Original Article

Preventive Measures of Vaso-Occlusive Crisis Among Sickle Cell Disease Patients in South-Eastern Nigeria: How Much Do Our Patients Know?

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Abstract

Background: Vaso-occlusive crisis (VOC) is a feature of sickle cell disease (SCD), an inherited medical condition which is more common in Sub-Saharan Africa especially Nigeria and is saddled with multi-systemic complications. Knowledge and practice of preventive measures of VOC by patients may reduce the risk of morbidity and mortality. **Aim:** The purpose of this manuscript is to determine the knowledge, attitude, and practice of the various preventive measures of VOC by patients and their relationship with their frequency of painful crisis. **Materials and Methods:** A cross-sectional descriptive survey was used. This study was conducted between August, 2018, and February, 2019. Participants were selected consecutively as they came for clinic consultations. Demographic characteristics and the knowledge, attitude, and practice of various preventive measures of VOC were obtained using a structured interviewer-administered questionnaire. The analysis was performed using Statistical Package for the Social Sciences software version 23.0. Descriptive and inferential statistics were used with $P < 0.05$ which was considered statistically significant. Ethical clearance and informed consent were obtained before the commencement of the study. **Results:** A total of 154 SCD patients participated in the study and were made up of 73 (47.4%) males and 81 (52.6%) females, with a male-to-female ratio of 1: 1.1. Majority of the respondents (76.6%) had good knowledge of preventive measures toward VOC in SCD. This study also found that 59 (38.3%) participants had good practice of preventive measures toward VOC while 95 (61.7%) had fair practice. Majority (68% [40/59]) of those who had good practice of preventive measures were less likely to have more than three crises in a year compared to those who do not practice good preventive measures, though not statistically significant (OR = 2.489, $P = 0558$) (confidence interval = 0.480 – 0.637). **Conclusion:** Majority of the patients have good knowledge and practice of preventive measures of VOC and this appears to have reduced the frequency of their crisis in a year. Health education for sickle cell patients is an important tool that may reduce morbidity and mortality by reducing the frequency of crisis.

Keywords: Painful crisis, preventive measures, sickle cell anaemia, south-east Nigeria, vaso-occlusive crisis

Introduction

Sickle cell disease (SCD) is an inherited disorder of the red blood cells in which the abnormal variant of hemoglobin S (HbS) is found in the place of the normal hemoglobin A.[1] This abnormality was discovered over a 100 years ago.[1] At the sixth position in the beta globin gene, glutamic acid was replaced with valine,[2] leading to polymerization of the abnormal HbS during periods of deoxygenation which accounts for increased rigidity of the red cells.[3] This presents with various symptoms ranging from dactylitis, acute and repeated painful crisis, priapism, acute chest syndrome, and severe anaemia from chronic hemolysis.[4]

Over time, these signs and symptoms result in severe complications that often lead to significant morbidity, chronic organ damage, and shortened life expectancy when compared

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to the general population.[5,6] These complications include avascular necrosis of the bones, cerebrovascular accidents, renal failure, chronic leg ulcers, and pulmonary hypertension. SCD is a broad term for all hemoglobinopathies with abnormal HbS. These include sickle cell anaemia (HbSS), hemoglobin SC disease, and other compound hemoglobinopathies such as sickle beta-thalassemia.[7] The most common and most severe form is the HbSS[6] which is found in sub-saharan Africa [3]and among people of African descent living in other continents. It affects about 2% to 3% of the Nigerian population of more than 160 million.[8]

The clinical severity of SCD is dependent on both genetic and environmental factors.[9,10] There are factors that have been known to exacerbate the severity of sickle cell vaso-occlusive crises. These include high cell counts such as hemoglobin level of 11 g/dl and above, high white cell counts and neutrophilia, thrombocytosis, low fetal hemoglobin (HbF) level, and the haplotype.[11]

Studies have shown that painful crisis in SCD is worsened by physical[12] and emotional stress,[13] as well as some environmental factors such as quality of air, climate, exercise, socioeconomic status, and infection.[14] Association of cold weather and frequency of painful crisis has also been documented.[15]

Preventive measures for repeated crises are often taught by doctors and counselors. The sufferers are encouraged to rehydrate, avoid smoking, and avoid exposure to extremes of temperature and physical as well as emotional stress.

In general, in outpatient clinics, we come across a number of these patients, of which the new ones are unaware of these preventive measures and therefore do not practice them. They also seek help in the wrong places due to poverty and ignorance. This may lead to worsening of symptoms and complications and increased likelihood for increased morbidity and mortality. The cost of hospital care for SCD patients is high and far-fetched for majority of these patients.[16] This is also complicated by lack of comprehensive health-care insurance scheme. Most of the new interventions such as gene therapy and stem cell transplantation are either very expensive or inaccessible.

Awareness and practice of these preventive measures may be beneficial in reducing the frequency of Vaso-occlusive crisis (VOC) in some of these patients and therefore reducing the risks of mortality.

There is a dearth of information on how much patients know about the risk factors for VOC and the available preventive measures. The purpose of this manuscript is to determine the knowledge, attitude, and practice of the various preventive measures of VOC by the patients and their relationship with their frequency of painful crisis. Findings of the study may aid in making health policies that will improve the quality of life of patients living with SCD as well as reduce morbidity and mortality from frequent VOC in SCD patients in resource-poor environment like ours.

Materials And Methods

Study area and study population

A multicenter, descriptive, cross-sectional study was carried out between August 2018 and February 2019 at the Federal Teaching Hospital Abakaliki, Ebonyi State; Emeka Odumegwu Ojukwu University Teaching Hospital, Awka, Anambra State and University of Nigeria Teaching Hospital (UNTH), Ituku/Ozalla, Enugu State, all in southeastern Nigeria. The study population comprised adult SCD patients seen at the hematology clinics of the health institutions where the studies were carried out.

Sampling method

Convenient sampling technique was used. Patients were consecutively recruited as they walked into the consulting rooms. Only those patients who gave an informed verbal consent were recruited for this study.

Study instrument and data collection

A pretested, semi-structured, interviewer-administered questionnaire was used for data collection. A total of 300 questionnaires were distributed, out of which only 154 questionnaires were correctly and completely filled. The response rate was 51.3%. These questionnaires were considered valid, study data were extracted from them, and data analysis was done.

Information sought in the questionnaire included sociodemographic characteristics, knowledge about preventive measures against VOC in SCD, practice of preventive measures against VOC, and attitude toward SCD.

Data management and analysis

Knowledge about sickle cell disease and practice of preventive measures against vaso-occlusive crisis

Ten variables on the study instrument were used to assess participants' knowledge on the preventive measures of VOC. One mark was awarded for every correctly answered question and zero for every wrongly answered or unanswered question. Getting all the ten questions on general knowledge of SCD correctly were scored as 100%. Those who scored 80% and above had good knowledge, whereas those that scored 51%–79% had fair knowledge. Participants who scored 50% and below were categorized as having inadequate knowledge.

Attitude of people living with sickle cell disease

Seven questions on the study instrument were used to assess respondents' attitude toward SCD, with each question having five Likert items. The Likert items were strongly agree, agree, indifferent, disagree, and strongly disagree. The number of responses to each of the Likert items was graded as 1 for strongly disagree, 2 for disagree, 3 for indifferent, 4 for agree, and 5 for strongly agree, giving a total minimum score of 7 and maximum score of 35.

The total attitude scores were obtained and converted to percentages and graded as negative attitude (scores 49.9% and below) and positive attitude (scores 50% and above).

Data analysis

Data collected were cleaned for inconsistencies in the responses, were coded, and analyzed using statistical package for social sciences (SPSS) computer software version 23 (SPSS Inc. Chicago, IL, USA). Descriptive statistics was used to compute percentages and averages. Chi-square and Fisher's test were used to test for associations between variables as appropriate. $P < 0.05$ ($P < 0.05$) was considered significant. The results were presented in tables and charts and expressed as percentages/proportions, means, and standard deviation.

Ethical considerations

Ethical clearance for this study was obtained from the UNTH Health Research Ethics Committee.

Results

A total of 154 SCD patients participated in the study and were made up of 73 (47.4%) males and 81 (52.6%) females, with a male-to-female ratio of 1: 1.1. The mean age of the participants was 23.9 ± 7.2 years. Most of the participants, 141 (91.6%), were single [Table 1].

One hundred and eighteen (76.6%) participants had good knowledge about preventive measures toward VOC in SCD, while 36 (23.4%) had fair knowledge. None of the participants had poor knowledge. Knowledge about preventive measures toward VOC displayed by the participants are shown in Table 2.

This study also showed that majority (131 [85.1%]) of the participants had a positive attitude toward sickle cell disease [Figure 1], as 130 (84.4%) agreed that they have accepted their medical condition, 151 (98.1%) rejected the notion that their parents' hatred for them was the cause of the sickle cell anaemia [Table 3].

This study also found that 59 (38.3%) participants had good practice of preventive measures toward vaso-occlusive crises while 95 (61.7%) had fair practice.

Relationship between practice and frequency of crises

This study showed that majority, 68% (40/59), of those who had good practice of preventive measures were less likely to have more than three crises in a year compared to those who do not practice good preventive measures, though not statistically

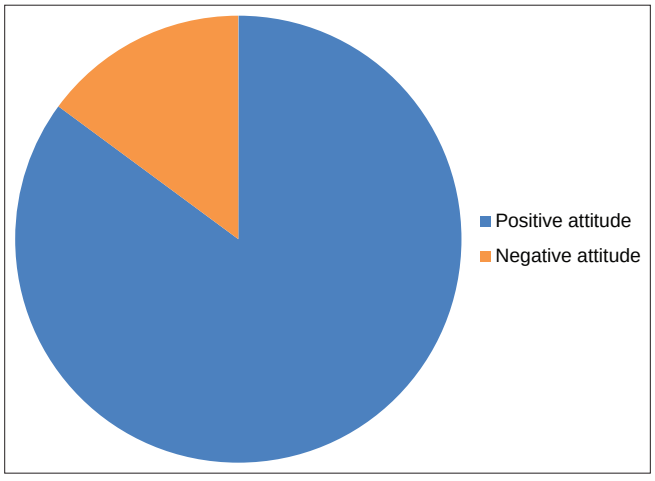

Figure 1: Attitude displayed by the participants to sickle cell disease

Table 1: Sociodemographic characteristics of the participants

Characteristics	Frequency (%)
Gender	
Male	73 (47.4)
Female	81 (52.6)
Total	154 (100)
Educational status	
Primary	20 (13.0)
Secondary	44 (28.6)
Tertiary	90 (58.4)
Total	154 (100)
Marital status	
Single	141 (91.6)
Married	12 (7.8)
Separated	1 (0.6)
Total	154 (100)
Financial status	
Comfortable	44 (28.6)
Fair	57 (37.0)
Not enough	53 (34.4)
Total	154 (100)
Occupation	
Civil servant	16 (10.4)
Student	95 (61.7)
Self-employed	25 (16.2)
Unemployed	18 (11.7)
Total	154 (100)

Table 2: Participants knowledge about preventive measures toward vaso-occlusive crisis

Variables	Responses	
	Correct, *n* (%)	Incorrect, *n* (%)
Drinking enough fluid	136 (88.3)	18 (11.7)
Avoiding extremes of temperature	126 (81.8)	28 (18.2)
Daily intake of folic acid	129 (83.8)	25 (16.2)
Daily intake of Paludrine	105 (68.2)	49 (31.8)
Daily intake of iron	122 (77.3)	35 (22.7)
Regular hospital visits on doctor's appointment	128 (83.1)	26 (16.9)
Avoid mosquito bites	145 (94.2)	9 (5.8)
Minimizing stress of all types	129 (83.8)	25 (16.2)
Avoiding strenuous physical activities	132 (85.7)	22 (14.3)
Sleeping under insecticide-treated nets	126 (81.8)	28 (18.2)
Getting pneumococcal vaccines	51 (33.1)	103 (66.9)
Regular attendance to spiritual homes	124 (80.5)	30 (19.5)
Reporting to the emergency at the onset of severe crisis	125 (81.2)	29 (18.8)
Knowing your steady state hemoglobin	88 (57.1)	66 (42.9)
Smoking tobacco	152 (98.7)	2 (1.3)
Drinking alcohol	148 (96.1)	6 (3.9)
Regular light exercises	91 (59.1)	63 (40.9)

Table 3: Attitude toward sickle cell disease

Attitudinal statements	Attitude	Responses
	Positive, *n* (%)	Negative, *n* (%)
I am a sickle cell patient because my parents hate me	151 (98.1)	3 (1.9)
It is a spiritual illness from ancestral spirit	147 (95.5)	7 (4.5)
I have accepted my medical state	130 (84.4)	24 (15.6)
I strive to be the best I can	148 (96.1)	6 (3.9)
I feel hopeless, scared and alone	135 (87.7)	19 (12.3)
I have good support from family and friends	145 (94.2)	9 (5.8)
I have given up in life	141 (91.6)	13 (8.4)
I know I can live long with SCD if I live healthy	145 (94.2)	9 (12.3)
Being part of support group is beneficial	130 (84.4)	24 (15.6)

SCD: Sickle cell disease

significant (odds ratio = 2.489, *P* = 0558) (confidence interval = 0.480 – 0.637).

Discussion

Health education on the preventive measures of VOC is necessary due to the high rate of poverty and low threshold to seeking medical care in our environment. SCD poses a major scourge in developing countries like Nigeria due to poor socioeconomic status, nonavailability of new therapies, ignorance, and lack of medical facilities. These lead to increased morbidity and mortality in the patients, with its attendant physical and emotional stress. Considering the fact that there are known triggers for vasocclusive crisis, it may be beneficial to ensure that patients understand these triggers and ensure they avoid them. This may help reduce morbidity among the patients, given the poor socioeconomic status and poor health-care delivery.

We studied 154 sickle cell patients that seek care from three different tertiary health-care institutions. We found that majority of these patients were aware of the preventive measures. This is a very encouraging number and it also shows that health workers seen by these patients educate them on healthy living. There is generally a scanty literature on previous studies on this topic, but our findings are contrary to the findings of a study done in Saudi by Al Nasir and Niazi[17] and another in Bahrain[18] that showed a poor knowledge of preventive measures in their patients. These studies were done over 25 years ago when there was still poor understanding of the triggers for VOC and poor internet access. There is better understanding of SCD as years evolved, patients are more aware now through access to the internet and can source information on their own. It is essential that patients understand the nature of their illness to enable them seek medical care early enough. They also need to be aware that frequent painful crisis may lead to increased morbidity and mortality.

Most of our patients had a good attitude toward their medical condition. Over 80% accepted their medical condition and did not think their medical condition was due to superstitious beliefs. Gil *et al*. stated that the acquisition of knowledge about a disease is important in behavioral adaptation, especially if it is paired with belief that one's behavior will have positive impact on health.[19] This statement might explain the positive attitude the patients have because of the good knowledge they have on the subject matter.

While it is good to have a good knowledge of these preventive measures, it is more important that the patients practice them. We reported that over 70% of the patients had a good knowledge of these preventive measures, but only about a third of them actually practice these measures. Reasons given by some patients while being seen at the clinic for this were distractions by daily activities such as school or work. Some also complained of not having a convenient place to ease themselves as their reasons for not being well hydrated. Others may not be able to get sufficient rest because they are dependent on their daily income for survival and hence the tendency to forget to rest. These patients require continuous counseling to enable them modify their life style so as to incorporate these preventive measures.

We also reported that those who had a good practice of these preventive measures were less likely to have recurrent VOC, although the finding was not statistically significant. This is contrary to a finding in a study by Amoran *et al*., carried out in Abeokuta, Nigeria,[6] that reported increasing painful crisis despite adequate knowledge of the preventive measures. It appears that the problems patients may have is with practicing the knowledge they already have, which may explain the findings by Amoran *et al*.[6] It is therefore important to engage the patients more through counseling by medical personnel, to encourage them practice what they already know so as to help reduce the frequency of VOC with the possibility of reducing mortality.

Conclusion

The study found that the patients had adequate knowledge and good attitude toward the preventive measures of SCD but practice of this knowledge was poor and might negatively impact their quality of life. Enacting policies with regards to knowledge and practice of these preventing measures would be a viable and cost effective way of managing sickle cell anaemia VOC. This can be achieved via counseling the patients, to confirm they have the correct information and understand their challenges in practicing these preventive measures. It is hoped that this will help reduce frequency of painful crisis and possibly, the attendant complications.

Recommendations

Based on the findings of this study, regular educational and enlightenment activities or programs should be organized to improve the practice of use of these preventive measures to reduce the frequency of vaso-occlusive crises in sickle cell anaemia patients.

Limitations of the study

Recall bias may have affected the ability of the patients to give details of what they do to prevent crisis.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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