Assessing the quality of care for haemophilia at the Yaoundé reference treatment Centre of Cameroon

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With a recently established Haemophilia Treatment Centre (HTC) in Yaoundé, Cameroon, over a hundred people living with haemophilia have been recruited and followed up at this centre. This study aimed at assessing the quality of haemophilia care provided at the HTC, in order to monitor and improve patient care. In February 2014, the HTC was assessed using recommended markers. Although few, the logistics and reagents for the diagnosis and treatment of haemophilia were available. There were seven trained workers involved with haemophilia care, but the multidisciplinary care team was incomplete. A total of 113 people living with haemophilia (all males) had been registered and regularly followed up at the HTC. This study showed that the HTC of the Yaoundé University Teaching Hospital, although not yet ideal, allows for some degree of haemophilia patient care. Hence, it may be recommended to improve the centre and make it fully established in Cameroon. ***Blood Coagul Fibrinolysis*** **27:000–000**

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Introduction

Haemophilia is a sex-linked congenital bleeding disorder caused by a deficiency of coagulation factor VIII (FVIII) for haemophilia A or factor IX for haemophilia B. According to Stonbraker *et al.* haemophilia has an estimated frequency of approximately one in 10 000 births, with Haemophilia A being more common (80–85%) than haemophilia B [1]. Estimations based on the World Federation of Haemophilia (WFH)'s annual global surveys indicate that the number of people living with haemophilia (PLWH) in the world is approximately 400 000 [2], with about 75% of them living in developing countries [3]. Haemophilia is a chronic disease and managing it is both complex and costly because of its clinical manifestations and complications. However, with adequate treatment products and proper care, PLWH can live perfectly healthy lives. Unfortunately, about 75% of them receive little or no treatment. Implementing haemophilia care in sub-Saharan Africa remains a great challenge because this disease is not yet considered a national health problem in many countries [4]. In Cameroon, based on the estimations by the WFH, they may be up to 1800 PLWH. Challenges faced in the care of these PLWH include not only the multidisciplinary and costly nature of the care per se, but also the identification and diagnosis of these individuals. With traditional medicine still a dominant part of the Cameroonian culture, many with bleeding disorders may attribute it to witchcraft and other spiritual causes, and hence may not necessarily attend routine medical institutions for diagnosis. Although some degree of diagnosis was possible between the 1980s and 1990s using specific coagulation factor assays, this procedure was arrested because of the rising costs of coagulation screening tests and the low demand. Thus, until recently, management of bleeding disorders was mainly through the transfusion of blood and blood products, not even affordable to many. Hence, there is paucity of data on haemophilia and its management in Cameroon. To date, few available data report just over 100 diagnosed and registered cases [5]. The role of the WFH is to work together with national associations to promote the well-being of PLWH worldwide. It is in collaboration with the WFH initiative that a Twinning Programme was established in 2009, under the umbrella of the WFH, between the Haematology and Transfusion Service of the Yaoundé University Teaching Hospital of Cameroon and the Haemostasis Unit of the University Hospitals of Geneva. The aim of this twinning program was to create a Haemophilia Treatment Centre (HTC) in the hospital that would contribute to improve on the diagnosis and management of PLWH and other bleeding disorders in Cameroon. Because monitoring and evaluation is key to the success of any program, this study was carried out to assess the activities of this HTC and the quality of haemophilia care provided there using the structure and process aspects for assessing care quality.

Study design and methods

The cross-sectional descriptive study was conducted in February 2014 at the Haematology and Transfusion unit

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of the Yaoundé University Teaching Hospital of Cameroon, using a structured pretested data collection sheet which included selected indicators for assessment of care, adapted from: structure and function of centres for treatment and global management of haemophilia [6], Guidelines for the management of haemophilia, second edition and Diagnosis of haemophilia and other bleeding disorders, second edition [7]. Some of the data collected included demographic data, information on the structure and the activities of the treatment centre since creation. This study was approved by the Director of the University Teaching Hospital and ethical clearance was requested from the Ethical and Research Committee of the Faculty of Medicine and Biomedical Sciences. At the treatment centre, staff interviews were conducted. A checklist was then used to assess the laboratory section of the treatment centre. The storage of clotting factors, reagents and other treatment products was assessed based on international norms, for their availability and sufficiency's in quantities. The conditions under which all these are stored, the presence or absence of continuous electricity supply, and the current state of the equipment for storage and the storage products were all assessed. The registry of the HTC was also consulted to verify the quality of information that was recorded in it. Information was also obtained about the profile of the patients seen and followed up at the treatment centre. Other aspects assessed included the number of offices available for consultation, dedicated clinical areas for the management of patients, and geographical accessibility of the treatment centre within the hospital to the patients.

All data were coded in EPIDATA 3.1 and analysed using STATA 11.0 (STATA Corp. College Station, Texas, USA). The use of FVIII in international units per persons with haemophilia A (IUs per PWHA) for a Country was calculated by dividing the reported number of IUs of FVIII used by the reported number of PWHA. The FVIII use (IUs per capita) for a country was calculated by dividing the reported number of IUs of FVIII used by its total population in the appropriate year.

Results

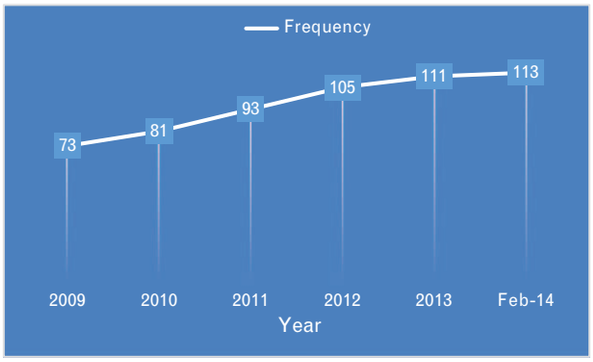
The treatment centre is an integral part of the Haematology and Transfusion unit of the hospital. This includes the main haematology laboratory, the blood bank and three consultation rooms. There are seven members of staff at the HTC including three physicians. The Chief of Service who is a haematologist, assisted by two full-time clinical biologists and a part time occasional haematologist. There is also a nursing staff and one physiotherapist. Most of the staff has received specialized training ranging from six weeks to 2 years in haemophilia care. Whenever appropriate, patients are referred to other services including Paediatrics, Radiology, Surgery and others. There was equipment available both for storage of reagents and clotting factors, including equipment needed for the diagnosis of haemophilia and other bleeding disorders except an analytic balance. They all met international norms and were all in good shape and functioning properly. Clotting factors were available in insufficient quantities. These factors were stored at +4 to +6°C. There was no fresh frozen plasma, no cryoprecipitate, desmopressine and splint cast as requested by WFH. Tranexamic acid was also not available at the treatment centre although it could be found in the hospital pharmacy for the patients to purchase as required. There were both adult and paediatric crutches available at the treatment centre. The reagents for the diagnosis of haemophilia and other bleeding disorders were available at the treatment centre, as well as for monitoring the prothrombin time and activated partial thromboplastin times during treatment. These were in sufficient quantities to last several months, and were stored at +4 to +6'C. Their storage conditions were considered average. The registry of the treatment centre was found in the sector of the laboratory used for haemostasis, which served as office space for one of the Clinical Biologists. This registry was created effectively at the start of the treatment centre in 2009. There were two separate consultation rooms used and another consultation room was lodged where the registry is found and which is also used as the laboratory for the diagnosis of haemophilia and other bleeding disorders. Patients requiring admission were admitted in the different services of the hospital as the case may be (e.g., paediatric cases were admitted in the Paediatrics Service). There are two entrances to access the treatment centre: the main entrance goes through a staircase and another one, which is level all through, goes through the back entrance.

By February 2014, with the help of the Cameroon Association of Haemophilia, the staff members of the treatment centre have been able to register and were following up a total of 113 PLWH (all males) (Fig. 1). The youngest patient was 1 year old and the oldest was 62 years old. There were three deaths recorded, which occurred because of internal bleeding. One had an intracranial haemorrhage following a head trauma, whereas the two others had bleeding from the gastro-intestinal tract. There were 94 (85%) of the 110 patients alive who had haemophilia A. Ten (9%) had haemophilia B and five (5%) were still unidentified, whereas one (1%) had both haemophilias A and B. They were also nine suspected cases of vWD disease. AQ5

No new cases were diagnosed in the year 2009 at the HTC, but since the year 2010, after the acquisition of a coagulometer in 2010, the working staff at the centre has been able to diagnose 38 new cases of haemophilia (Fig. 2).

A total of one million (1 589 182) international units of both clotting factors A and B had been used to treat

Fig. 1



Cumulative evolution of cases registered and followed up at the HTC of the YUTH since 2009.

patients by February 2014. The highest amount was used in 2011, but there was a significant decrease in the use of clotting factors in 2012–2013 (Table 1).

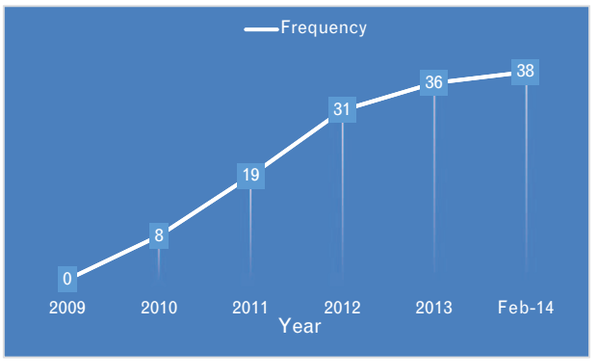
The use of FVIII in international units per person with haemophilia A (IUs per PWHA) for the reported number of PLWH A in Cameroon varied greatly at the HTC, with the highest consumption in 2011 and lowest in 2012 and a mean consumption of FVIII of 3 IUs per PWHA. This was similar in IUs per capita with a mean of 0.00021 (Table 2).

Five research activities have been carried out and of these; three have been published in international journals, whereas the two others were presented as posters during WFH congresses (Table 3).

Discussion

The aim of this study was to monitor and improve the quality of care of PLWH in Cameroon.

Fig. 2



Cumulative evolution of diagnosed cases at the HTC of the YUTH since 2009.

Table 1 Amount of clotting factor used at the HTC per year

Year	Amount used in IUs
2009	185 640
2010	345 880
2011	845 210
2012	69 002
2013	143 450
Total	**1 589 182**

HTC, Haemophilia Treatment Centre; IUs, International Units.

The HTC of the hospital was the only one in Cameroon. This made it difficult for patients found in different regions of the country to receive adequate care. This could explain why the three cases died in the Littoral, North and Far North regions of the Country. Even though it is the lone one, we found out that it was not also separate, but incorporated in the Haematology and Transfusion unit of the hospital. Most patients usually find it very difficult descending the staircase to get to the treatment centre, as a majority of them already have musculoskeletal complications. Hence, accessibility to the treatment centre may not necessarily be easy. Nevertheless, the way through the back entrance which is flat throughout and without steps, seems easier for them.

The fact that this treatment centre is incorporated into the Haematology and Transfusion unit without any increase in personnel could negatively affect the quality of care offered to PLWH because the workers are overwhelmed with the workload and may not optimally cater for PLWH. The incompleteness of a multidisciplinary care team could also greatly affect the quality of care delivered because all PLWH are supposed to be reviewed by the complete multidisciplinary care team every six months for children and yearly for adults following the recommendations of WFH [8,9], to assess the complete wellbeing of the patient, and develop individual comprehensive management plans. However, the treatment centre refers patients to appropriate quarters as the need arises. For example, surgical issues are referred to surgery and others disciplines as required, even if they are not members per se of the multidisciplinary team for treatment. This is not the case in South Africa where haemophilia care is highly organized and effective in the delivery of haemophilia health services [10].

Table 2 Amount of clotting FVIII in IUs per PWHA and IUs per capita used at the HTC per year

Year	Amount of FVIII used in IUs per PWHA	Amount of FVIII used in IUs per capita
2009	1924	0.00011
2010	3644	0.0002
2011	8988	0.0005
2012	651	0.00004
2013	No available data	No available data
Mean	3802	0.00021

PWHA, people with haemophilia A.

Table 3 Research and outcome done at the HTC since its creation in 2009

Researcher	Topic	Purpose	Academic year and place of defence	Published
Tiga Fombutu	Serological markers in people living with haemophilia	MD Thesis	2009–2010 in FMBS	Poster presentation in WFH Congress, 2011.
Didjoo Chantal	*Profile épidémiologique et Clinique des patients vivant avec hémophilie au Cameroun*	MD Thesis	2010–2011 in FMBS	In process
Annick Ndoumba	Conservation of fresh frozen plasma	*DES*	2010–2011 in FMBS	Poster presentation in WFH Congress, 2012
NGO Balogog Pauline	Frequency of inhibitors; in people living with haemophilia in Cameroon	*DES*	2012–2013 in FMBS	International Journal of Laboratory Haematology
Larissa Tene	Haemostatic changes in HIV cases	MD Thesis	2011–2012 in ISSS, UDM	The American Journal of Tropical Medicine and Hygiene

DES, Diplome d'Etudes Specialisés; FMBS, Faculty of Medicine and Biomedical Sciences; MD, Medicinae Doctorae; UDM, Université Des Montagnes.

Treatment was mostly donations from the WFH and from the twin partners of the Haemostasis Unit of the University Hospitals of Geneva. With the remarkably high cost of these clotting factors, this will explain why they are not readily available. The insufficiency of the two refrigerators could explain why the storage conditions of the treatment products were only average, as most of them were wet and others were sticking to the walls of the refrigerators, including the reagents and treatment products, which were all together in the same refrigerator. This sub-optimal storage conditions could alter the quality of the products.

Prophylaxis and home therapy are not offered by the treatment centre. This could be because of the fact that treatment products are not readily available and also because the few staff available are already stretched between haemophilia care and routine haematology. According to the Malmo protocol, clotting factors are given 25–40 IU/kg three times a week for haemophilia A and twice a week for haemophilia B; or Utrecht protocol where clotting factors are given 15–30 IU/kg three times a week for haemophilia A and twice a week for haemophilia B, in countries where prophylaxis and home therapy are practiced. The Malmo protocol has been effectively applied in Sweden for primary prophylaxis and many PLWH are now living normal lives in Sweden [11]. Meanwhile, home care for PLWH is already effective in South Africa and Zimbabwe because their respective governments are actively involved in funding for treatment products [12].

Also at the treatment centre, there was neither fresh frozen plasma nor cryoprecipitates, nor desmopressin, a synthetic hormone used in emergencies to stop bleeding in people with haemophilia A and Von Willebrand disease. This is not the case in the national haemophilia treatment centre in Cairo Egypt where they are now producing, storing and using well-tolerated cryoprecipitates to treat their patients [13]. As such, patients have other alternatives in the absence of clotting factors, which are more costly. Jennings *et al.* found out in England in 2009 that cryoprecipitates contain 200–1000 IU/dl FVIII, Von Willebrand's factor, fibrinogen and factor XIII [14]. All reagents for the diagnosis of haemophilia were available and in sufficient quantities, except those to fully diagnose vWD which were absent. This explains why none of the nine cases of the disease suspected at the treatment centre was confirmed.

All the 113 patients registered and followed up at the HTC were males, with the highest number of cases registered in 2009. This is because haemophilia is a sex-linked congenital disease that generally affects males because of a mutation in the X chromosomes of their mothers who serve as carriers. The youngest patient in our study was 1 year old, whereas the oldest was 62 years old. The death of three patients registered, as the creation of the HTC was because mainly of the absence of other haemophilia treatment centres in different regions of the country. Of the 110 alive, haemophilia A was predominant representing 94/110 (85%). This is similar to the results of a study carried out by Stonebraker *et al.* who found out that haemophilia A represents 80–85% of the estimated 400 000 PLWH around the globe by the WFH [15]. There was also one case of mixed haemophilia (haemophilias A and B).

The 38 new cases diagnosed at the treatment centre was unlike in the past where all suspected cases of haemophilia in Cameroon were sent through Centre Pasteur to European countries for diagnosis and were paid for. Today it is done freely and easily at the treatment centre.

From 2009 to the end of 2013, a total amount of 1 589 182 IU clotting factors have been used to treat patients at the HTC. The highest amount used in 2011, but there was a significant decrease in the use of clotting factors in 2012–2013. This significant decrease in treatment reagents in 2012–2013 could be explained by the fact that there are irregular supplies of clotting factors from the few available donors and the Cameroon government is not yet actively involved in the care of PLWH in the Country.

The use of FVIII in IUs per PWHA at the treatment centre varied greatly since its creation in 2009, with a mean of 3802 IUs per PWHA. This result is similar to

those obtained by Stonebraker *et al.* who found out in 2010 that the mean consumption of FVIII in IUs per PWHA for low-income countries ranged from 48 in Uzbekistan to 24 721 in Eritrea. The mean consumption of FVIII in IUs per capita was 0.00021 at the treatment centre. This is lower than those obtained by J.S. Stonbraker *et al.* in the same study who found out that FVIII use (IUs per capita) for low-income countries varied from 0.0004 in Nigeria to 0.1287 in Eritrea, where a low income country according to the World Bank's Economic classification of 2006 had a Gross National Income per capita (in US dollars) of $905 or less [16]. One international unit per capita is approximately 20 000 IUs per patient, which is what the WFH suggested as a minimum clinical target for haemophilia care [17]. The low FVIII use (IUs per capita) per patient at the treatment centre could explain why Didjo'o Chantal found out in her study at the treatment centre of Cameroon in 2012 that 72.3% of 88 PLWH (mostly PWHA) had developed osteoarticular complications.

The researches done at the HTC of Cameroon have helped to improve on patient care. For example, close monitoring and doses of clotting factors were adjusted for some patients who were found with complications and inhibitors in two of the studies.

Conclusion

The study was limited by the absence of some files and data at the HTC of Cameroon, which made it difficult to report all the activities of the centre. Taking into account this limit, we concluded that the treatment centre of the hospital has been evolving since its creation, in equipment and other facilities and logistics for haemophilia care, but was not yet ideal. The few available trained personnel (07) were stretched between general haematology and haemophilia care. Although not yet comparable to the recommended standards, the creation of this HTC in 2009 allowed for some degree of improved haemophilia patient care. Hence, it may be recommended that support and guidance be offered to enhance haemophilia care throughout the national territory of Cameroon.

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The manuscript has been seen and approved by all authors, it is not under active consideration for publication, has not been accepted for publication, nor has it been published, in full or in part.

Conflicts of interest

There are no conflicts of interest. AQ6

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