

Article

Sickle Cell Disease in the Democratic Republic of Congo: Assessing Physicians' Knowledge and Practices

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Abstract: Background: Sickle cell disease is a major public health issue in the DRC while it is still poorly understood by health professionals. The objective of this study was to assess the knowledge and practices of Congolese physicians treating sickle cell disease (SCD) in order to identify areas for improvement in clinical care. Methods: This is a descriptive observational study conducted on Congolese physicians using a questionnaire. Participants were evaluated on a pre-established answer grid. Results: A total of 460 physicians participated, including 81 women (18%) with an average age of 35 years (range 25–60 years). Most physicians were general practitioners. Although self-assessment of their level of knowledge on SCD was estimated as average to good, less than half of the participants (n= 460; 46%) reported adequate management of vaso-occlusive crisis but only 1% of them had received specific training on SCD. Most physicians reported difficulties both in terms of diagnostic (65%) and management (79%) options of SCD patients. This study showed also that 85% of them did not have access to diagnostic tools for SCD. Conclusions: Insufficient knowledge on SCD, poor diagnostic and treatment options may contribute to morbidity and mortality of patients living in the DRC. Interventions aiming at improving physician's knowledge, patient's follow-up and treatment access are needed. Specific training alongside existing programs (HIV, malaria), early diagnosis of the disease, and creation of patients' advocacy groups should improve SCD patient's care.

Keywords: sickle cell disease; k knowledge assessment; practices; physicians; Democratic Republic of Congo

1. Introduction

Haemoglobinopathies, mainly comprising thalassemia and sickle cell anemia, are inherited conditions. Currently, almost 5% of the world's population has a mutation in one of the globin genes. Each year, nearly 300,000 infants worldwide are born with thalassemia syndrome (30%) or sickle cell disease (SCD) (70%)[1]. The major forms require rapid diagnosis and treatment but are still poorly recognized by healthcare professionals, causing misdiagnosis and inadequate support, factors that explain high morbidity and mortality rates in these patients [2]. It is estimated that more than 75% of sickle cell patients live in sub-Saharan Africa. The fact that the epidemiology of thalassemia in the Democratic Republic of Congo (DRC) is not known can be partly explained by the rarity of major forms [1]. In contrast, SCD is a common disease in the DRC, with 2% of newborns homozygous for hemoglobin S, representing around 40,000 births per year [3,4]. Although this figure is epidemiologically significant, the disease remains relatively unrecognized, resulting in a high mortality rate in a country with limited resources [5]. SCD is recognized as a public health problem in the DRC and benefits from national guidelines for the control and management of SCD through the national SCD program (PNLCD in french) established in 2001.

SCD is clinically expressed from the age of three months. It manifests variably concerning its frequency and intensity, classically represented as a triad of episodes of acute anemia, painful episodes of vaso-occlusive crisis (VOC) and susceptibility to infection [6].

In the DRC, high population growth combined with significant health challenges has created an urgent need for healthcare workers. Also, human resources for health are proving to be a serious problem for the sector as a whole: a decline in professional quality, the flowering of schools and universities offering cheap medical education, few qualified staff. The flight of qualified doctors out of the country in search of better working conditions and lives affects hundreds of Congolese doctors who will pursue careers in South African health training. In 2012 the Ministry of Health's Human Resources Directorate estimated the DRC's staff of 5,967 doctors for a population of 70 million. That equates to 1 doctor per 11731 inhabitants [7]. We did not find reliable and up-to-date data on the total number of physicians and their distribution according to age, sex and between provinces. This is explained by the recent mass exodus of physicians to Southern Africa, by the number of physicians working in the private sector and by the migration of health personnel within the country [8,9]. There are three major traditional universities for training physicians (University of Kinshasa, Lubumbashi University, and the University of Kisangani), and the emergence of private universities or under-agreement medical training is not sufficient to meet the healthcare needs. This is partly explained by the use of training programs that are not adapted to local needs, a poorly organized training framework, a lack of appropriate internship places, a qualitative and quantitative shortage of teachers, and a lack of model schools for testing and adapting the training programs [8].

To overcome some of the shortcomings of physicians' knowledge in certain pathologies such as HIV / AIDS or malaria, the National HIV and malaria programs receiving support from world Health Organization (WHO) and regularly organize capacity building training of caregivers and physicians, which result in improved care and management of these diseases. Significant efforts are being made to integrate vertical disease control programs into the primary health care package, particularly in the fight against HIV and Malaria [10,11].

The majority of Congolese have no access to quality health care. Moreover, the insufficient knowledge among physicians may contribute to the high mortality and morbidity rates related to sickle cell disease. This assumption led us to evaluate the knowledge and practices of Congolese physicians concerning SCD patient's care and to propose concrete actions to improve it.

2. Methods

This is a descriptive observational study conducted on a population that included physicians, predominantly in 5 cities within the DRC. To collect the information, a questionnaire was delivered online or directly by hand via existing health structures or existing networks in Kinshasa (DRC

capital), Lubumbashi (Haut Katanga), Mbuji-Mayi (Kasai-Oriental), Bukavu (South Kivu) and Kananga (Kasai-central) and other cities. The selected cities are the most heavily populated in the DRC [12] and contain, at minimum, a university medical school.

The study comprised any general practitioner (GP) or specialist practising in the DRC and registered in the national council of the order of physicians who agreed to participate in the study. Any doctor registered with the national council of the order of physicians obtains a serial number of doctors (CNOM in French) which authorizes him to practice medicine in the DRC.

Questionnaires excluded were those with less than 50% questions answered.

The questionnaire was developed in collaboration with the medical faculty of the University of Mbuji-Mayi in the DRC and the department of clinical chemistry at the Université Libre de Bruxelles (ULB). It was trialled on 30 physicians in Mbuji-Mayi. Answers from the trial were used to improve the questionnaire, especially for the use of unequivocal formulations. The finalized questionnaire was submitted to the Department of Epidemiology and Biostatistics (School of Public Health, ULB) for validation.

The questionnaire comprised 57 questions divided into five groups: (1) physician profile; (2) SCD clinical practice; (3) diagnosis and clinical management of haemoglobinopathies; (4) organizational management of SCD; and (5) concerns regarding haemoglobinopathies and self-assessment of knowledge of these.

Questions were either closed (dichotomous or nominal) or open. Open questions were evaluated as either a right answer (the answer was correct according to a grid of pre-established responses) or an inappropriate response (the answer was irrelevant, inadequate or wrong) and self-assessment of knowledge on hemoglobinopathies was recorded through a scale of Likert values [13]. The self-assessment scale comprised 5 numeric values ranging from 1 to 5. The scale was 1 = poor knowledge, 2 = insufficient knowledge, 3 = average knowledge, 4 = good knowledge and 5 = very good knowledge.

Recommendations of management of vaso-occlusive crises was defined by the prescription of analgesic (paracetamol, non-steroidal anti-inflammatory or morphine), hyperhydration and the treatment of the factor favoring the crisis (infections: malaria, sepsis etc.), while recommendations of management of acute anemia was defined by the prescription of red blood cell transfusions.

A large city is defined by its urban landscape and the presence of an international airport directly connected to foreign countries while a remote city is defined by a semi-rural landscape and is located inside the country, without direct contact with foreign countries.

No instructions were given that could guide responses. In the case of a non-response, doctors were contacted repeatedly by either mail, phone, SMS, Facebook, LinkedIn or direct contact.

3. Ethical Approval

The study protocol had been reviewed and approved by the Ethics Committee of the Medical Faculty at the University of Mbuji-Mayi (Fac-Méd / UM / CE / 032/2016). Informed consent was included at the beginning of the survey questionnaire. The questionnaire was anonymous and used for scientific purposes and the participant was free to participate or not and at any time during the questionnaire, he could stop his participation. The recording of the responses to the questionnaire did not contain any information that could identify the doctors interviewed, unless one of the questions was asked explicitly and they were free to fill in or not. If the interviewee has completed the questionnaire and has given his identity and contact details, he was assured that no information concerning his identity could be recorded in connection with his responses. The identities or details were managed on a separate database where it is only indicated that the respondent had (or not) completed the questionnaire and if necessary in future projects on hemoglobinopathies, we can use this information to contact them. There is no way to match the identities of the respondents with their responses on the questionnaire.

4. Statistical Analysis

Statistical analysis of the collected data was performed using EPI Info software (USA, CDC Atlanta, version 5.0, 2008). The data, as appropriate, is represented by the mean and confidence interval at 95% for numerical parameters and absolute or relative frequency for temperamental parameters. The chi-square test was used to compare the level of knowledge of the participants, with a p -value < 0.05 considered significant.

5. Results

5.1. Physicians profile and practice

The questionnaire was sent out from June 2017 to September 2018 to 956 physicians, 475 responses were recorded. Of these, 15 were excluded: 7 practising outside the DRC, 3 did not give their serial number of doctors and 5 responded to less than 50% of the questions.

The response rate was 48% (460/956). The median age was 35 years (range 25-60 years) and a minority of participants (18%; 81/460) were women.

The percentage of participants was geographically distributed as Mbuji mayi 23.6%, Kinshasa 23%, Lubumbashi 22.6%, Kananga 17.6%, Bukavu 7% and other towns 6%. Most of the participants were general practitioners (82.4%) and few specialists: i.e. pediatricians (4.1%), public health specialists (4.1%) or other specialists (9, 4%). The majority of them (86%) had a clinical practice while a few of them (1%) had taken a diploma course on major SCD syndromes. Almost half of them (49%) had less than 5 years of clinical experience and few (10%) had more than 10. The majority (52,2%) worked in a General Reference Hospital (HGR), the others (26% and 13%) worked in health centers or a private hospital, respectively. Sickle cell disease clinical practice and knowledge among the physicians are described in Table 1.

5.2. Diagnosis and Clinical Management of SCD

Physicians reported that the circumstances of SCD diagnosis are mainly clinical (80%). Emmel test (sickling test) [14,15] was the most frequently requested test (83%). Diagnostic test was requested most often at the time of disease complication (99%) and rarely for neonatal screening (1%). Approximately two-thirds (65%) reported the difficulty of performing hemoglobin electrophoresis due to lack of equipment. For the vast majority of the participants (91%) the sole disease is sickle cell anemia (homozygosity for hemoglobin S). Most physicians (99%) are not aware of the other syndromes (thalassemia, HbSC, etc.). The different SCD syndromes encountered are presented in Table 1.

Regarding the recommendations of management for VOC and acute anemia, were met for less than half (44%) and the majority (86%) of physicians, respectively. After managing sickle cell crisis, few physicians (24%) refer patients to an appropriate structure or a support center for future monitoring.

Few physicians (9%) prescribed hydroxyurea and only 26% of them thought that hydroxyurea is available in a local pharmacy. If most of them weren't aware of the price of hydroxyurea (74 %), they responded that it is not affordable for their patients (91%). The majority (95%) also reported that the therapeutic means for SCD management are not sufficient and most of them (79%) suggested that conditions have to be implemented to improve SCD patient's care.

The physicians interviewed noted certain concerns: in terms of training, the lack of local training of a university degree in sickle cell syndrome (79%), but also in terms of patient follow-up, the difficulty of organizing regular medical follow-up and the rigorous application of management recommendations.

Table 1. Clinical practices and opinions of Congolese physicians about sickle cell disease (n= 460)

1. Clinical practices	n	(%)
Do you follow patients who are?		
- Homozygote for HbS	452	(98%)
- Heterozygous for HbS	4	(1%)
- Compound heterozygous for SBeta-thalassemia	4	(1%)
- Compound heterozygous for other sickle cell syndromes	0	(0%)
Do you treat sickle cell patients?		
- ≤15 years old	452	(98%)
- >15 years old	8	(2%)
Physicians who prescribe hemoglobin electrophoresis	101	(22%)
Physicians who announce diagnoses to their patients	301	(66%)
Physicians who giving appropriate advice to sickle cell patients *	147	(32%)
Physicians who refer patients for psychological support to patient's associations	46	(10%)
Physicians who use traditional medicines to treat sickle cell disease	24	(5%)
Physicians who collaborates with NGOs involved in the fight against sickle cell anemia	60	(13%)
Physicians who receive patients referred by traditional healers	24	(5%)
Physicians in possession of the national protocol for the management of sickle cell disease in their structures	67	(15%)
Who knows at least a reference center for sickle cell disease in his hometown	318	(69%)
2. Opinions of physicians		
Organization of a university diploma in major sickle cell syndromes in his hometown	0	(0%)
Physicians who know their phenotypes	212	(46%)
Physicians who knows at least one sickle cell association in their city of residence	78	(17%)
Physicians who think that sickle cell patients are afraid of the disease	386	(84%)
Physicians who think that sickle cell patients keep hope	294	(64%)
Knowledge of sickle cell patients is coming from:		
- Patient association	66	(14%)
- Support structure	69	(15%)
- Association aid	124	(27%)
Physicians who know the different types of major sickle cell syndromes		
- HbSS	454	(99%)
- HbSβThal	4	(1%)
- HbSC	2	(0%)
- HbSOarabic	0	(0%)
- HbSDpunjab	0	(0%)
- HbSE	0	(0%)

NGOs: non-governmental organizations, HbSS: homozygous S, Composite heterozygote (SC, S-beta thalassemia, SOarabic, SDpunjab and SE),

* (rehydration, regular medical follow-up, adherence to prophylaxis (folic acid, oral penicillin, dewormers and antimalarials) ± 1 one of the advice on factors triggering crisis: intense physical exercises, thermal variations, emotional stress etc).

5.3. Self-Assessment and Concerns

For most physicians (84%), hemoglobinopathies are a concern in their clinical practice and they (79%) reported that there are conditions to be met to improve services and treatment burden of sickle cell patients. Training on major sickle cell syndromes, the availability and accessibility of

hydroxyurea and diagnostic means (hemoglobin electrophoresis) were the most often reported aspects that must be improved.

Self-assessment of the level of SCD knowledge was reported as between average and good, while it was low or insufficient for thalassaemia (Table 2).

Table 2. Self-assessment of the level of knowledge of Congolese doctors on hemoglobinopathies.

Parameters	Quotation/5*
	Sickle Cell Anemia
Advice to patients	3.8
Biological diagnosis	3.7
Clinical diagnosis	3.9
Genetic level	3.2
Pathophysiology map	3.9
Medical management	3.7

* Likert scale.

Compared to those practising in remote cities of the DRC (138/348; 40%) or having a clinical practice of more than 10 years (33/212; 24%), physicians practising in large cities (Kinshasa and Lubumbashi) or having a practice of less than 10 years had a better knowledge of the management of vaso-occlusive crises 66% (74/112) and 76% (179/212) ($p = 0,000$), respectively.

5. Discussion

Sickle cell disease is a chronic disease with multi-organ involvement. Its management is complex and in particular in the DRC where access to care, the low socioeconomic level, and other factors such as malaria play a significant role in its morbidity and mortality rates. For optimal management, a first step is to diagnose the disease, then to establish appropriate follow-up and treatment.

This study aimed to determine, through a descriptive survey of physicians, factors that could improve patient's care in the DRC. The survey aimed to assess the knowledge and practices of 460 Congolese physicians spread across the country but mainly in three big cities in terms of population like Kinshasa, Lubumbashi and Mbuji-Mayi, but also smaller cities like Kananga, and Bukavu.

The participation rate was 48% as observed for similar surveys [16,17]. However, it took over a year to reach this participation rate at the cost of diligent follow-up and several reminders. The average age of the physicians was 35 years (range: 25–60) and in a 2012 study, the average age of the person's health care age was 33 [7]. These results correspond to the 25-54 age group that constitutes the majority of the population in the DRC [12]. The very low participation of women (18%) may reflect the low level of education of women in the DRC [18]. Physicians lived in the three most populous big cities of the DRC i.e. Mbuji-Mayi (23.6%), Kinshasa (23%) and Lubumbashi (22.6%) and are mainly general practitioners (82.4%) [12]. Only 1% (6/460) of them had already received formal training on major sickle cell syndromes. This is not surprising because specialized medical training is limited to a few universities meeting the criteria for postgraduate organization in the DRC that cannot absorb all the specialization requests of the country [19]. The physicians also encountered difficulties in organizing the systematic follow-up of sickle cell patients. This is partly due to the local context. In Africa, for geographic and socio-economic reasons, and particular representations of the disease, patients have little access to health structures. They consult most frequently during complications and most often in health facilities where specialists are not present [20]. However, even these specialists have received little or no specific training on the management of SCD. If the training and health support center (CEFA) based at Monkole Hospital in Kinshasa organizes training on sickle cell anemia, these are chargeable and do not cover needs throughout the country while other training course offers are rare [21]. Health personnel must be regularly trained and equipped for the basic management of SCD, in particular on early detection, crisis prevention, and care, this concern was raised among the participants [22].

Common diagnostic methods in newborns and beyond are Hb separation techniques such as isoelectric focusing, high-performance liquid chromatography and capillary electrophoresis [23]. In this study, 65% (298/460) of the physicians reported they had diagnostic difficulties limiting them to clinical diagnosis and biological confirmation only by a sickle cell test. However, the Emmel test is a non-specific test with its limits of interpretation and it is only routinely available in university hospitals and some private laboratories [24]. Newborn screening for SCD was adopted in 1972 in the United States and was spread to other continents [25]. Newborn screening made it possible to identify newborns suffering from SCD before the onset of symptoms to prevent infectious complications, VOC and to reduce the mortality rate of sickle cell children by up to 84% [25,26]. However, in this study, the circumstances of SCD diagnosis were mainly in the presence of symptomatic patients and neonatal diagnosis was only reported by 1% of the physicians. This confirmed data in the literature demonstrating that neonatal screening is still very exceptional in Sub-Saharan Africa and no African country has so far maintained neonatal screening for SCD in its national program [26–28]. Currently, there are rapid tests for sickle cell disease who have good sensitivity and good specificity [29]. The use of these rapid tests which could be coupled with the rapid malaria tests and by equipping the reference centers with a confirmation technique, such as the iso-electrofocusing technique, could also be a strategy to be taken into account in order to improve the diagnosis and management of sickle cell patients.

The management of vaso-occlusive crises is not optimal with more than half of the participants reporting its inadequate or insufficient management. The same observation was made in Nigeria [22]. Nevertheless, most physicians (95%) thought that the therapeutic means for SCD management are not sufficient and is unsatisfactory. They point out that there are conditions to be met in order to improve patient's care. In the management of SCD related pain, it is recommended to use good hydration and the administration of level three pain relievers such as morphine in case of uncontrollable pain [30]. However, in the DRC, access to morphine is very limited, pain management is most often restricted to level two analgesics. Only physicians who resided in large cities and had less than 10 years of practice had good knowledge of the management of vaso-occlusive crises. Training opportunities in large cities are probably one of the reasons for this better knowledge.

If, on the other hand, 86% of participants had a good attitude towards an acute anemia and recommended a blood transfusion. Anemia of whatever origin is frequent in Africa and allows to explain its management is well known. This is essential because acute anemia is responsible for a high mortality rate in sickle cell patients in sub-Saharan Africa [31,32]. Its prevention and effective management dictate specific strategies adapted to the African context [24].

The use of hydroxyurea has proven its effectiveness and its long-term tolerance in northern countries [33,34]. Because of its cost, its availability and the risk often put forward of male sterility, it is rarely used in Africa. The same observation was made in our study. However, monitoring hydroxyurea treatment must not be overlooked, and therefore the ability to guarantee the biological and clinical controls necessary for monitoring this treatment. The assurance of regular follow-up, affordability for families and knowledge of the disease and its treatment by health staff are prerequisites for the initiation of this therapy [24].

Almost all of the doctors surveyed (99%) are unaware of the different forms of major sickle cell syndromes (SS, SC, S β -thalassemia, etc.) and the scarcity of specific training on sickle cell anemia and the lack of organization of this specialization in DRC can explain this result; on the other hand, a majority of patients in the DRC are homozygous for hemoglobin S [4].

The associations of sickle cell patients are very little known by the doctors interviewed. Due to lack of funding, associations carry out little activity and, like other pathologies such as malaria, their visibility is very reduced because very little publicized in the DRC.

Physician's self-assessment of their level of knowledge of SCD shows that their level is good to medium. However, this self-assessment does not seem to be objective. For example, 90% of doctors interviewed reported not know the other major sickle cell syndromes (S β -Thalassemia, SC etc.), but also they have the difficulty of having a diagnosis of certainty of SCD and the majority of them uses Emmel test, which is unreliable to differentiate those who have the disease from the carriers. These

items are to be considered in the interpretation of the self-assessment of our respondents. Other studies carried out in sub-Saharan Africa have come to the same conclusions [35]. In Nigeria, the level of care and knowledge of health professionals in the diagnosis and prevention of sickle cell crises is low [22]. Neonatal screening can be a means of improving the knowledge of healthcare teams and helping to prevent complications, as has been shown in an experiment in Burkina Faso and Nigeria [36].

Improvement in sickle cell care can be achieved in different fields or areas. SCD is a serious public health issue in the DRC but only a few hours are dedicated to the disease in the physician training course content. Para-university training, like those offered at the national level for HIV and malaria programs, but also the development of a university diploma on sickle cell disease are necessary to strengthen the level of knowledge of health care providers. In resource-limited countries, implementation of a neonatal screening program constitutes a real challenge; use of new rapid sickle cell tests, that could be coupled with rapid malaria tests, could be another option for all children under 5 years with signs of SCD (anemia, infections, pain). The implementation of a reference center that could confirm the diagnosis for example by iso-electrofocusing is also needed. It must be based on past experiences obtained by African teams. One of the other key elements for better patient management is patient associations. Supporting and encouraging the creation of associations for sickle cell patients makes it possible to ensure various essential actions; among these is the awareness of the population about this disease and support from the government for various concrete actions such as that of setting a minimum monthly cost, in the absence of free care, for all sickle cell children to ensure them a regular follow-up.

6. Conclusions

The level of knowledge and practices of Congolese doctors on SCD could be improved. Targeted training, both theoretical and practical, adapted to the local context and which can be combined with existing programs (HIV, Malaria) would improve the level of knowledge and optimize sickle cell patient's care. The extension of national guidelines for the management of SCD to health workers is needed to help improve the level of knowledge and practice about the disease. Diagnostic tools (rapid sickle cell test) and therapeutic means accessible to patients in fragile socio-economic situations (folic acid and oral penicillin, anthelmintic and antimalarials), pain management by morphine as well as treatment with hydroxyurea must constitute a complementary line of thinking for the training of health personnel.

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