rate of 89.4%. The respondents comprised of 72 (37.1%) males and 122 (62.9%) females. Knowledge of the pathogenesis of Haemophilia among the respondents was good 178 (91.8%). Male predominance of the disorder was ascertained by 148 (76.3%) respondents. About half, 105 (54.1%) of respondents were aware that pain is a symptom among haemophiliacs. One hundred and seventy seven (91.2%) of the respondents knew that prolonged post circumcision bleeding is a symptom while 160 (82.5%) were aware that joints and muscles are mainly affected by bleeding in Haemophilia. One hundred and eighteen (60.8%) and 145 (74-4%) of the respondents were aware that HIV and Hepatitis B and C respectively were possible complications of treatment. The role of genetic counseling in prevention of Haemophillia was ascertained in 88-7% of the respondents. There is a fairly good knowledge of Haemophillia among the surveyed health workers. There is a need for improved knowledge about Haemophilia among health workers through continuing education to ensure prompt diagnosis and appropriate care of people with this disorder.

Keywords: Assessment, Knowledge, Health Workers, Haemophilia

1. Introduction

Haemophilia is a hereditary bleeding disorder that occurs primarily among males. It is characterized by bleeding into muscles and joints and causes chronic pain and joint disease [1]. Haemophilia is the most common hereditary coagulation factor deficiency, which is characterized by an impaired ability to stop bleeding when a blood vessel is broken [2]. According to Valizad et al, the underlying molecular defect is a reduced or dysfunctional synthesis of coagulation factor VIII (Haemophilia A) or coagulation factor IX (Haemophilia B), or coagulation factor XI (Haemophilia C) [3]. While haemophilia 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 and it impacts approximately 400,000 people worldwide [4]. Haemophila A is the most common form with an incidence of 1:10,000 male births worldwide [2, 5], and Haemophilia B has an incidence of 1:20,000-30,000 male births worldwide [6]. Haemophilia remains a lifelong non-curative chronic condition and is associated with significant morbidity [7].

People with bleeding disorders can live healthy and effective lives, however, if left untreated bleeding disorders, specifically haemophilia, can lead to disabling pain, acute joint injury and lethal internal bleeding [8]. Although people at all haemophilia severity levels can bleed as a result of an injury, those with severe forms of the disease often experience spontaneous bleeding [1]. In rare cases, bleeding into vital organs can cause death. Bleeding contributes to the progressive deterioration of joints and muscles and this affects patients' well-being and their everyday activities [9]. Other clinical presentations include haemathrosis, haematoma, pseudotumours, haematuria, unusual bleeding after vaccinations, pain, bloody stool, idiopathic epistaxis, and unexplained irritability in infants [6]. Complications associated with haemophilia includes, hematomas of the soft tissues, intracranial hemorrhages, prolonged bleeding, poor wound healing, prolonged oozing following minor injuries, epistaxis, and ecchymosis [2].

Fahimeh et al in an earlier study opined that early treatment of severe hemorrhage episodes is necessary to reduce long-term complications [10]. Severe bleeding episodes commonly affect the knee, elbow and ankle joints. Early treatment with replacement of essential clotting factor at the time of a bleeding episode can prevent severe joint disease [11]. Prophylaxis can be used to stabilize or improve function in a joint that has bled in the past (secondary prophylaxis) or to prevent bleeding (primary prophylaxis). According to the Universal Data Collection Program of the Centers for Disease Control and Prevention (CDC) [12], 44% of people with severe hemophilia A (clotting factor VIII deficiency, the more prevalent form of hemophilia) and 33% of people with haemophilia B (clotting factor IX deficiency) are treated with some form of prophylaxis. The first randomized clinical trial of young children with haemophilia A on prophylaxis showed improved joint function [13]. Until the past decade, treatment of haemophilia involved intravenous infusion of a human blood-derived clotting factor that was effective in stopping most bleeding episodes, but transmitted blood-borne infections, including HIV and hepatitis A, B, and C. According to the CDC's Haemophilia Surveillance System, 7.5% of youths with haemophilia were infected with HIV, and 53% contracted hepatitis C from tainted blood products (CDC, unpublished data, 1998) cited by Nazzaro *et al* [1]. Newer manufacturing processes such as recombinant technology, implemented in the early 1990s, have since increased the safety of blood products [1]. Adequate knowledge of clinical presentation, early diagnosis and prompt management of haemophillia by health workers can help to prevent or minimize complications when it occurs. This study was conducted to assess the knowledge of health workers at the UNIMEDTHC, Nigeria about the clinical presentation, diagnosis and management of haemophillia

2. Methodology

The cross-sectional survey was conducted at the University of Medical Sciences Teaching Hospital Complex, Ondo State, Nigeria in November 2019. The respondents were health workers of the University of Medical Sciences Teaching Hospitals Complex, Ondo State, The teaching hospital has annexes in Akure and Ondo the two largest towns in the state. The study was conducted in the two centers. The research employed a well-structured questionnaire in obtaining data from the respondents. The structured questionnaire was in the following parts: Socio demographic information of the respondents; General knowledge of Haemophilia; Signs and symptoms of Haemophilia; and Treatment, Complications and prevention of Haemophilia. The study sample size was determined using the Krejcie and Morgan table of (1970) at 95% level of confidence. Hence the sample size of the study was two hundred and seventeen (217).

The respondents were selected using simple random sampling technique, targeting mainly doctors and nurses. Questionnaires were self-administered by the respondents. Ethical clearance for the study was obtained from the Research and Ethics Committee of the hospital. Data was analyzed using Statistical Package for Social Science (SPSS) version 25. Results are represented as descriptive statistics in tables using frequencies and percentages.

3. Results

Table 1 on demographics shows there were 194 respondents comprising 72 (37.1%) males and 122 (62.9%) females. About half of the respondents (49%) were above 30 years old. Doctors and nurses accounted for 81.4% of the respondents while the remaining health workers accounted for 18.4%.

Table 2 showed 178 (91.8%) of the respondents were aware that hemophilia is hereditary while 148 (76.3%) agreed that males are predominantly affected. One hundred and eighteen (60.8%) of the respondent knew that affected females may have heavy menstrual flow. One hundred and twenty (61.9%) knew that the chance of a couple having a boy with Haemophilia if the mother is a carrier is 1 in 4 births. Forty nine (25.3%) of the respondents had the impression that haemophilia is curable. About 40.7% and 21.6% of the respondents had seen and managed a haemophiliac patient respectively. One hundred and seventy nine (92.3%) of the respondent knew that prolonged bleeding in haemophilia is due to deficient or dysfunctional coagulation factor.

Table 3 showed one hundred and seventeen (91.2%) of the respondents knew prolonged post circumcision bleeding as a common clinical presentation of haemophilia. One hundred and five (54.1%) of the respondents knew haemophiliacs could present with pain. One hundred and seventy-seven (91.2%) and 130 (67%) of the respondent were aware that internal and external bleeding and bleeding into weight bearing joints respectively could occur in haemophiliacs. One hundred and seventy six (90.7%) of the respondents were aware that prolonged bleeding may occur following tooth extraction.

Table 4 showed one hundred and eighty three (94.3%) of the respondents knew that diagnosis of haemophilia is mainly by deficient coagulation factor assay while 161 (83%) thought that replacement therapy is the mainstay of treatment. One hundred and sixty one (83%) of the respondents agreed that prophylaxis with factor concentrate in haemophilia management is beneficial. One hundred and eighteen (60.8%) of the respondents knew HIV infection as a complication of treatment of haemophilia while one hundred and forty five (74.7%) thought hepatitis B and C infections could also complicate treatment. One hundred and sixty eight (86.6%) of the respondents agreed that a comprehensive multidisciplinary approach to the management of haemophilia is the ideal. One hundred and seventy two (88.7%) and one hundred and fifty four (79.4%) of the respondents knew that genetic counseling and prenatal diagnosis were respectively relevant in the prevention of haemophilia. One hundred and sixty six (85.6%) of the respondents were aware that life expectancy is close to normal with appropriate care and treatment for haemophillia..

Table 1. *Demographic Characteristics of Respondents.*

		Frequency	Percent (%)
Gender	Male	72	37.1
	Female	122	62.9
Age	Below 20 years	13	6.7
	20 – 29 years	86	44.3
	30 – 39 years	62	32.0
	40 – 49 years	27	13.9
	50 years	6	3.1
Designation of Health Workers	Doctors	92	47.4
	Nurse	66	34
	Laboratory workers	25	12.9
	Health attendants	2	1.0
	Midwives	9	4.6
Nature of Employment	Full	134	69.1
	Contract	60	30.9
Insurance Coverage	Yes	49	25.3
	No	145	74.7
Duration of Employment	Less than 1 year	74	38.3
	1 – 5 years	74	38.3
	6 – 10 years	18	9.3
	above 10 years	27	14.0

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

Questions	Measures	Frequency	Percent (%)
Haemophilia is an inherited rare blood disorder?	Yes	178	91.8
	No	13	6.7
	I Don't Know	3	1.5
Haemophilia is a common blood disorder?	Yes	78	40.2
	No	111	57.2
	I Don't Know	5	2.6
Haemophilia is an infectious disease?	Yes	19	9.8
	No	169	87.1
	I Don't Know	6	3.1
Prolonged bleeding in Haemophilia is due to deficient or dysfunctional coagulation factor?	Yes	179	92.3
	No	11	5.7
	I Don't Know	4	2.1
Haemophilia predominantly affects the male?	Yes	148	76.3
	No	27	13.9

Questions	Measures	Frequency	Percent (%)
	I Don't Know	19	9.8
Haemophilia affects females?	Yes	110	56.7
	No	64	33
	I Don't Know	20	10.3
Haemophilia is curable?	Yes	49	25.3
	No	125	64.4
	I Don't Know	20	10.3
Affected females may have heavy menstrual flow?	Yes	118	60.8
	No	36	18.6
	I Don't Know	40	20.6
The chance of a couple having a boy with Haemophilia, if the mother is a carrier is: (1 in 4 births)?	Yes	120	61.9
	No	33	17.0
	I Don't Know	41	21.1
Have you ever seen a Haemophiliac before?	Yes	79	40.7
	No	112	57.7
	I Don't Know	3	1.5
Have you managed a Haemophiliac before?	Yes	42	21.6
	No	149	76.8
	I Don't Know	3	1.5

Table 3. *Respondents' Knowledge of Laboratory Diagnosis, Care, Treatment, Complications and Prevention of Haemophilia.*

Questions	Measures	Frequency	Percent (%)
Acute or Chronic Pain	Yes	105	54.1
	No	61	31.4
	I Don't Know	28	14.4
Prolonged Post Circumcision bleeding	Yes	177	91.2
	No	6	3.1
	I Don't Know	11	5.7
Prolonged Internal and external bleeding	Yes	177	91.2
	No	3	1.5
	I Don't Know	14	7.2
Bleeding into joints spaces spontaneously	Yes	160	82.5
	No	15	7.7
	I Don't Know	19	9.8
Re – bleeds are a common feature	Yes	158	81.4
	No	11	5.7
	I Don't Know	25	12.9
Bleeding does not affect weight bearing joints	Yes	26	13.4
	No	130	67.0
	I Don't Know	38	19.6
Prolonged bleeding after tooth extraction	Yes	176	90.7
	No	10	5.2
	I Don't Know	8	4.1
Knee Joint is mostly frequently affected	Yes	107	55.2
	No	36	18.6
	I Don't Know	51	26.3
Bleeding into the brain may occur	Yes	122	62.9
	No	29	14.9
	I Don't Know	43	22.2
Bleeding maybe spontaneous	Yes	179	92.3
	No	5	2.6
	I Don't Know	10	5.2

Table 4. *Respondents' Knowledge ofTreatment, Complications and Prevention (TCP) of Haemophilia.*

Questions	Measures	Frequency	Percent (%)
Coagulation factor assay is most important for diagnosis	Yes	183	94.3
	No	3	1.5
	I Don't Know	8	4.1
Deficient coagulation factor replacement is the mainstay in management	Yes	161	83.0
	No	10	5.2
	I Don't Know	23	11.9
Prophylaxis with factor concentrate is proactive and beneficial	Yes	161	83.0
	No	4	2.1
	I Don't Know	29	14.9
Care and treatment is lifelong	Yes	164	84.5

Questions	Measures	Frequency	Percent (%)
	No	8	4.1
	I Don't Know	22	11.3
Life expectancy is close to normal with appropriate care and treatment.	Yes	166	85.6
	No	8	4.1
	I Don't Know	20	10.3
Genetic counselling is relevant in the prevention of Haemophilia	Yes	172	88.7
	No	7	3.6
	I Don't Know	15	7.7
Female relatives of the Haemophiliac needs genetic testing.	Yes	163	84.0
	No	14	7.2
	I Don't Know	17	8.8
Pre – natal diagnosis is important in prevention	Yes	154	79.4
	No	14	7.2
	I Don't Know	26	13.4
HIV infection may complicate treatment	Yes	118	60.8
	No	39	20.1
	I Don't Know	37	19.1
Hepatitis B and C may complicate treatment	Yes	145	74.7
	No	18	9.3
	I Don't Know	31	16
A multidisciplinary comprehensive approach of care and treatment is critical	Yes	168	86.6
	No	7	3.6
	I Don't Know	19	9.8
Plasma clothing factor level assay is most important	Yes	157	80.9
	No	13	6.7
	I Don't Know	24	12.4
On demand factor replacement is as effective as prophylaxis	Yes	122	62.9
	No	26	13.4
	I Don't Know	46	23.7
A specialist team for comprehensive care of Haemophilia is an ideal practice	Yes	169	87.1
	No	8	4.1
	I Don't Know	17	8.8

4. Discussion

This survey assessed the knowledge of the pathogenesis, clinical presentation, diagnosis as well as treatment, complication and prevention of haemophilia among health workers in the University Teaching Hospital Complex, Ondo State, Nigeria. We observed that 178 (91.8%) of the respondents were aware that haemophilia is hereditary while 148 (76.3%) agreed that males were predominantly affected by the disorder. An earlier study reported more than 80% of the respondents as having high medium knowledge levels about the inheritance pattern of haemophilia A [14]. The inheritance of haemophilia is X-linked recessive, with rare exceptions; males are affected while females are carriers of the trait. Majority of respondents 177 (91.2%) agreed that prolonged post circumcision bleeding is a common clinical presentation of haemophilia in the newborn period. It is documented that prolonged post circumcision bleeding in the neonate may be the first sign of severe haemophilia A in a male infant [6]. We noted that 177 (91.2%) respondents agreed that prolonged internal and external bleeds occur, while 130 (67%) were aware that weight bearing joints are mainly affected by spontaneous bleeding in this disorder. We found that 105 (54.1%) of the respondents were aware that pain is a symptom in haemophiliac bleed. The bleeding in haemophilia 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 [6, 15]. Both Haemophilia A and B are characterized by prolonged and repeated bleeding episodes particularly into muscles and joints [6]. Dental extraction is the most frequent surgical procedure performed in them and the resultant bleeding could be prolonged [16, 17]. In this study 176 (90.7%) of the respondents were aware that prolonged bleeding may occur post tooth extraction. A large number of the respondents in this study knew that the diagnosis of haemophilia is mainly by coagulation factor assay. Diagnosis of haemophilia has been established to be by specific quantitative coagulation factor assay [2]. One of the major challenges of management of haemophilia in most developing countries is with case definition, with resultant missed diagnosis [18, 19]. It has been reported that complications of haemophilia can be decreased or prevented with improved quality of life through early diagnosis and appropriate treatment [5]. Most of the respondents 161 (83.0%) in this study agreed that replacement of deficient coagulation factor is the mainstay of management of haemophilia. This is similar to the result of a previous study where most of the respondents had good knowledge of treatment of haemophilia [20]. One hundred and sixty-one respondents (83.0%) agreed that prophylaxis with coagulation factor concentrate in haemophilia management is both proactive and beneficial. Specific coagulation factor replacement following every bleeding episode, or as prophylaxis given twice or thrice weekly is reported as the therapy of choice in Haemophilia A [21]. In

most developing nations, there is little or no access to coagulation factor concentrates hence a greater morbidity and mortality among the haemophiliacs in these nations [22]. Recombinant products have been increasingly regarded as the preferred choice for management of haemophilia due to its low risk for viral infection [21].

One hundred and eighteen (60.8%) and 145 (74-4%) of the respondents were aware that HIV and Hepatitis B and C respectively were possible complication of treatment. Considerations for prevention of transfusion transmissible infection is important while deciding on therapeutic choices. The use of virus inactivated concentrates or detergent treated plasma derivates are considered safe when available while the use of untreated concentrates and plasma products is discouraged [23].

We observed that only 79 (40.7%) of respondents had seen a haemophilia patient before while 42 (21.6%) had managed one. This might suggest under diagnosis of the disease and not a true reflection of how prevalent the disorder is. Studies have shown that majority of haemophiliacs living in developing countries were being under diagnosed and undertreated [18, 20]. One hundred and sixty-eight (86.6%) of the respondents in this study opined that a comprehensive multidisciplinary approach to treatment is the ideal. Comprehensive multidisciplinary approach has been judged as the optimal health care option for haemophiliacs [24, 25]. One hundred and seventy two (88.7%) and 154 (79.4%) of the respondents were aware that genetic counseling and prenatal diagnosis were relevant in the prevention of haemophilia respectively. Genetic counseling services integrated with antenatal care services will enable women with risks for having a child with haemophilia make an informed choice in relation to family planning. Among the respondents, 166 (85.6%) were aware that life expectancy may be close to normal with appropriate care and treatment for haemophiliacs. Life expectancy of a child born with haemophilia in Ireland is essentially normal with an excellent quality of life [2]. However this might not be the case in most developing nations where haemophiliacs were being under diagnosed and undertreated

5. Conclusion

Knowledge of clinical features and management of haemophilia should be improved among health workers, through continuing education. This would facilitate prompt recognition of cases and referral to experts for appropriate management. The use of multi-disciplinary approach which has been judged as the optimal model of care as well as training and re-trainning of experts will go a long way to stem the incidence of under diagnosis and under treatment of patients with haemophilia.

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