Knowledge and Health Beliefs of Caregivers toward the Complications and Triggers of Sickle Cell Disease at-Dr Gaafar Ibnauf Paediatric Hospital, (2022)

Fatma Abdalla Mohamed[1] ,Fathelrahman Elawad Ahmed[2] ,Egbal Abbashar Algmair [3]

1. BSc, MSc Lecturer, Department of Nursing, College of Nursing and Health Sciences, Jazan University, Kingdom of Saudi Arabia. MBBS, MSc, PhD Professor of pediatrics and consultant pediatric hematologist, Dr. Gaafar Ibnauf Paediatric Hospital- Sudan

2. BSc, MSc, PhD Associate professor in pediatric Nursing, faculty of nursing science, University of Alnelain-Sudan

Corresponding author: Fatma Abdalla Mohamed; Lecturer, Department of Nursing, College of Nursing and Health Sciences, Jazan University, Kingdom of Saudi Arabia. Tel: +966536773859; Email: fatimaabdalla008@gmail.com

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Abstract:

Introduction: Sickle cell disease (SCD) is a potentially devastating condition that results in vaso-occlusive and hemolytic crisis. It is complicated by various life-threatening events. The severity of complications associated with this disorder is widely variable. The overall patient mortality is much higher, and life expectancy is shorter when compared to the general population. The study aimed to assess the knowledge and health beliefs of caregivers toward the complications and triggers of sickle cell disease.

Methods: A descriptive cross-sectional study design was conducted in the Sickle cell referral clinic, Dr. Gaafar Ibnauf Paediatric Hospital, Khartoum, Sudan, with a simple random sampling of 286 caregivers who attended the referral clinic during the study period between September and December 2021. Data were collected using a pre-structured investigator-filled questionnaire. The Likert scale was used to assess the health beliefs of caregivers toward the complication and triggers of sickle cell disease. A parametric one-sample T-test was used to investigate the statistical significance of the knowledge and health beliefs of caregivers toward the complications and triggers of sickle cell disease.

Results: The vast majority of participants were female (85.7%). Most of them were from the second class, followed by those from the third class. Almost all of the participants were not employed; the rest were workers. Most of the caregivers' educational level was intermediate, followed by secondary level. The study findings indicate the level of knowledge of caregivers about the complication of SCD was unsatisfactory. In contrast, the knowledge about the triggers of SCD and the health beliefs of caregivers toward the complication and triggers of SCD were satisfactory.

Keywords: Knowledge, Health Beliefs, Caregivers, Complications, Triggers, Sickle cell disease.

المستخلص

المقدمة: مرض فقر الدم المنجلي (SCD) هو حالة مدمرة محتملة تؤدي إلى نوبات فقر الدم الانحلالي وانسداد الأوعية الدموية. كما انه يصبح صعبا ومعقدا بسبب الأحداث المختلفة التي تهدد الحياة. تختلف شدة المضاعفات المرتبطة بهذا الاضطراب على نطاق واسع. معدل الوفيات الإجمالي للمرضى أعلى بكثير ومتوسط العمر المتوقع أقصر بالمقارنة مع عامة السكان. هدفت الدراسة إلى تقييم المعرفة والمعتقدات الصحية لمقدمي الرعاية تجاه مضاعفات ومسببات مرض فقر الدم المنجلي.

منهجية الدراسة: تم إجراء دراسة مقطعية وصفية في العيادة الخارجية لفقر الدم المنجلي التابعة لمستشفى الدكتور جعفر بن عوف التخصصي للأطفال، الخرطوم، السودان. من خلال أخذ عينة عشوائية بسيطة، حيث تم اختيار ٢٨٦ من مقدمي الرعاية للأطفال المصابين بفقر الدم المنجلي والذين كانوا على متابعة منتظمة وحضروا العيادة خلال فترة الدراسة ما بين سبتمبر وديسمبر ٢٠٢١. تم جمع البيانات عن طريق الاستبانة المنظمة مسبقا. كما تم استخدام مقياس ليكرت لتقييم المعتقدات الصحية لمقدمي الرعاية تجاه مضاعفات ومحفزات مرض فقر الدم المنجلي. تم استخدام اختبار (T) لعينة واحدة للتحقيق في الدلالة الإحصائية للمعرفة والمعتقدات الصحية لمقدمي الرعاية تجاه مضاعفات ومحفزات مرض فقر الدم المنجلي.

نتائج الدراسة: كانت الغالبية العظمى من المشاركين من الإناث (٨٥.٧٪). كان معظمهم من الدرجة الثانية، يليهم أولئك من الدرجة الثالثة. وأغلبية المشاركين في الدراسة تقريبا لا يعملون، أما الباقون فكانوا عمالا. كان المستوى التعليمي لمعظم مقدمي الرعاية متوسطا، يليه المستوى الثانوي. تشير نتائج الدراسة إلى أن مستوى معرفة مقدمي الرعاية بمضاعفات مرض فقر الدم المنجلي غير كافية. في المقابل، كانت المعرفة بمحفزات مرض فقر الدم المنجلي والمعتقدات الصحية لمقدمي الرعاية تجاه مضاعفات ومحفزات مرض فقر الدم المنجلي كافية.

الكلمات المفتاحية: المعرفة، المعتقدات الصحية، مقدمو الرعاية، المضاعفات، المحفزات، مرض فقر الدم المنجلي.

Introduction:

Sickle cell disease (SCD) is a potentially devastating condition that results in vaso-occlusive and hemolytic crisis. It is complicated by various life-threatening events. The severities of complications associated with this disorder are widely variable. The overall patient mortality is much higher and life expectancy is shorter when compared to the general population [1].

One of these life-threatening events is splenic sequestration which leads to a sudden and severe decline in the hemoglobin level. In a plastic crisis, the parvovirus B19 infects the precursors of the red blood cells in the bone marrow, causing transient RBC aplasia with reticulocytopenia. Vaso-occlusive pain can occur in any organ of the body and can be manifested by either significant dysfunction or pain [2].

A new pulmonary infiltrate in children with SCD is known as acute chest syndrome [1]. neurologic complications that can occur in children with SCD are varied and complex, about 11% and 20% will have either overt or silent strokes, respectively, before they reach their 18th years. One of the most common complications that can occur in children with SCD is priapism. The highest prevalence of priapism in males was found in 48.0% of males [3]

Research questions:

1-What do caregivers know about complications of sickle cell disease?

2- What do caregivers know about triggers of sickle cell crisis?

3-What do caregivers believe in the complications and triggers of sickle cell disease?

4-What kinds of sources of information are available to caregivers about sickle cell disease?

Significance of the study:

Sickle cell disease (SCD) is a life-threatening genetic disorder affecting approximately 100,000 individuals in the United States and is associated with many acute and chronic complications that require immediate medical attention [4].

The lack of standardized diagnostic criteria for the complications of SCD is confusing to the patients, their families, the public, and healthcare providers. It has hindered the efforts of clinical researchers to collect outcome data and improve the quality of care for those with this condition [5].

A previous study was conducted to empower mothers to overcome the sickle cell crisis in their children in the maternity and children hospital at Makkah Al-Mukarama. The study results revealed poor knowledge about complications of sickle cell anemia in 57.5% of the mothers. The study also showed that most mothers had poor knowledge about various triggers of SCD, such as fever, infection, dehydration, pain, and nutritional deficiency [6]. Based on the findings of this study and the work experiences, it was expected that the lack of knowledge about complications and various triggers of SCD would be presented in Sudanese mothers. This study aims to analyze caregivers' knowledge and health beliefs toward the complications and triggers of sickle cell disease.

The study objectives:

The study aimed to assess the knowledge and health beliefs of caregivers toward the complications and triggers of sickle cell disease.

Methods

This was a descriptive cross-sectional study conducted, at Dr. Gaafar Ibnauf Paediatric Hospital, Khartoum, Sudan. This hospital was selected because it is the largest pediatric hospital in Sudan. Being located in Khartoum, the referral clinic provides services to patients coming from all parts of Sudan.

Approximately, 50 patients are seen weekly in a regular follow-up every 1-2 months.

The sample consisted of 286 out of 1000 caregivers of children with SCD who were on regular follow-up and attended the clinic during the period between September and December 2021. Caregivers of patients with other chronic diseases besides SCD were excluded from the study. By simple random sampling, research subjects were assigned to groups completely at random. Each subject was labeled then a table of random numbers was used to select from labeled subjects. Interviews were used to collect the data. Data were collected by the researcher using a pre-structured investigator-filled questionnaire. A structured questionnaire was developed to assess socio-demographic data and socioeconomic status based on WHO classification, it included basic knowledge regarding the triggers of crisis such as high altitude, fever, infection in crowded places too much exposure to the sun, furthermore possible complications of SCD, for instance, splenic sequestration crisis, acute chest syndrome, stroke, retinopathy, osteomyelitis, and vascular necrosis.

The Likert scale was used to assess the health beliefs of caregivers toward the triggers and complications of sickle cell disease.

The score of knowledge was computed on a binary basis with 1 point for a correct answer and 0 points for an incorrect response. While the score of health beliefs was sum on a five-point scale with 1 point graded for a strongly disagree statement and 5 points for a strongly agree statement, The hypothetical mean (test value) of both knowledge and health beliefs was assumed to be equal to the mean of the knowledge and health beliefs score separately.

Ethical approval was obtained from the Institutional Review Board of Alnelain University and the Ethics Committee of the Ministry of Health, Khartoum State. The researcher maintained the anonymity and confidentiality of the participants after informed consent was obtained. The researcher collected the data during the rest time of the study groups. The researcher maintained the preventive measures against COVID-19.

The statistical analysis was performed by using Statistical Packages for Social Sciences (SPSS) version 25 Parametric one-sample T-test was used to investigate the statistical significance of the variable of interest.

Results

The data were grouped and analyzed to assess the knowledge and health beliefs of caregivers toward the complications and triggers of sickle cell disease.

Table (1): Socio-demographic characteristics of the studied group

Socio-demographic characteristics				
	N	**Mean**	**Minimum**	**Maximum**
Age (years)	286	34.6	17.00	65.00
		N	**Frequency**	**Percent**
Gender	Male	286	41	14.3
	Female	286	245	85.7
Occupation	Not employed	286	202	70.6
	Workers	286	39	13.6
	Professional	286	8	2.8
	Self employed	286	25	8.7
	Student	286	12	4.2
Resident area	Slum	286	7	2.4
	Third class	286	25	8.7
	Second class	286	248	86.7
	First class	286	6	2.1

The data depicted in a table (1) indicates the vast majority of participants were female (85.7%). most of them were from the second class followed by those from the third class. Almost all of the participants were not employed, the rest were workers. Most families depend on the low-income sector.

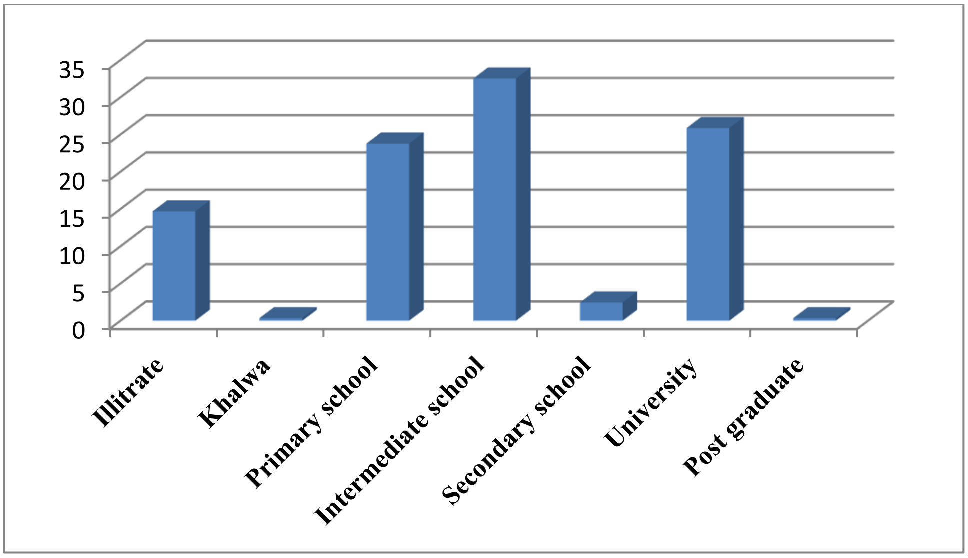

Fig (I): The education level of caregivers

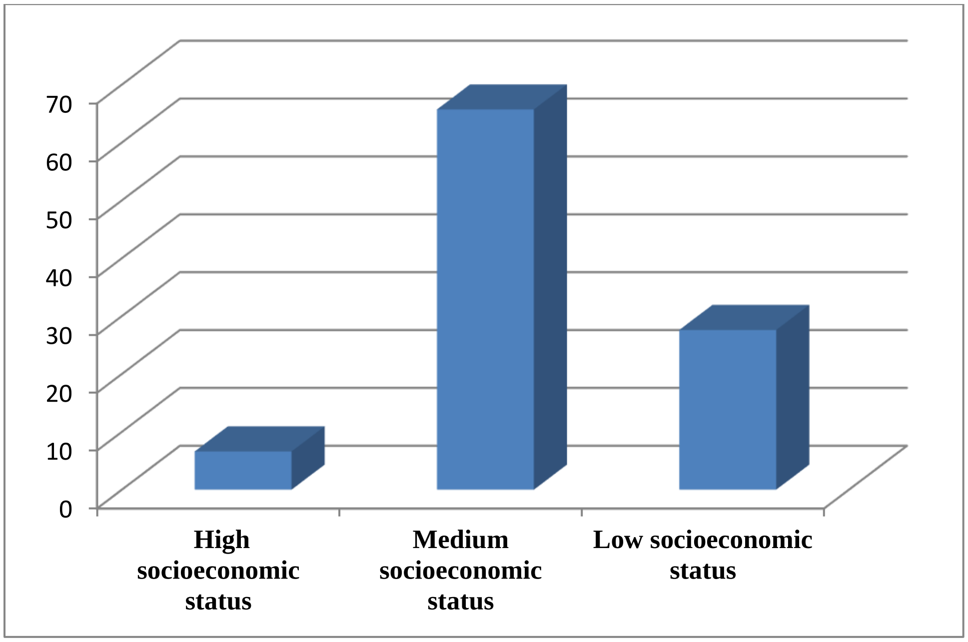

Figure (II): The socioeconomic status categories of the studied group

Table (2) Descriptive statistics of the knowledge of caregivers about the complications of SCD

Descriptive Statistics						
	N	**Range**	**Minimum**	**Maximum**	**Mean**	**Std. Deviation**
Knowledge about the complications of SCD	286	17.00	.00	17.00	9.0524	5.22014

Table (3): The statistical test of the knowledge of caregivers about the complications of SCD

One-Sample T-Test						
Knowledge about the complications of SCD	Test Value = 8.5					
	N		**Mean**	**Std. Deviation**	**Std. Error Mean**	
	286		9.0524	5.22014	.30867	
	t	**df**	**Sig. (2-tailed)**	**Mean Difference**	**95% Confidence Interval of the Difference**	
					Lower	**Upper**
	1.790	285	.075	.55245	-.0551	1.1600

The data depicted in table (3) display the mean in the level of knowledge of caregivers about the complications of SCD, along with the statistic t, together with an associated degree of freedom (df) (285), at 95% confidence interval of the difference, the p-value was (0.075) which mean the level of knowledge of caregivers about the complications of SCD was unsatisfactory.

Table (4): Descriptive statistics of the knowledge of caregivers about the triggers of SCD crisis

Descriptive Statistics						
	N	**Range**	**Minimum**	**Maximum**	**Mean**	**Std. Deviation**
Knowledge about the triggers of SCD crisis	286	13.00	.00	13.00	8.3531	3.15858

Table (5): The statistical test of the knowledge of caregivers about the triggers of SCD crisis.

One-Sample T-Test						
Knowledge about the triggers of SCD crisis	**Test Value = 6.5**					
	N		**Mean**	**Std. Deviation**	**Std. Error Mean**	
	286		8.3531	3.15858	.18677	
					95% Confidence Interval of the Difference	
	t	**df**	**Sig. (2-tailed)**	**Mean Difference**	**Lower**	**Upper**
	9.922	285	.000	1.85315	1.4855	2.2208

The data depicted in a table (5) display the mean in the level of knowledge of caregivers about the triggers of SCD, along with the statistic t, together with an associated degrees-of-freedom **(df)** (285), at 95% confidence interval of the difference, the **p-value** was (0.000) which means the level of knowledge of caregivers about the triggers of SCD was satisfactory.

Table (6): Descriptive statistics of the health beliefs of caregivers toward the complications and triggers of SCD

Descriptive Statistics						
Health beliefs toward the complications and triggers of SCD	**N**	**Range**	**Minimum**	**Maximum**	**Mean**	**Std. Deviation**
	286	64.00	12.00	76.00	36.9615	6.64186

Table (7) The statistical significance of the health beliefs of caregivers toward the complications and triggers of SCD

One-Sample T-Test						
Health beliefs toward the complications and triggers of SCD	**Test Value = 32**					
	N		**Mean**	**Std. Deviation**	**Std. Error Mean**	
	286		36.9615	6.64186	.39274	
					95% Confidence Interval of the Difference	
	t	**df**	**Sig. (2-tailed)**	**Mean Difference**	**Lower**	**Upper**
	12.633	285	.000	4.96154	4.1885	5.7346

The data depicted in table (7) display the mean of caregivers' health beliefs toward the complications and triggers of SCD, along with the statistic t, together with an associated degrees-of-freedom **(df)** (285), at 95% confidence interval of the difference, the **p-value** was (0.000) which mean the health beliefs of caregivers toward the complications and triggers of SCD was satisfactory.

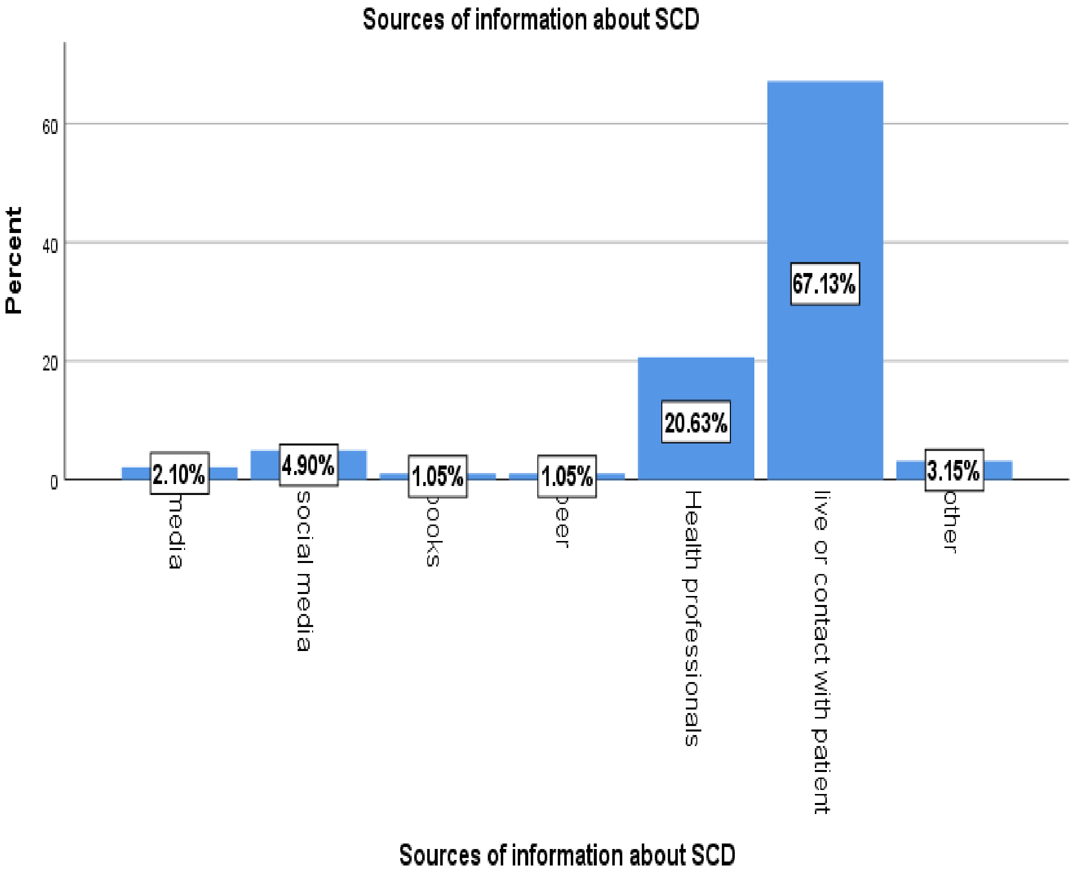

Fig (III): Descriptive statistics of sources of information about SCD

Table (8): Reliability and validity statistics of variables of interest.

Variables of interest	**Reliability**	**Validity**	**N of Items**
Knowledge of caregivers about the complications of SCD	0.915	0.956	17
Knowledge of caregivers about the triggers of SCD crisis	0.784	0.885	13
Health beliefs of caregivers toward the complications and triggers of SCD	0.182	0.426	10

The data depicted in a table (8) revealed the knowledge of caregivers about the complications of SCD is described through 17 items, the Cronbach's alpha = 0.915, which indicates good reliability of participants in the knowledge about the complications of SCD indicator. The validity statistic is acceptable, and it is equal to 0.956. As well as the knowledge of caregivers about the triggers of SCD described through 13 items, the Cronbach's alpha = 0.784, which indicates good reliability of participants for the knowledge of caregivers about the triggers of SCD indicator. The validity statistic is acceptable, and it is equal to 0.885. In contrast, the health beliefs of caregivers toward the complications and triggers of SCD are described through 10 items, the Cronbach's alpha = 0.182, which indicates the unreliability of participants for the health beliefs of caregivers toward the complications and triggers of SCD indicator. The validity statistic is unacceptable, and it is equal to 0.426.

Discussion

The study analyzed the knowledge and health beliefs of caregivers toward the complications and triggers of SCD. It was conducted to determine the extent of their knowledge and health beliefs toward the complications and triggers of SCD**.**

The findings of this study showed the level of knowledge of caregivers about the complications of sickle cell disease was unsatisfactory. It could be related to the lack of social awareness and negative cultural beliefs about the condition. Hence it is important to provide effective and comprehensive health education and counseling programs. This finding was in alignment with the result of the previous study, which showed poor knowledge regarding complications of sickle cell anemia in 57.5% of the participants [6], and inconsistent with the result of a recent study that revealed a good level of knowledge of complications of sickle cell disease [7].

The current study showed that caregivers' knowledge about triggers of SCD crisis was satisfactory. This study's result was similar to the previous study's finding which indicated that respondents had average knowledge about sickle cell and factors that precipitate crises [8], and incongruent with the previous study, which demonstrated that most mothers had poor knowledge about various triggers of SCD, such as fever, infection, dehydration, pain, and nutritional deficiency [6].

In 1989, a study conducted by Gil stated that having adequate knowledge about a disease is important in behavioral adaptation. It also explained that knowledge can influence one's attitude toward the condition [9]. However, this is not the case in the current study because caregivers had poor knowledge about the complications of sickle cell disease and positive health beliefs of caregivers toward the complications and triggers of sickle cell disease. It could be explained by the unreliability of participants in the health beliefs of caregivers toward the complications and triggers of sickle cell disease indicator which was confirmed by Cronbach's alpha test. The current study result agreed with Daaks' [10] and Jaffers' [11] study. At the same time, it misaligned with a study result that concluded that most participants (71.2%) had a poor attitude toward sickle cell disease and premarital genetic counseling [12].

The major sources of information about SCD include live or contact with patients (67.1%), followed by a health professional (20.6%), and social media (4.9%). These findings were aligned with the results of the previous study, which revealed that the primary source of information includes health professionals (36.5%), the Internet (11.1%), friends (13.8%), and family (18.2%) [13]. Therefore, it is crucial to innovate community sensitization via different media and social media channels to provide reliable information to create awareness about the health issue.

In conclusion, this study indicates the level of knowledge of caregivers about the complications of SCD was unsatisfactory. In contrast, the knowledge about the triggers of SCD and the health beliefs of caregivers toward the complications and triggers of SCD were satisfactory.

Recommendations:

Further research studies should be undertaken on a caregiver to investigate the factors affecting the knowledge and health beliefs of caregivers toward the complications and triggers of SCD.

Conflict of interest: The authors have no relevant financial or non-financial interest to disclose to declare

Acknowledgments: The authors thank the manager of Dr. Gaafar Ibnauf Paediatric Hospital. The authors are grateful to all working staff of the sickle cell referral clinic. Last but not least all love and prayers to my patients and their caregivers who participated in this study.

References

1- Neville AK, Panepinto JA. Pharmacotherapy of sickle cell disease. 18th Expert Committee on the Selection and Use of Essential Medicines. 2011 Sep 28;1(1):1-5.

2- Kliegman RM, Behrman RE, Jenson HB, Stanton BM. Nelson text book of pediatrics.18th edition. saunders publishing ; Aug 15.2007. P2026- 2037.

3- Giovanna A.O. Arduini & Alessandra B. Trovó de Marqui. Prevalence and characteristics of priapism in sickle cell disease. Hemoglobin. 2018 Mar 4;42(2):73-7. DOI: 10.1080/03630269.2018.1452760

4- Yawn BP, Buchanan GR, Afenyi-Annan AN, Ballas SK, Hassell KL, James AH, etal . Management of sickle cell disease: summary of the 2014 evidence-based report by expert panel members. JAMA. 2014 Sep 10;312(10):1033-1048. doi:10.1001/jama.2014.10517

5- Ballas SK, Lieff S, Benjamin LJ, Dampier CD, Heeney MM, Hoppe C, etal. Definitions of the phenotypic manifestations of sickle cell disease. American journal of hematology. 2010 Jan;85(1):6-13.

6- Abd El-Gawad SM. Empowering mothers to overcome sickle cell crisis in their children through engagement and education. American Journal of Nursing Research. 2017;5(5):182-90. DOI:10.12691/ajnr-5-5-4

7- Oluwole EO, Okoye CD, Ogunyemi AO, Olowoselu OF, Oyedeji OA. Knowledge, attitude and premarital screening practices for sickle cell disease among young unmarried adults in an urban community in Lagos, Nigeria. The Pan African Medical Journal. 2022;42. DO -
10.11604/pamj.2022.42.8.27705

8- Nakazwe E, Mwanakasale V, Siziya S. Knowledge attitude and practices of parents with children suffering from sickle cell disease towards factors that precipitate sickle cell crises, at arthur davidson children's hospital in Ndola Zambia. AGE. 2017;18:18-27.

9- Gil KM, Abrams MR, Phillips G, Keefe FJ. Sickle cell disease pain: relation of coping strategies to adjustment. JCCP. 1989 Dec;57(6):725-731
https://doi.org/10.1037/0022-006X.57.6.725

10- Daak AA, Elsamani E, Ali EH, Mohamed FA, Abdel-Rahman ME, Elderdery AY,etal. Sickle cell disease in western Sudan: genetic epidemiology and predictors of knowledge attitude and practices. Trop Med Int Health. 2016 May; 21(5):642-53.
Available from:
https://pubmed.ncbi.nlm.nih.gov/27028397/

11- Dihya EJ, Fatima KA, Khadija MA, Nafeesa AM, Ridha AH, Zainab MH. Adult sickle cell diseased patients' knowledge and attitude toward the preventive measures of sickle cell disease crisis. International Journal of Nursing and midwifery. 2009 Nov 30;1(2):010-8.

12- Al-Qattan HM, Amlih DF, Sirajuddin FS, Alhuzaimi DI, Alageel MS, Bin Tuwaim RM, Al Qahtani FH. Quantifying the levels of knowledge, attitude, and practice associated with sickle cell disease and premarital genetic counseling in 350 Saudi adults. Advances in Hematology. 2019 May 2;2019 https://doi.org/10.1155/2019/3961201

13- Olakunle OS, Kenneth E, Olakekan AW, Adenike OB. Knowledge and attitude of secondary school students in Jos, Nigeria on sickle cell disease. Pan African Medical Journal. 2013;15(1).