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THE KNOWLEDGE AND PARTICIPATION OF PHYSIOTHERAPISTS FROM THE NIGERIAN SOUTH-WEST RELATED TO THE CARE AND MANAGEMENT OF INDIVIDUALS WITH SICKLE CELL DISEASE

Chidozie E. Mbada, Overcomer T. Binuyo, Omotola A. Onigbinde, Samuel O. Ayelawa, Abiodun J. Olaniyi

Department of Medical Rehabilitation, Obafemi Awolowo University, Ile-Ife, Nigeria

Abstract

Rehabilitative modalities such as incentive spirometry and physiotherapy interventions, e.g. aerobic exercises, have been shown to produce positive outcomes in patients with sickle cell disease (SCD). Hemispheric CVA and other complications arising from SCD are amenable to physiotherapy. There have been few studies on the effectiveness of physiotherapy in the management of individuals with SCD. The present study attempts to assess the SCD-related knowledge, attitude and level of participation of physiotherapists in the Nigerian healthcare system. It also examines a possible association between the foregoing and practice settings. The study was carried out among physiotherapists in South-West Nigeria registered on professional e-platforms. The respondents filled in an adapted electronic version of a questionnaire. The IBM SPSS 20 was used for statistical analysis, and a chi-square test was used to determine an association between the levels of knowledge, practice settings, and participation among the respondents ($p < 0.05$). The study findings revealed that only 19% of respondents had received a specific training related to the care of SCD patients, although 65% had been involved in the treatment of at least one patient with SCD. Only 29.5% of respondents had "Good Knowledge" of SCD pathophysiology. A statistically significant association ($\chi 2 = 75.357$, $p = 0.012$) was found between respondents' level of knowledge and their practice settings, i.e. a teaching hospital mostly. The physiotherapists from the Nigerian South-West, mostly those working in teaching hospitals, reported an average level of knowledge of SCD care. Since the management of sickle cell disease is of multidisciplinary character and requires an all hands-on deck approach, controlled trials should be undertaken by physiotherapy researchers regarding the effects of modalities and interventions on pro-inflammatory bio-molecules.

Key words: *sickle cell disease, physiotherapy, rehabilitation*

Introduction

Sickle cell disease (SCD) is the most prevalent genetic disorder among the Black people in Nigeria [1] affecting an estimated 150,000 children annually [2]. SCD in fact comprises a group of autosomal recessive disorders resulting from homozygotic or compound heterozygotic inheritance of mutated genes encoding the production of β-globulin dimer of hemoglobin A (HbA). The replacement of glutamate on position 6 of the HBB gene located on the short arm of chromosome 11 with valine causes a mutation in the β-globulin subunit that distorts the shape of the haemoglobin which is then named HbS [3, 4]. Lysine replacing the glutamate gives rise to HbC [5], a less prevalent form of the abnormality. The heterozygous inheritance of the HbS gene with a normal HbA makes a person a carrier of the sickle cell gene in a condition called sickle cell trait [6]. The homozygous inheritance of the HbS gene precipitates sickle cell anaemia [6].

Heterozygous inheritance can occur with other abnormalities of the beta globin such as HbS/β0/+ thalassemia and the HbSC [6]. These occurrences lead to various crises and increased susceptibility to infections which are responsible for recurrent illness in patients with SCD. HbS carries oxygen effectively, but on

deoxygenation the β-globulin peptides polymerise forcing the red blood cells (RBC) into the crescentic shape of a sickle in the process called "sickling." This weakens the red blood cell membranes and ultimately leads to a rapid and excessive destruction of RBCs [6], a precipitate for anaemic crises experienced by individuals with SCD. Anaemic crises are caused by the hyperhaemolysis of RBCs, aplastic, and acute sequestration crises. Hyperhaemolysis is amplified by infections, glucose-6-phosphate dehydrogenase (G6PD) deficiency, acidosis and dehydration. The body responds by stimulating the production of reticulocytes from haematopoietic sources (long bones, skull and cheek bones), and extramedullary sources, mostly the liver. The work overload on these sources leads to the hypertrophy of bones featuring a "hair-on-end" appearance on diagnostic imaging and to hepatomegaly. The accumulation of by-products of the destruction of RBCs and unconjugated hemoglobin elicits sclera jaundice and painful bilirubin gallstones [7].

Aplastic crises are characterized by an acute failure of erythropoiesis often following viral infections, especially parvovirus B19. During these crises individuals experience fatigue and progressive pallor with associated bone and debilitating joint pain. Blood transfusions are often necessary to preserve the patient's life. Acute sequestration is caused by pooling of blood in the spleen and the liver, and is characterized by a sudden onset of progressive anaemia, splenic enlargement, abdominal pain and shock. In addition, aggregation of the sickled cells within blood vessels causes excruciatingly painful vaso-occlusive crises. Vaso-occlusive crises do not begin until about the 6th month of life [7] resulting from the abundance of HbF (foetal haemoglobin).

Their clinical presentation at infancy may include dactylitis, i.e. inflammation of the digits because of occlusion of blood vessels supplying the fingers. When vaso-occlusion occurs in vessels supplying the brain, a cerebral stroke ensues, while avascular necrosis arises due to loss of blood supply to the bones. Auto splenectomy following fibrosis of the spleen as a result of vessel blockage also complicates SCD. This exposes persons with SCD to infection from encapsulated bacteria such as Streptococcus pneumoniae, N. meningitidis, H. influenza etc. This infection causes or amplifies the acute chest syndrome in the presence of vaso-occlusion of pulmonary blood vessels, which in young children causes chest pain, fever, cough, tachypnoea, leucocytosis, and pulmonary infiltration in the upper lobes, often difficult to distinguish from pneumonias, whereas adults are usually afebrile, dyspnoeic with severe chest pain and multilobar/lower lobe disease. Pulmonary hypertension is being increasingly recognized as a serious complication of SCD. Vaso-occlusion in the kidneys results in necrosis of the renal papillae causing proteinuria and haematuria. Painful episodes of priapism also occur in men due to the occlusion of blood vessels in the penis.

The clinical rehabilitation of persons with SCD should employ a multidisciplinary approach of clinical practitioners, including physiotherapists [8].

Medical and pharmacological interventions involve oxygen and fluid replacement therapy to correct the dehydrated and hypoxic internal environment; opioids and NSAIDs to alleviate pain; antibiotics to treat infections; and blood transfusions and bone marrow transplants to replace reticulocytes and augment blood levels, respectively. Prophylactic hydroxyurea administration, penicillin and vaccination are approaches seeking to increase HbF levels and prevent bacterial infection in infants. Gene therapy and gene editing are the newest treatment techniques.

A number of studies indicate that physiotherapy may be highly beneficial in reducing the crises and hospital visits, as well as preventing complications by using individually tailored and monitored moderate intensity endurance exercises [9-11]. Aerobic dancing positively affects the platelet count, pack cell volume, health status, frequencies of crisis and hospitalization, and the length of hospitalization in individuals with sickle cell anaemia [12].

In the event of painful crises, heat modalities might be useful for pain management, but there is little or no evidence to support their direct benefits. Also, the application of TENS

has not been shown to display any potential benefits or harm in the management of pain in populations with SCD [8, 13].

Incentive spirometry performed regularly every 2 hours was shown to be beneficial in patients with chest pain, back pain, chest infection or hypoxia, reducing the risk of acute chest syndrome and atelectasis [14], and the role of physiotherapy in the early mobilisation of hospitalized patients for a relatively long time cannot be overlooked. Stroke developed secondary to SCD is amendable to physiotherapy in the restoration of function and mobility. Despite the evident relevance of physiotherapy in the management of persons with SCD, and the seeming unreadiness to engage physiotherapists in the management of SCD cases [15], there is an apparent paucity of data reflecting active physiotherapy interventions in managing persons with SCD in Nigeria. This study seeks to assess the knowledge, awareness and participation of physiotherapists in South-West Nigeria in the management of persons with SCD.

The very few studies into effectiveness of physiotherapy interventions in the management of individuals with SCD do indicate the need for research to assess the level of knowledge and awareness as well as attitude of physiotherapists to SCD in the Nigerian healthcare system.

The aims of the study were to assess:

- Nigerian physiotherapists' level of knowledge of SCD, its pathophysiology, and clinical presentation;
- Nigerian physiotherapists' awareness of physiotherapy interventions in individuals with SCD;
- the level of participation of Nigerian physiotherapists in the holistic management of individuals with SCD;
- the association between participants' level of knowledge and practice setting as well as their participation in the management of persons with SCD;
- the association between Nigerian physiotherapists' participation and practice setting in the management of persons with SCD.

The findings of this study were to reveal the current state of knowledge, awareness and participation of physiotherapists in Nigeria related to the care and management of individuals with sickle cell disease. The study results may affect the appropriation of physiotherapeutic practices in SCD management as a cogent aspect of a holistic approach to clinical rehabilitation of individuals with SCD.

Material and methods

This cross-sectional study using a convenient sampling technique was delimited to physiotherapists in South-West Nigeria registered on professional e-platforms of the Medical Rehabilitation Therapists Board of Nigeria, the Association of Academic and Clinical Physiotherapists of Nigeria, and Nigeria Society of Physiotherapy. Cochran's sample size formula for proportions was used to determine the study sample size.

A self-structured adapted electronic version of a questionnaire assessing the knowledge of, attitudes to, and awareness of Nigerian physiotherapy students of their roles in the prevention and management of COVID-19 by Mbada et al. [16] was used after having been tested for its face and content validity.

The IBM SPSS 20 was used to calculate data using descriptive statistics and chi-square inferential statistics and to determine the association between respondents' levels of knowledge, practice settings, and participation ($p < 0.05$).

Results

The physiotherapists from South-West Nigeria reported a 51.33% response rate. The respondents' mean age was 29.44 ± 6.94 (minimum age: 20 years, maximum age: 50 years). The majority of respondents were men (52.6%), single (70.5%), and with the first academic degree (79.5%) as their highest qualification. Most respondents were either interns or members of the youth corps (46.2%), with 10 years or less experience (65.4%), who worked in teaching hospitals (33.3%).

Table 1. Respondents' socio-demographics

Variable	Frequency	Percentage
Sex		
Male	41	52.6
Female	37	47.4
Marital Status		
Single	55	70.5
Married	23	29.5
Highest Educational Qualification		
BSc/BMR/BPT	140	79.5
MSc	15	19.2
PhD	1	1.3
Years of Experience		
1 – 10	51	65.4
11-20	10	12.8
Less than 1 year	17	21.8
Place of Work		
Federal Medical Centre	9	11.5
Private Practice	16	20.5
Specialist Hospital	7	9.0
State Hospital	15	19.2
Teaching Hospital	26	33.3
University	5	6.4

Table 2. Assessment of respondents' knowledge of SCD

No.	Item	Percentage of respondents who gave the correct answer n (%)
1.	SCD is a genetic disorder of red blood cells.	75 (96.2%)
2.	Being a carrier means you are anaemic.	46 (59.0%)
3.	SCD is caused by inheritance of a sickle allele from both parents.	76 (97.4%)
4.	SCD occurs when glutamic acid replaces valine in the heaemoglobin protein.	60 (76.9%)
5.	SCD is diagnosed at birth.	49 (62.8%)
6.	SCD symptoms occur at birth.	38 (48.7%)
7.	Pain is a common symptom in people living with SCD.	76 (97.4%)
8.	The most experienced form of pain is stabbing.	54 (69.2%)
9.	Swelling on both hands may occur.	28 (35.9%)
10.	Swelling on both feet may occur.	31 (39.7%)
11.	Jaundice may occur.	62 (79.5%)
12.	Easy and frequent fatigability.	73 (93.6%)
13.	Pre-marriage genotype counselling helps to reduce SCD prevalence.	77 (98.7%)
14.	Persons with SCD are susceptible to infantile stroke.	67 (85.9%)
15.	Persons with SCD commonly suffer from ischemic complications at the hip.	75 (96.2%)
16.	Persons with SCD suffer from chronic respiratory distress syndrome.	59 (75.6%)
17.	Persons with SCD may experience joint and low back pain.	75 (96.2%)
18.	Persons with SCD may experience chest and abdominal pain.	60 (76.9%)

Table 3. Respondents' levels of knowledge of SCD

	Frequency	Percent
SCD knowledge among respondents		
Good	23	29.49
Fair	55	70.51

Although 65% of respondents had been involved in the treatment of at least one patient with sickle cell disease before, only 19% had received specific SCD training on prevention measures and therapeutic interventions, and 21% received training on collaborative activities in SCD management. The assessment of respondents' knowledge on SCD is presented in Table 3. 96.2% correctly responded that SCD is a disorder of red blood cells caused by inheritance of a sickle allele from both parents; however, only 46 (59.0%) responded correctly to the fact that being a carrier does not mean being anaemic. 60 (76.9%) responded correctly regarding the biochemical basis of the replacement of glutamate with valine in the hemoglobin protein. Most respondents (98.7%) agreed that pre-marital counselling helped to reduce SCD prevalence, and 75 (96.2%) knew that SCD patients commonly suffer from ischemic complications. While most respondents had a fair knowledge of SCD, only 23 (29.5%) had a good knowledge of SCD. Most respondents (69.2%) had participated in the care of individuals with SCD and in specially prepared preventive programs.

There was a significant association ($\chi 2$ = 75.357, p = 0.012) between respondents' knowledge levels and their practice settings, as most of them worked in a teaching hospital. However, no significant association was found between respondents' level of knowledge and participation (Table 5). Moreover, the association between respondents' participation levels and practice setting was non-significant (Table 6).

Table 4. Participation of physiotherapists in SCD patient care

Item	Yes	No
I have taken part in the care of SCD patients.	69.2%	30.8
I have cared for a SCD stroke patient.	39.7	60.3
I have provided chest physiotherapy to a SCD patient in ICU.	37.2	61.5
I have prepared preventative programs for persons with SCD.	69.2	30.8
I have employed pain relief modalities in SCD patients.	48.7	51.3
I have treated low back pain in an SCD patient.	21.8	78.2
I have treated a leg/foot ulcer in a SCD patient.		100%

Table 5. Association between respondents' level of SCD knowledge level and practice settings

	Level of SCD knowledge	
	$\chi 2$	p-value
Practice setting	75.357	0.012
Participation		
I have taken part in the care of SCD patients.	10.436	0.403
I have cared for a SCD stroke patient.	7.407	0.687
I have provided chest physiotherapy to a SCD patient in ICU.	20.078	0.453
I have prepared preventative programs for persons with SCD.	7.391	0.688
I have employed pain relief modalities in SCD patients.	11.133	0.347
I have treated low back pain in an SCD patient.	8.537	0.577
I have treated a leg/foot ulcer in a SCD patient.	4.948	0.895

Table 6. Association between respondents' practice settings and participation in SCD management

Item	Respondents' practice settings	
	χ^2	p-value
I have taken part in the care of SCD patients.	7.027	0.219
I have cared for a SCD stroke patient.	3.955	0.556
I have provided chest physiotherapy to a SCD patient in ICU.	15.129	0.127
I have prepared preventative programs for persons with SCD.	3.566	0.613
I have employed pain relief modalities in SCD patients.	3.902	0.564
I have treated low back pain in an SCD patient.	7.964	0.158
I have treated a leg/foot ulcer in a SCD patient.	5.915	0.315

Discussion

SCD survivors experience periodic painful crises which leave them physically exhausted, with an indication for possible physical therapy interventions. The aim of this study was to assess the knowledge, awareness as well as participation of physiotherapists from South-West Nigeria in the management of persons with sickle cell disease (SCD). The study also evaluated the association between physiotherapists' participation in the management of persons with SCD, practice settings and their levels of knowledge.

The highest qualification of most respondents was a bachelor's degree with a professional experience between 1 and 10 years, which corresponded with the results of most other surveys carried out in Nigeria [17]. Most of the respondents worked in a teaching hospital. As expected the vast majority (96.2%) of them understood that SCD is a genetic disorder of red blood cells; however, more than one half of the respondents could not differentiate between being a carrier and being anemic. With regard to SCD pathophysiology, one-third of respondents know that SCD occurs because of a replacement of the amino-acid glutamate with valine in the haemoglobin protein. Most respondents also know that SCD is caused by inheriting a sickle allele from both parents.

SCD diagnosis usually occurs between 3 and 6 months after birth when foetal haemoglobin becomes almost completely replaced by adult haemoglobin, with symptoms initially appearing between 5 and 6 months of age [7]. A little more than one-half of the respondents know that SCD is not diagnosed at birth, but less than 50% know that SCD symptoms do not begin at birth.

Pain is the most common SCD symptom and complication [18] as well as the topmost reason for hospitalization of individuals with SCD [19]. Stabbing, aching, or throbbing (20) pain is usually felt in the chest, arms, legs, fingers, and toes. Pain is a significant cause of disease-related morbidity and is negatively associated with quality of life of persons living with SCD [21]. Efforts to reduce the debilitating effect of pain in SCD patients are paramount and have been largely pharmacological. Evidence of the effectiveness of physiotherapy modalities in managing pain is marginal; however, kinesiotherapy and aquatic rehabilitation have been shown to reduce pain episodes and the likelihood of pulmonary complications [22].

Most respondents know SCD patients may experience joint, low back, chest and abdominal pain. Swelling usually occurs in places where pain is felt in the hands and feet because of recruitment of vasodilators during the inflammation process in vaso-occlusive crises [23]. Nearly 40% of the respondents know that swelling occurs both in the hands and feet of SCD patients. Because of easy and frequent fatigability experienced by SCD survivors physiotherapy interventions are usually mild, with frequent breaks, and gradually raised to moderate levels. Most respondents did know that SCD patients experience frequent fatigability. Beside pain, other SCD complications include jaundice, cerebrovascular diseases, ischaemic complications of the hip leading to avascular necrosis of the femoral head, and chronic respiratory distress syndrome, all of which are implications for physiotherapy. Most respondents knew that SCD patients experience these symptoms. Premarital counselling has been shown to reduce SCD prevalence [24]. All respondents but one knew that pre-marriage genotype counselling helps to reduce the SCD prevalence. Overall, a fair level of knowledge about SCD pathophysiology, diagnosis and symptoms was reported by most of the respondents.

Although the management of persons with SCD is usually multidisciplinary, more oft than not, physiotherapeutic interventions are usually limited to awareness, health promotion and preventative programs, in which one-third of respondents have been involved in. About one-third of the respondents have participated in the care of individuals with SCD, but nearly one half had managed stroke, low back pain or chest pathologies. Sickle-cell leg ulcers are usually severe, chronic, and recurrent with no official treatment recommendations [25] and with no literature data confirming amenability to physiotherapy. As such no respondents had treated leg/foot ulcers in SCD patients.

The present study revealed a statistically significant association between respondents' practice setting (mostly a teaching hospital) and level of knowledge of SCD. However, no significant associations were found between the physiotherapists' level of knowledge and participation, or between practice settings and participation in the management of SCD patients.

Conclusions

Physiotherapists from South-West Nigeria, mostly those working in teaching hospitals, demonstrate an average level of knowledge of sickle cell disease. Since SCD management is multidisciplinary and requires an all-hands-on-deck approach, controlled trials should be undertaken by physiotherapy researchers regarding the effects of modalities and interventions on the pro-inflammatory bio-molecules.

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CORRESPONDENCE

Chidozie E. Mbada
Department of Medical Rehabilitation, Obafemi Awolowo University
Ile-Ife, Nigeria
E-mail: doziembada@yahoo.com