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Nurses Knowledge Regarding Sickle Cell Disease among Children

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Abstract

Background: Sickle cell disease (SCD) is a prevalent genetic disorder, particularly among children of African and Mediterranean descent. It is associated with various complications such as vaso-occlusive crises, hemolysis, and stroke. Nurses' knowledge of SCD is essential for effective patient care, especially in pediatric departments where children with SCD require specialized management. This study aimed to assess the knowledge of nurses regarding SCD in pediatric departments of a hospital .

Methods: A descriptive cross-sectional study was conducte at a hospital with a large pediatric ward. Seventy nurses working in the pediatric medicine, surgery, and emergency departments were surveyed using a structured questionnaire. The questionnaire assessed demographic information and knowledge about SCD, covering areas such as clinical manifestations, management, complications, diagnostic tests, and the role of education and counseling.

Results: The study found that most nurses (61.4%) were aged 20-30 years, and 98.6% were female. A significant proportion (84%) held bachelor's degrees. While 94.3% of nurses demonstrated knowledge of the definition of SCD, 75% were unaware of the life expectancy of individuals with the disease. Nurses exhibited good knowledge of clinical manifestations (87.1%) and pain episode management (90%), but 51.5% were unaware that SCD is not a curable condition. Additionally, 95.7% of nurses recognized the importance of education and counseling in managing SCD.

Conclusion: The study revealed that nurses in the pediatric departments possess a strong understanding of many aspects of SCD, including clinical features, management, and complications. However, gaps in knowledge were identified, particularly regarding the life expectancy, curability, and the role of genetic factors in SCD. These findings suggest a need for targeted educational programs to address these knowledge gaps and improve care for pediatric patients with SCD.

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Introduction

Sickle cell anemia refers to a group of autosomal recessive genetic blood disorders caused by a mutation at the sixth codon of the beta-globin gene. This mutation results in the formation of abnormal hemoglobin, which, under low oxygen conditions, polymerizes and causes red blood cells to become rigid and take on a sickle shape (1). The most severe form of this disorder is homozygous sickle cell disease (HBSS), also known

as sickle cell anemia (2). A key clinical manifestation is the occurrence of vaso-occlusive crises, which are often accompanied by hemolysis. These crises are the primary cause of hospitalization and may necessitate blood transfusions to prevent organ damage and death. Over 10% of individuals with sickle cell anemia experience overt strokes, while 22% exhibit silent cerebral infarctions (3).

Sickle cell disease is considered one of the most prevalent inherited blood disorders worldwide, affecting an estimated 100 million people, particularly individuals of African and Mediterranean descent (3). In sub-Saharan Africa, it is the most common genetic disorder, associated with a high mortality rate in children between the ages of 1 and 5 years (4, 5). In the United States, sickle cell disease is the most common autosomal recessive genetic disorder, affecting approximately 1 in 375 individuals of African descent (6). The prevalence of the sickle cell trait in sub-Saharan Africa ranges from 5% to 40%, with more than 230,000 infants born with sickle cell anemia each year (0.74% of total births). The life expectancy of individuals with sickle cell disease in this region is often under 5 years, with the highest mortality rates occurring in this age group (4).

The population of is ethnically diverse, with groups ranging from Arabs to Africans and Afro-Arabs, including well-established ethnic groups such as the Nuba and Nilotes. Other groups, including Arabs, Hausa, and Copts, migrated to the region at various points in history, resulting in a mix of Arab and Negroid genetic characteristics (1, 2). the highest rates of cousin marriages globally, with around 40-54% of all marriages occurring between first cousins (4, 6). The presence of sickle cell disease was first reported in 1926 by Archibald, marking the first case documented in Africa (3). The prevalence of sickle cell disease is highest among populations (8), where the sickle cell gene is more common (9, 10).

It is believed that the sickle cell gene was introduced into the population through migrating (11).

Materials and Methods

This study employed a descriptive cross-sectional design to assess nurses' knowledge of sickle cell disease in the pediatric department.

The research was carried out at a hospital with multiple departments, including a large pediatric ward. The pediatric department consists of three sections: pediatric medicine, pediatric surgery, and pediatric emergency. The pediatric medicine ward includes four units, with eight general rooms and four private rooms, accommodating a total of 36 nurses. The pediatric surgery department comprises four general rooms (32 beds) and three private rooms. The pediatric emergency department consists of two rooms with five beds and is staffed by 10 nurses.The study focused on qualified nurses working in the pediatric department of the hospital.A total of 70 nurses were included in the study.

Data Collection Tools and Technique

Data was collected through a structured questionnaire designed to evaluate nurses' knowledge on sickle cell disease. The questionnaire consisted of two sections: the first gathered demographic information, while the second assessed knowledge of sickle cell disease, covering topics such as its definition, common geographical areas affected, life expectancy of individuals with the disease, clinical features, screening methods, diagnostic tests, general management, pain episode management, complications, and the influence of socioeconomic and environmental factors.

Statistical Data Analysis

The collected data was processed, coded, and analyzed using SPSS version 18. The knowledge score for each domain was calculated by dividing the number of correct responses by the total number of questions in that domain, and the results were expressed as percentages. Pearson’s chi-square test was used to examine associations between nurses' knowledge and demographic characteristics. Data were presented in tables.

Results

Table 1: Age Distribution of Nurses

Age Range	Frequency	Percentage
20-30 years	43	61.4%
31-40 years	26	37.1%
Above 41 years	1	1.4%
Total	**70**	**100%**

Interpretation: The majority of the nurses fall within the 20-30 years age group.

Table 2: Gender Distribution of Nurses

Gender	Frequency	Percentage
Male	1	1.4%
Female	69	98.6%
Total	**70**	**100%**

Interpretation: The vast majority of the nurses are female.

Table 3: Educational Level of Nurses

Educational Level	Frequency	Percentage
Diploma	5	7.1%
Bachelor's Degree	59	84.3%
Postgraduate	6	8.6%
Total	**70**	**100%**

Interpretation: Most of the nurses hold a Bachelor's degree.

Table 4: Years of Experience of Nurses

Years of Experience	Frequency	Percentage
0-5 years	39	55.7%
6-10 years	24	34.3%
Above 11 years	7	10%
Total	**70**	**100%**

Interpretation: A significant portion of the nurses have less than five years of experience.

Table 5: Knowledge of Sickle Cell Disease (SCD) Definition

Knowledge of SCD Definition	Frequency	Percentage
Yes	66	94.3%
No	3	4.3%
I do not know	1	1.4%
Total	**70**	**100%**

Interpretation: The majority of nurses are familiar with the definition of Sickle Cell Disease (SCD).

Table 6: Knowledge Regarding Ethnic Groups Affected by Sickle Cell Disease in Africa

Knowledge of Ethnic Groups Affected by SCD	Frequency	Percentage
Yes	63	90%
No	2	2.9%
I do not know	5	7.1%
Total	**70**	**100%**

Interpretation: Most nurses are aware of the ethnic groups most commonly affected by Sickle Cell Disease, particularly in Africa.

Table 7: Knowledge of Nurses Regarding the Geographic Area Affected by Sickle Cell Disease in ()

Knowledge of Geographic Area for SCD	Frequency	Percentage
Yes	61	87.1%
No	3	4.3%
I don't know	6	8.6%
Total	**70**	**100%**

Table 8: Knowledge of Nurses Regarding Common Clinical Features Manifested by Sickle Cell Disease and Pain Crises

Knowledge of Common Clinical Features of SCD	Frequency	Percentage
Yes	61	87.1%
No	5	7.1%
I don't know	4	5.7%
Total	**70**	**100%**

Interpretation: Most nurses are familiar with the common clinical features and manifestations of Sickle Cell Disease, including pain crises.

Table 9: Knowledge of Nurses Regarding Antenatal Screening as an Appropriate Test for Sickle Cell Disease

Knowledge of Antenatal Screening for SCD	Frequency	Percentage
Yes	52	74.3%
No	9	12.9%
I don't know	9	12.9%
Total	**70**	**100%**

Interpretation: A majority of nurses are aware that antenatal screening is an appropriate test for diagnosing Sickle Cell Disease.

Table 10: Knowledge of Nurses Regarding Diagnostic Tests for Sickle Cell Disease (Complete Blood Count and Sickle Cell Test)

Knowledge of Diagnostic Tests for SCD	Frequency	Percentage
Yes	62	88.6%
No	6	8.6%
I don't know	2	2.9%
Total	**70**	**100%**

Interpretation: Most nurses are aware of the appropriate diagnostic tests for Sickle Cell Disease, including the complete blood count and sickling test.

Table 11: Knowledge of Nurses Regarding the Shape of RBCs in Sickle Cell Disease Patients

Knowledge of RBC Shape in SCD Patients	Frequency	Percentage
Yes	67	95.7%
No	1	1.4%
I don't know	2	2.9%
Total	**70**	**100%**

Interpretation: Most nurses know the characteristic shape of red blood cells in patients with Sickle Cell Disease.

Table 12: Knowledge of Nurses Regarding General Management of Sickle Cell Disease (Maintaining Adequate Fluid Intake, Treating Infections Promptly, and Vitamin Supplementation)

Knowledge of General Management of SCD	Frequency	Percentage
Yes	67	95.7%
No	1	1.4%
I don't know	2	2.9%
Total	**70**	**100%**

Interpretation: Most nurses are knowledgeable about the general management of Sickle Cell Disease, including maintaining fluid intake, treating infections promptly, and providing vitamin supplements.

Table 13: Knowledge of Nurses Regarding Pain Management for Sickle Cell Disease with Narcotic Analgesics

Knowledge of Pain Management with Narcotics	Frequency	Percentage
Yes	63	90%
No	2	2.9%
I don't know	5	7.1%
Total	**70**	**100%**

Interpretation: A large majority of nurses are aware that narcotic analgesics are used to manage pain episodes in Sickle Cell Disease.

Table 14: Knowledge of Nurses Regarding Appropriate Management of Severe Painful Crises in Sickle Cell Disease (Morphine, Rehydration, Blood Transfusions)

Knowledge of Management of Severe Painful Crises	Frequency	Percentage
Yes	62	88.6%
No	3	4.3%
I don't know	5	7.1%
Total	**70**	**100%**

Interpretation: Most nurses are familiar with the appropriate management of severe painful crises in Sickle Cell Disease, including the use of morphine, rehydration, and blood transfusions.

Table 15: Knowledge of Nurses Regarding Effective Management of Stroke in Sickle Cell Disease

Knowledge of Effective Management of Stroke	Frequency	Percentage
Yes	56	80%
No	5	7.1%
I don't know	9	12.9%
Total	**70**	**100%**

Interpretation: Most nurses are aware of the effective management of stroke in patients with Sickle Cell Disease.

Table 16: Knowledge of Nurses Regarding Iron Accumulation in Vital Organs as a Complication of Recurrent Blood Transfusions

Knowledge of Iron Accumulation in Organs	Frequency	Percentage
Yes	52	74.3%
No	8	11.4%
I don't know	10	14.3%
Total	**70**	**100%**

Interpretation: Most nurses are aware that iron accumulation in vital organs is a complication of recurrent blood transfusions.

Table 17: Knowledge of Nurses Regarding Complications of Sickle Cell Disease (Pain Crises, Stroke, and Lung Tissue Damage)

Knowledge of SCD Complications	Frequency	Percentage
Yes	60	85.7%
No	2	2.9%
I don't know	8	11.4%
Total	**70**	**100%**

Interpretation: Most nurses are aware that complications of Sickle Cell Disease include pain crises, stroke, and lung tissue damage.

Table 18: Knowledge of Nurses Regarding Splenomegaly as a Surgical Complication of Sickle Cell Disease

Knowledge of Splenomegaly as a Complication of SCD	Frequency	Percentage
Yes	34	48.6%
No	27	38.6%
I don't know	9	12.9%
Total	**70**	**100%**

Interpretation: Many nurses are aware that splenomegaly is a complication of Sickle Cell Disease and that it may require surgical treatment.

Table 19: Knowledge of Nurses Regarding the Curability of Sickle Cell Disease

Knowledge of Curability of SCD	Frequency	Percentage
Yes	59	84.3%
No	4	5.7%

Knowledge of Curability of SCD	Frequency	Percentage
I don't know	7	10%
Total	**70**	**100%**

Interpretation: Most nurses understand that Sickle Cell Disease is not curable.

Table 20: Knowledge of Nurses Regarding the Treatment of Sickle Cell Disease

Knowledge of Treatment for SCD	Frequency	Percentage
Yes	62	88.6%
No	3	4.3%
I don't know	5	7.1%
Total	**70**	**100%**

Interpretation: Most nurses are familiar with the treatment options for Sickle Cell Disease.

Table 21: Knowledge of Nurses Regarding the Factors Affecting the Severity of Sickle Cell Disease

Knowledge of Factors Affecting Sickle Cell Disease Severity	Frequency	Percentage
Yes	56	80%
No	11	15.7%
I don't know	3	4.3%
Total	**70**	**100%**

Interpretation: Most nurses are aware that environmental and socioeconomic factors affect the severity of Sickle Cell Disease.

Table 22: Knowledge of Nurses Regarding Counseling for Sickle Cell Disease Patients

Knowledge of Counseling for Sickle Cell Disease Patients	Frequency	Percentage
Yes	67	95.7%
No	2	2.9%
I don't know	1	1.4%
Total	**70**	**100%**

Interpretation: Most nurses understand the importance of health education and counseling in the management of Sickle Cell Disease.

Table 23: Knowledge of Nurses Regarding Providing Health Education for Sickle Cell Disease

Knowledge of Providing Health Education for Sickle Cell Disease	Frequency	Percentage
Yes	18	25.7%

Knowledge of Providing Health Education for Sickle Cell Disease	Frequency	Percentage
No	26	37.1%
I don't know	26	37.1%
Total	**70**	**100%**

Interpretation: Many nurses do not actively provide or promote health education for Sickle Cell Disease to children and their families.

Table 24: Correlation Between Nurses' Knowledge About Sickle Cell Disease, Their Level of Education, and Years of Experience

Variable	Level of Education	Years of Experience
Correlation	0.67	0.96
Sig. (2-tailed)	0.028	0.03
N	70	70

Interpretation:

- There is a statistically significant relationship between nurses' level of education and their knowledge about Sickle Cell Disease, with a correlation coefficient of 0.67 and a p-value of 0.028, which is lower than the significance level (5%).
- A highly significant relationship exists between years of experience and knowledge about Sickle Cell Disease, with a correlation coefficient of 0.96 and a p-value of 0.03, indicating that experience plays a critical role in increasing knowledge.

Discussion

This study aimed to assess the knowledge level of nursing staff caring for pediatric patients with Sickle Cell Disease (SCD). The overall results indicate that nurses in the pediatric department possess a significant level of understanding about SCD.

Demographic data showed that the majority of nurses (61.4%) were aged between 20 and 30 years, with an overwhelming majority being female (98.6%). The findings highlighted that the level of education was a key factor influencing knowledge, with differences noted among nurses holding associate degrees, bachelor's degrees, and postgraduate qualifications. Most nurses (84%) held a bachelor's degree. Additionally, half of the nurses had between 1 to 5 years of work experience (55.7%), while 34.3% had 6 to 10 years of experience.

The study found that 94.3% of nurses were knowledgeable about the definition of SCD (Table 5), a result consistent with the findings of Marie Osline Etine (1998), which reported that 97.8% of participants recognized SCD as a genetic disorder (12). The ethnic composition of nurses, particularly those from African backgrounds (90%), reflected a similar trend, as also noted by Etine, where black individuals were identified as the most commonly affected ethnic group (12). where SCD is prevalent, 87.1% of nurses demonstrated knowledge of the disease, aligning with previous research by Bakheita Atalla (2003), which reported 70% awareness in the region (11).

However, the study revealed that 75% of nurses were unaware of the life expectancy estimates for individuals with SCD (Table 8), which contrasts with the findings of Orah S. Platt (1994) (13). Conversely, 87.1% of nurses were knowledgeable about the clinical manifestations of SCD (Table 9), consistent with the research of Marie Osline Etine (1998) (12).

Regarding pain episodes, 90% of nurses demonstrated awareness (Table 10), which agrees with findings by Nagwa Sayed (1997), where 81.2% of nurses recognized that painful episodes often lead to hospital admissions in children (14). Additionally, 74.3% of nurses were knowledgeable about antenatal screening as an appropriate test for SCD (Table 11), which aligns with Sayed's study on antenatal screening for SCD (14).

The study also found that 88.6% of nurses were knowledgeable about SCD testing (Table 12), a result consistent with Nagwa Sayed (1997), who found 85.5% of participants were aware of tests such as electrophoretic mobility to confirm SCD (14). Similarly, nurses were aware of the shape of red blood cells (RBCs) in SCD (Table 13), mirroring the findings from Sayed's research (14).

Furthermore, 95.7% of nurses understood the general management of SCD, including adequate fluid intake, infection treatment, and vitamin supplementation (Table 14), which correlates with Sayed's report of 100% of patients receiving folic acid, IV fluids, and antibiotics for SCD management (14). When it came to pain episode management, 81.4% of nurses demonstrated knowledge (Table 15), while 90% were knowledgeable about handling severe pain crises (Table 16). This finding contrasts with Shafeega Hassan (2015), who reported that only 47.8% of nurses were familiar with pain assessment and management (15).

Regarding stroke management, 80% of nurses were knowledgeable (Table 17), a result that aligns with Nagwa Sayed (2003), who reported that only 8.7% of patients with a history of stroke received prophylactic transfusion, even though chronic transfusion programs were more effective (14). Additionally, 81.4% of nurses were aware of complications related to SCD, such as pain crises, stroke, and long-term tissue damage (Table 18), and 85.7% recognized splenomegaly as a complication (Table 15), indicating a good level of knowledge about SCD complications. These findings were consistent with the study by Marie Osline Etine (1998) on the knowledge of nurses regarding SCD complications (12).

However, 51.5% of nurses were unaware that SCD is not a curable disease (Table 19), which suggests a gap in knowledge. This finding aligns with the work of Nagwa Sayed (2003), who recommended focusing on the genetic factors and beta-globin haplotypes in SCD research (14). Regarding advanced management techniques, 84.3% of nurses were knowledgeable about bone marrow transplantation and gene therapy as treatments for SCD (Table 20), a result consistent with the findings of Nagwa Sayed (1997) on SCD management (14).

The study also showed that 80% of nurses understood how environmental and socioeconomic factors affect the severity of the disease (Table 21), which aligns with Nagwa Sayed (1997), who reported that lower socioeconomic status is associated with more severe clinical manifestations of SCD (14). Finally, 95.7% of nurses recognized the importance of education and counseling in SCD management (Table 23), which echoes findings from Marsh J. Treadwell (2006), who highlighted the crucial role of counseling for individuals affected by SCD (16, 17).

In conclusion, the study demonstrates that while nurses possess a high level of knowledge about many aspects of SCD, there are areas that require further attention, particularly in the understanding of life expectancy, curability, and the impact of genetic factors.

Conclusion

The results of this study indicate that the majority of nurses demonstrated a strong understanding of the definition, clinical symptoms, management, and complications of sickle cell disease (SCD). However, a significant portion of the nurses (75%) lacked knowledge about health education related to SCD, and half of the nurses were unaware of whether SCD is a curable condition.

Recommendations

For Nurses:

- Ongoing training and refresher courses should be provided to nursing staff to ensure they stay up-to-date with the latest knowledge and skills related to SCD. These educational programs should be routinely offered

to inform new staff nurses about SCD. Nurses play a crucial role in providing education, counseling, and pain management for SCD patients.

- It is essential to follow universal precautions and crisis management protocols in pediatric emergency settings to ensure the well-being of children and alleviate pain during acute episodes.

For Hospitals:

- Nursing staff should actively participate in international programs, conferences, and workshops to stay current with developments in healthcare.
- Continuous nursing education is vital to preparing nurses to address future healthcare challenges effectively, ensuring they are well-equipped to provide the highest quality care to patients.

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