Improving the outcome of Sickle Cell disease patients in a resource limited setting Sudan Sickle Cell Anemia Center (SSCAC): a promising and developing experience

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ABSTRACT

This is a descriptive health service research and quantitative study, based on a population of 1400 patients with sickle cell disease, pediatric and adult population. Their age, gender, and frequency of follow-up and hospitalization are recorded from the statistical records and analyzed. And the description of the health facility and the services provided by it was compiled from observations and interviewing the staff.

Specific clinics for children and adults with sickle cell disease are available in several African countries but only a few countries have newborn screening programs [17]. Sudan Sickle Cell Anemia Centre (SSCAC) is the only specialized center for SCD in Sudan for children and adults. Its location in Al Obeid in the West is Justified by the highest percentage of patients in Sudan being in the West region[6].

During the six years of its work, the number of registered patients increased from 112 to 1400 patients with the age range of 3 months to 34 years. The percentage of male patients is 56%. More than half of the registered patients were of the under 5 age groups. This is in concordance with a previous study done in Al-Obeid where the under five years age group was dominating[11].

It is recommended that:
-The World Health Organization is recommended to provide technical and financial support to SSCAC in framing policies and strategies for the prevention and management of sickle-cell disease.
-The collaboration of the Sudan Ministry of Health, the WHO, and the non-governmental organizations with the center is needed to support constructing buildings, providing ambulance, the diagnostic equipment including Genetic screening, needed treatment facilities, transportation methods for patients from areas of Kordofan and Darfur to the center, and help funding research on sickle-cell disease to increase life expectancy and improve patient's quality of life.
-Funding resources for sickle-cell disease prevention and control, including training of staff to offer genetic counseling and health education, in addition to the introduction of newborn screening services in the center.
-Establishing similar specialized centers in Khartoum and different states of Sudan to provide easier access to services for most of the patients.

Key words: **Sickle Cell disease, Sudan Sickle Cell Anemia Centre (SSCAC)**

Introduction

Sickle cell disease is an autosomal recessive disorder that consists of a group of disorders that are characterized by the presence of sickle hemoglobin (Hb S). In Sickle cell disorders, hemoglobin S is formed as a result of the substitution of valine to glutamic acid in position number six in the Beta globin chain [1]. There are more than 700 structural hemoglobin variants but the commonest in Africa are Hb S and Hb C. [2]. When the red blood cells are exposed to hypoxia, their membrane is distorted producing the characteristic sickle-shaped cell which occludes small capillaries and venules and causes tissue ischemia, acute pain, and gradual end-organ damage [3][4].

In 2010, the percentage of newborns with SCD in sub-Saharan Africa was 79%, and this proportion is expected to increase to 88% by 2050[5].

Africa has high mortality rates ranging from 50 to 90% for those aged less than 5 years. This high mortality is due to the lack of several facilities like prenatal diagnostic services, systematic follow-up, and life-saving measures such as penicillin prophylaxis, vaccination for common bacterial diseases, and the provision of disease-modifying treatment with Hydroxyurea, and poor access to hematopoietic stem cell transplantation [4]. Infectious diseases like malaria may also play a role in increasing the severity and mortality[5].

The prevalence rate of sickle cell anemia in Sudan ranges from 2 to 30.4%. The highest prevalence of SCA in the Sudanese population is found in Western Sudan residents[6]. Studies performed in different cities in Sudan relating SCA to Sudanese tribes also concluded that most of the cases were from tribes belonging to the western area in Sudan and those migrating from Western Africa and Sudan, for example, a study done in Gedarif state in Eastern Sudan showed a high rate of sickle cell gene among the population that migrated from the west[7][8][9][10]. The Misseriya tribe in Kordofan and Darfur showed the highest rate of sickle cell disease in Sudan. This is related to the increased consanguinity rates and the rate of first-cousin marriages in Sudan[9]. In El Obeid, high frequencies of sickle cell disease were found among Falatah and Jawama, 80% of children's parents were relatives or from the same tribe[11].

Because of this increased prevalence of SCD in west Sudan, it is of high importance to improve the quality of care for sickle cell disease patients in this area. According to the WHO definition quality of care is defined as the extent to which health care services provided to individuals and patient populations improve desired health outcomes. To achieve this, health care must be safe, effective, timely, efficient, equitable, and people-centered [12].

These points are described by WHO as being safe; meaning delivering health care that minimizes risks and harm, effective service based on scientific knowledge and evidence-based guidelines, reducing delays in providing and receiving health care, efficient service that maximizes resource use and avoids waste, and equal service for all people from different genders, race, ethnicity, geographical location or socioeconomic status and providing care that takes into account the preferences and aspirations of individual service users and the culture of their community[13].

Providing health care for SCD patients requires trained professionals and a social support system so that their physical, emotional, psychological, and financial needs are fulfilled. Unfortunately, SCD patients in most sub-Saharan countries have limited access to clinical, health educational, social and psychological care[14].

The Specialized sickle cell centers are important in improving quality of life, reducing health care costs, and reducing health care utilization rates among patients with SCD[15]. Several strategies have been proposed to improve the quality of care for patients with SCD. System approaches include comprehensive sickle cell centers, hub and spoke models, satellite clinics, and telemedicine [13].

Sudan Sickle Cell Anemia Centre (SSCAC) is a voluntary, national, non-governmental organization accredited by the Federal Humanitarian Aid Commission. It was established in 2012 in El Obeid, North Kordofan State. As the city formerly acted as the capital for the whole Kordofan region, establishing SSCAC in El Obeid is justifiable. It was started as a sickle cell clinic and patients were observed by a senior pediatrician then evolved gradually when laboratory investigations, genetic counseling, and training packages were added. From 2015 onward, SSCAC spread in the form of sickle cell clinics in different states of Sudan; South Kordofan, West Kordofan, and South Darfur. The core mission of SSCAC is to improve the quality of all services dealing with Sickle Cell Disorders, thereby improve the quality of life for people with Sickle Cell Disorder.

Methodology and description

This is a descriptive health service research and quantitative study, based on a population of 1400 patients with sickle cell disease, pediatric and adult population. Their age, gender, and frequency of follow-up and hospitalization are recorded from the statistical records and analyzed. And the description of the health facility and the services provided by it was compiled from observations and interviewing the staff.

SSCAC interventions in El Obeid – the headquarters that is located in El Obeid Specialized Pediatric Hospital include; medical care for those presentingt with sickle cell crises at the emergency department of the hospital and sickle cell clinic for regular follow up for both pediatric and adult patients. The follow-up services are made up of clinical examination, growth assessment and routine laboratory tests. Basic laboratory investigations to establish the diagnosis and to continuously assess the patient status are available. Regularly patients, especially at age 6 – 16 years, undergo Transcranial Doppler assessment as an effective measure for CVA prevention. Echocardiography is an emerging service for selected cases.

Health promotion activities are carried out in the form of genetic counseling sessions for parents and some patients, as well as families. Raising awareness of the public is conducted through talks in schools, hospitals, universities and people gatherings,

radio programs, and television shows. As a part of prevention measures, pneumococcal vaccine is available for patients especially under-fives.

As building capacity will guarantee good performance in medical care and counseling, several training activities were conducted.

Research in sickle cell disorders is inconsistent in different aspects of the disease; SCD mapping, the natural history of the disease, accessibility, and sustainability of medical care, effectiveness and impact of dedicated centers and clinics, etc. Many kinds of research that touch on viable issues regarding sickle cell disease are conducted. The research ranges from KAP studies to molecular studies on the disease haplotypes and correlations between phenotypes and single nucleotides polymorphism (SNPs).

Ethical consideration

Informed consent was obtained from the managers of El-Obied specialized pediatrics Hospital to which the center belongs and is part of that hospital. They accepted to provide us, as authors, the free use of all the data and statistics of the SSCAC.

Findings

Six years have passed since the foundation of SSCAC in El Obeid. The number of registered patients increased from 112 to 1400 patients with an age range of 3 months to 34 years. Among patients, two thirds (63%) are from North Kordofan State, 52% are under five years, males are 56%.

Up to date, about 3400 patients have ben provided with the benefit of the sickle cell clinic for both pediatric and adult patients, with only 15 – 25% of total registered patients visiting the clinic regularly and the percentage of patients presenting with crises and the need to be hospitalized is only 10 – 20% of total registered patients. The number of patients who visit the pediatric clinic is around 100 – 160, while adults´ clinic is about 20 – 30 per month.

The training was started earlier; courses and workshops were organized for doctors, nurses, medical assistants. Limited activities were carried out due to a shortage of resources and lack of funding. Here are some events;

	Course / workshop	Target group	Number trained
1	Nursing sickle cell patients; basic skills	Nurses	20
2	Basic skill in TCD (Transcranial Doppler)	Doctors, Nurses	16
3	Genetic counseling course	Health workers	15

Research works performed at SSCAC till the year (2018)

SSCAC studies				
No	Title	Year	Level	Publication
1	Sickle Cell Anemia among Children in El Obeid Hospitals, Sudan: A clinical and hematological study	2013	Study	IJMRHS • International Journal of Medical Research &Health Sciences, 2018, 7(11): 66-71
2	Knowledge and attitude of health professionalstowards sickle cell disorders in El Obeid Teaching Hospital, Sudan	2014	Undergraduate	
3	University students in Sudan: knowledge, attitude toward sickle cell disease and pre-marital genetic counseling	2014	Undergraduate	
4	KAP study among families of patients with sickle cell disease in El Obeid, Sudan in 2013.	2014	Undergraduate	
5	Prevalence of alloimmunization in multiple blood transfusion sickers in El Obeid, 2015.	2015	MSc	
6	Management of sickle cell crises in El Obeid Hospitals, 2014	2015	Undergrad	
7	C - reactive Protein Level and WBC Count as Biomarkers for vaso-occlusive crisis among patients with Sickle Cell Disease	2015	MSc	American Journal of Medicine and Medical Sciences2015; 5(6): 283 – 286.
8	Relationship between Painful Crisis and Serum Zinc level in Children's with Sickle Cell Anemia	2016	MSc	
9	Association of Endothelial Nitric Oxide Synthase Gene T-786C polymorphism with Complication of Sickle Cell Anemia among Children in Khartoum 2017	2017	MSc	
10	Assessment of Microalbuminuria and Lactate Dehydrogenase as Early Indicators of Renal Impairment among Sudanese Children with Sickle Cell Anemia in North Kordofan State	2016	MSc	

11	Determination of Selected Trace Elements among Children with Sickle Cell Anemia in Elobied City - North Kordofan State	2017	Undergrad	European Journal of Pharmaceutical Sciences 2017, 4(10) 855-857.
12	Estimation of potential patients for stroke risk in children with sickle cell anemia using ultrasound in Jaffer ibn Aouf Pediatric hospital 2015	2018	PhD	
13	Prevalence of Sickle Cell hemoglobin in Shikan locality, North Kordofan State, 2017	2017	Undergrad	
14	Impact of Sickle Cell Disease in Renal Arteries Blood Flow Indices Using Ultrasonography	2017	PhD	International Journal of Medical Imaging 2017; 5(2): 9-13.
15	*Evaluation of Transcranial Doppler Abnormalities in Children with Sickle Cell Disease in Elobied Specialized Children`s Hospital in the Time Period from December 2016 to February 2017.*	2017	MD	
16	Frequency of HbS Gene Among Population of Northern Korodofan Area; Hematological and Molecular Analysis	2017	PhD	
17	Pregnancy and birth outcome and essential nutrient status of Sudanese pregnant women with sickle cell disease	2017	PhD	
18	Role of trans-cranial Doppler ultra-sound in Sickler patients as a screening tool, in EL-Obied North Kordofan State in Kuwaiti Hospital, within the period from November 2017 – January 2018	2017	MD	

Discussion

Specialized SCD centers are important and must fulfill three goals; affording direct and quick access to the health service, presence of a multidisciplinary team providing the best quality medical care, and cost-effectiveness because of their proximity to and location in an area with significant SCD populations[16]. Specific clinics for children and adults with sickle cell disease are available in several African countries but only a few countries have newborn screening programs [17]. Sudan Sickle Cell Anemia Centre (SSCAC) is the only specialized center for SCD in Sudan for children and adults. Its location in Al Obeid in the West is Justified by the highest percentage of patients in Sudan being in the West region[6].

During the six years of its work, the number of registered patients increased from 112 to 1400 patients with the age range of 3 months to 34 years. The percentage of male patients is 56%. More than half of the registered patients were of the under 5 age groups. This is in concordance with a previous study done in Al-Obeid where the under five years age group was dominating[11].
It is observed that the percentage of patients visiting the clinic regularly and that of patients who present with crises and need to be hospitalized is very low compared to the total registered number. This can be attributed to the distance from the center, low socioeconomic status, or underreporting [18]. The center is treating both pediatric and adult patients but the number of patients visiting the pediatric clinic is around 100 – 160, while adults´ clinic is only about 20 – 30 per month. This is in agreement with the results of the recent review of cross-sectional population surveys and cohort studies of SCD in Africa, which estimated that between 50 and 90% of SCA children died before age 5 years [17]. In the late 1970s, studies in Africa reported a childhood survival of less than 2% in sickle cell disease [18]. With improvements in healthcare, this has increased to nearly 50% [19].

The WHO Regional Office for Africa has recommended the need for developing national SCD control programs that include advocacy, prevention and counseling, early detection, treatment, surveillance, research, and community education and partnerships [20]. SSCAC has conducted some training activities; courses and workshops were organized for doctors, nurses, and medical assistants. Limited activities were carried out due to a shortage of resources and lack of funding.

More efforts and funds are needed in training and education. Some studies recommend the training of community health workers to improve the outcome of SCD, especially with urban or rural populations. [21] In our Sudanese society, if they get the required training, they will be of great help in education, early referral for treatment, and reducing the load from medical staff. Although results are not yet available, the high rate of patient acceptance of community health workers is an early indicator that their interventions can be feasible [22].

Another important point in education is educating the patients and their families by using educational manuals or by interactive learning environment in a friendly atmosphere that might be an efficient way to present disease education to patients, families, and the community [23]. All these need extra funds so that it will be achieved acceptably and continuously.

The capacity for doing clinical research is an important feature of the specialized health facility. This will help in testing the possibility of implementation of SCD guidelines and assess their impact on patient outcomes[24].

Eighteen research studies are done in the center by postgraduates and medical students in different aspects of the disease. The research ranges from KAP studies to molecular studies on the disease haplotypes and correlations between phenotypes and single nucleotides polymorphism (SNPs).

Establishing a quality improvement process in resource-limited settings faces lots of challenges. Stepping forward for improved quality care requires critical self-assessment, the willingness to change, and determined commitment and contributions from staff, management, patients, community and government.

Recommendations

Sudan Sickle Cell Anemia Center is unique in Sudan in form of services provided to patients with SCD.

-The World Health Organization is recommended to provide technical and financial support to SSCAC in framing policies and strategies for the prevention and management of sickle-cell disease.

-The collaboration of the Sudan Ministry of Health, the WHO, and the non-governmental organizations with the center is needed to support constructing buildings, providing ambulance, the diagnostic equipment including Genetic screening, needed treatment facilities, transportation methods for patients from areas of Kordofan and Darfur to the center, and help funding research on sickle-cell disease to increase life expectancy and improve patient's quality of life.

-Funding resources for sickle-cell disease prevention and control, including training of staff to offer genetic counseling and health education, in addition to the introduction of newborn screening services in the center.

-Establishing similar specialized centers in Khartoum and different states of Sudan to provide easier access to services for most of the patients.

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