Prevalence of Sickle Cell Disease among Children aged 1 – 15 Years attending the Sickle Cell Clinic at Luwero Hospital in Luwero District. A Cross-section Study.

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Abstract

Background:
To determine the prevalence of sickle cell disease among children aged 1-15 years at Luwero Hospital in the Luwero district.

Objectives:
To determine the prevalence of sickle cell disease among children aged 1-15 years at Luwero Hospital in the Luwero district, to determine the average haemoglobin concentration among sicklers aged 1-15 years at Luwero Hospital, and to find out the effectiveness of health supervision and management of sicklers aged 1-15 years by their guardians in Luwero district.

Study design:
The researcher conducted a cross-section study to obtain data among children aged 1-15 years attending the sickle cell clinic.

Results:
The age of children ranged from 1-to 15 years with a mean age of 6.9 years. The majority of the children were girls 66(68.75%). The majority of their parents/guardians were female 70(72.9%), divorced 57(59.4%), practicing farmers 43(44.8%) residing in rural areas 68(70.9%). Of the 96 children, 11 tested positive for SCD giving a prevalence of 11.5%. For haemoglobin concentration, the minimum Hb was 3.1 g/dl and the maximum Hb was 12.9g/dl with a mode of 5.6 g/dl for the blood samples that were analyzed with the CBC machine.

Conclusion:
The prevalence of SCD was high as revealed to be 11.5% during the study period and the average Hb level was 6.8 g/dl with the modal Hb 5.6 g/dl and many children were heading to the severe form of SCA.

Recommendation:
The government through the MOH should set aside funds to facilitate awareness projects encouraging premarital testing of couples for sickle cell trait and made mandatory to curb the increasing prevalence of SCD and providing enough funds for much stock of medication for the sicklers to prevent early mortality and for better management and supervision.

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1 Background of study.

The disease occurs due to a mutation of the beta globin gene of hemoglobin, causing a substitution of the glutamic amino acid for valine at position 6 of the beta chain, thereby producing abnormal hemoglobin called hemoglobin S (HbS) instead of the normal hemoglobin A (HbA).

When deoxygenated HbS polymerizes and precipitates, the erythrocytes change in form, a deformity of red blood cells which become sickle shaped resulting in hemolytic anemia which in turn causes acute complications like ischemic damage to tissues. SCD is a group of genetic diseases that is especially prevalent in tropical and subtropical regions, however, forced migrations and ongoing population movement have spread it throughout the world. (Karadag, 2018). Some sudden complications of SCD cause the body's red blood cell count to drop to life-threatening levels (severe anemia). When severe, these complications (including splenic sequestration, acute chest syndrome, and aplastic crisis) can be fatal if not treated with blood transfusion. (Steinberg MH, 2016).

SCD is a lifelong illness. A blood and bone marrow transplant is currently the only cure for SCD but there are effective treatments that can reduce symptoms and prolong life. (NHLBI, 2022). Specialized comprehensive medical care decreases morbidity and mortality during childhood. The provision of comprehensive care is a time-intensive endeavor that includes ongoing patient and family education, periodic comprehensive education and other disease-specific health maintenance services, psychosocial care, and genetic counseling. (Brousseau et al., 2010)

SCD is an increasing global health problem. Every year approximately 300,000 infants are born with sickle cell anemia (SCA) which is defined as homozygosity for the sickle hemoglobin (HbS) gene. (Steinberg MH, 2016). The global estimates for the birth of SCD were 42297 per 100,000 (42%) with the highest birth prevalence in Africa at 16121 per 100,000 (16%). (Igor Rudan, 2018).

As revealed by Assaf in 2020, most of the world's SCD burden is in Africa where it's a major contributor to child morbidity and mortality. A study in Nigeria showed that early diagnosis allows initiation of well-established public measures for primary prevention including penicillin prophylaxis, routine child vaccination, parental education about prompt medical management for fever, and detection of splenic sequestration. (Odunvbun et al, 2015) .

A study done in Uganda by MOH revealed that over 25,000 babies are born with SCD annually. (MOH, June 2019) even though Uganda's Ministry of Health has endeavored to scale awareness programs to encourage early diagnosis and treatment options in the central region of Uganda Luwero district inclusive but little success has been achieved. (MOH SCD Report, 2016)

2 Methodology

2.1 Study design

A cross-sectional research design was used in the study. This research design was used because it examines the relationship between the disease and other variables of interest as they exist in a defined population and this addressed objective two and analyzed data on the health supervision and management of children having sickle cell disease.

The study was quantitative because it enabled the researcher to get information from the selected population described and document aspects of the situation as they occur among them. The key information through interviews and document analysis was used through participants filling questionnaires. The research design was preferred because it involves appropriate statistical techniques like frequencies and percentage.

2.2 Study area

The study was conducted at Luwero hospital in Luwero district at the sickle cell clinic. Luwero hospital is in the Centre of Luwero town council. Luwero district is found in central Uganda along Kampala – Gulu high way boarded by Nakasongola district to the north, Kayunga to the east, Nakaseke to the west and Wakiso to south.

The study area was chosen because it was easily accessible by researcher as he stays within the study area and transport costs were reduced during data collection. Additionally, the hospital has special sickle cell clinic which increased on case availability as far was the study concerned

2.3 Study population

The study population comprised of all children aged 1 – 15 years presenting with signs and symptoms of sickle cell seeking health care services at Luwero hospital.

2.4 Sample size determination

The sample size was determined using the formula by Kish and Leslie (1965)

$$N = \frac{(Z^2PQ)}{d^2}$$

where;

Z = confidence limit corresponding to 95% confidence interval 1.96 for 95% confidence interval.

P = Estimated prevalence. In this case, the prevalence is not known and thus 50% (0.5) will be taken

Q = Percentage of people not affected (1-P), 1-0.5 = 0.5

d = Error of precision 10% (0.1).

Therefore;

$N = \{(1.96)^2(0.5)(0.5)\}$

$(0.1)^2$

$= 96.04$

The sample size consisted of 96 respondents from the whole population.

2.5 Sampling technique

A systematic random sampling technique was employed. Parents / guardians who brought their children with sickle cell signs and symptoms for testing and treatment at Luwero hospital were sampled to collect information.

2.6 Sampling procedure

A systematic random sampling technique was used as three children aged 1 – 15 years were selected and one was skipped for those who came to Luwero hospital at the sickle cell clinic. This was done following the subgroups formed i.e., 1 – 5 years, 6 – 10 years and 11 – 15 years.

2.7 Data collection method

In this study, questionnaire, facial interview, observation and screening solubility sickling test were used to collect primary data from attending patients.

Observation method looked for signs such as yellowing of the eyes and stunted growth. Collected blood was taken to the laboratory for screening and confirmed by hemoglobin electrophoresis and this was kept confidential.

2.8 Data collection tool(s)

Well- designed close and open-ended questionnaires were used to collect information from parents / guardians by the researcher. This tool was used because it minimized time wastage and allowed many respondents to be interviewed at the same time.

Laboratory request forms were used to collect information about the patient samples from the laboratory. Observation, the researcher observed for the signs present with children that were brought for screening like stunted growth making the researcher actively participate thus had a better understanding about the sickling cell testing during the research study.

2.9 Data collection procedures

After approval of my proposal, the researcher obtained an introductory letter from the school administration of St Francis school of health sciences. This was handed to the laboratory manager at Luwero hospital who granted me permission to carry out the study.

The researcher then proceeded and obtained informed consent from the participant's guardians, explained the purpose of the study to them. He had facial interview with the participants and handed the questionnaire to the guardians of the children and guided them where necessary.

Venous blood samples were collected from the children into the purple topped EDTA vacutainers for hemoglobin estimation and sickling solubility test. The completed questionnaires were kept in safe custody and were only accessible by the researcher. The exercise was repeated until a total of 96 respondents were obtained.

2.10 Study variables

The predictor variable comprised of hemoglobin levels among sicklers, the health supervision and management included access to the health facility, monitoring for anemic signs and follow up of children by their care takers and the economic status of the parents/guardians.

Quality control

The questionnaires were pre – tested among 15 parents with children attending the out- patient department. Two laboratory assistants who helped the researcher were thoroughly oriented about the objectives and the purpose of the study.

The SOPS and manufacturer's instructions on how to use the reagents were followed while performing the sickling solubility test. Proper labeling of the specimen was observed correctly and ensured that there was no interchange of samples. The patients' samples were collected in sterile specimen containers to avoid contamination. The samples were worked on immediately and not placed in direct sunlight to prevent hemolysis of the samples.

2.10.1 Inclusion criteria

Only children with sickle cell clinical signs and symptoms attending health care services at Luwero hospital whose parents had consented and present at the time of data collection were considered for the study. Only children in the age bracket of 1 – 15 years were involved.

2.10.2 Exclusion criteria

The study excluded all patients above 16 years and their parents had not consented and refused to take off blood samples from their children for hemoglobin estimation and sickle cell testing.

2.10.3 Piloting the study

A pilot study was conducted at Bishop Ceaser Asili Hospital to pretest the research tool i.e., Questionnaires and corrections were changed accordingly

3 Data analysis and presentation

The data was collected analyzed using Microsoft excel and in themes by use of descriptive concepts. Data was presented in frequency tables and this was used to describe the study population and the extent of sickle cell disease.

3.1 Ethical consideration

I acquired an introductory letter from the school administration and presented it to the laboratory manager at Luwero hospital who allowed me to carry out the research study.

The researcher properly identified himself to the respondents involved in the study. The participants had to consent before enrollment. The researcher explained to the parents of these children the intensions of the study and why it's necessary to participate thus consented voluntarily.

The parents were informed about their freedom to decline in responding to any of the questions that they feel uncomfortable with.

Confidentiality of the information given by the parents / guardians and their identities was not included anywhere in the research and this was achieved by discouraging them from including their names in the questionnaires.

4 Data analysis and presentation

4.1 Social demographic characteristics of the participants.

Most children were female 66(68.75%) with their age ranging from 1 – 15 years. When grouped by age the majority were between 6 – 10 years (55.2%) with the mean age of 6.9 years while the ones under 11-year-old age group were the smallest with 11.5%. The majority of the parents/guardians were female 70(72.9%), divorced 57(59.4%) and the highest education background was secondary 45(46.9%). Most parents were farmers 43(44.8%) residing in rural areas 68(70.9%).children's different age groups and their number.

4.2 The prevalence of sickle cell disease (SCD) among children aged 1- 15 years (N=96)

A total of 96 children were tested for SCD over the study period out of whom 11 tested positives were confirmed by Hemoglobin electrophoresis. This amounted to a prevalence of SCD among children at 11.5%

4.3 Average hemoglobin levels among children with sickle cell disease.

The blood samples that were collected from 96 sickle cell diseased children and analyzed by the CBC machine for hemoglobin concentration as the parameter of interest gave the minimum Hb concentration as 3.1 g/dl and the maximum Hb as 12.6 g/dl. The average Hb concentration was 6.8 g/dl with the mode being 5.6 g/dl and the median of 6.2 g/dl.

Children whose hemoglobin concentration was below 5g/dl were 20. According to age groups, 1 – 5 years of age were 13 (65%) and 6 – 10 years were 7 (35%). Thus, the haemoglobin concentration among sickle cell children decreased with age indicating the fact that those below 5years of age were at great risk of sickle cell anemia. (Represented in figure 4 below)

5 Discussion, conclusion, and recommendations

Discussion:

Prevalence of SCD among children aged 1 – 15 years of age

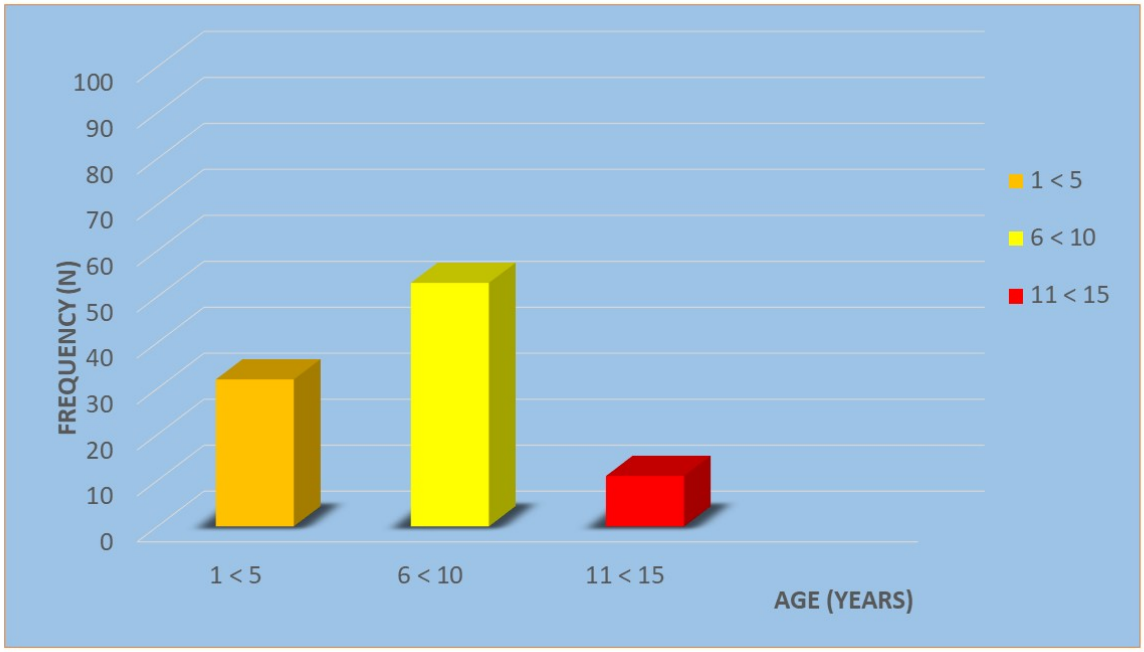

Figure 1. A bar graph showing the children's different age groups and their number

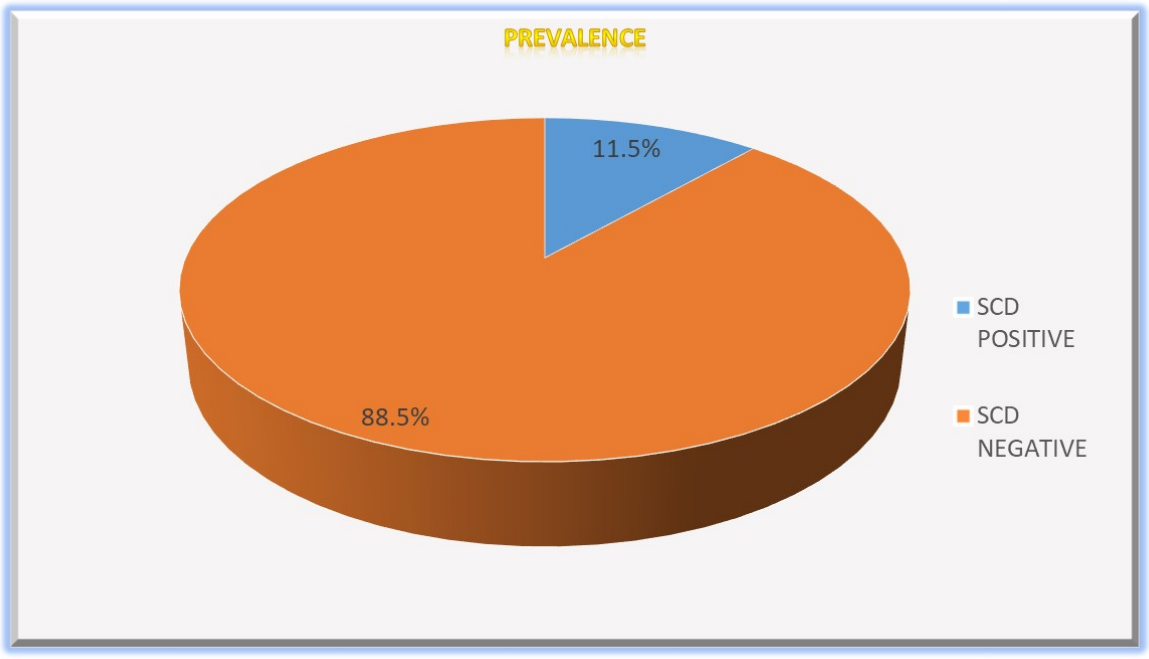

Figure 2. A pie chart showing the SCD test results for children. (N= 96)

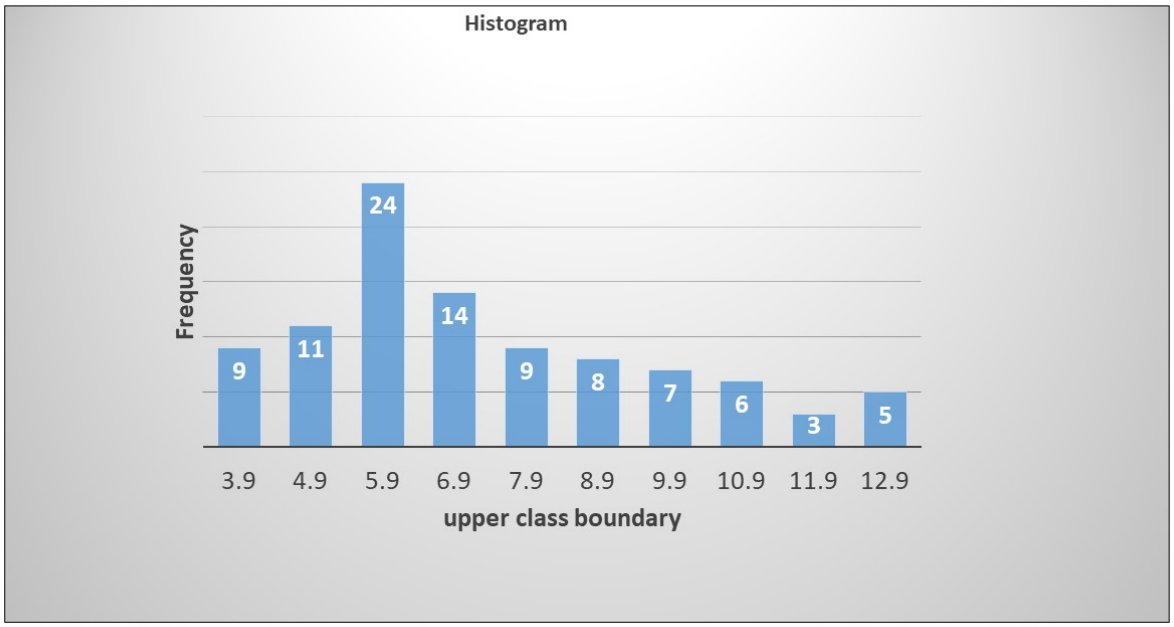

Figure 3. A histogram showing the hemoglobin levels among the sickle cell children.

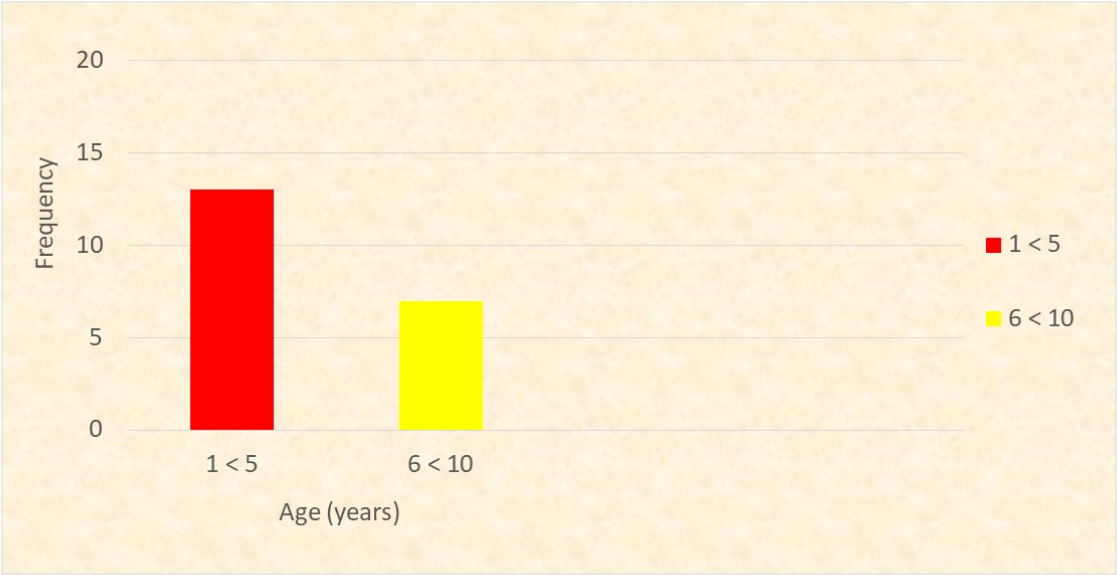

Figure 4. A bar graph showing the number of children with their Hb below 5g/dl according to age groups.

Table 1. Showing the demographic characteristics of the parents and their children

Sex	Frequency (N)	Percentage (%)
Male	26	27.1
Female	70	72.9
Marital status		
Married	39	40.6
Divorced	57	59.4
Education		
Primary	36	37.5
Secondary	45	46.9
Tertiary	15	15.6
Occupation		
Civil servant	15	15.6
Farmer	43	44.8
Self employed	38	39.6
Residence		
Rural area	68	70.9
Urban area	28	29.1

Source: Bio data 2021

Table 2. showing demographic characteristics for the children.

Sex	Frequency (N)	Percentage (%)
Male	30	31.25
Female	66	68.75
Age (years)		
1 – 5	32	33.3
6 – 10	53	55.2
11 – 15	11	11.5

Source: Bio data 2021

Table 3. A table showing the test results for SCD among the children.

SCD TEST RESULTS	NUMBER (N=96)	PERCENTAGE (%)
SCD positive	11	11.5
SCD negative	85	88.5

Source: Bio data 2021

The objective of the study was to determine the prevalence of SCD among children aged 1-15 years at LH. Data analysis and interpretation revealed the major findings under this objective. It revealed that out of the 96 children 11 tested positive. These findings indicate that the prevalence estimate was 11.5% which is lower than the prevalence study which was conducted by Ambrose in 2017 in Tanzania at 12.1%. This could be a result of individual factors such as negligence and inadequate health education about SCD.

As partners divorce learning of having a child with SCD, this indicates the knowledge gap about SCD and premarital testing for one to know their haemoglobin genotype having tested for sickle cell is very low which agrees with the study conducted in Nigeria by Olarewaju (2013) where the study showed that comprehensive knowledge was low concerning testing to know one's haemoglobin genotype.

As revealed in the previous studies by Ndeezi et al., 2016 showed that in Uganda, recent estimates

Table 4. A frequency distribution table showing the hemoglobin levels.

Class (Hb g/dl)	Frequency(f)	Cumulative frequency(cf)	Lower limit	Upper limit
3.0 – 3.9	9	9	3.0	3.9
4.0 – 4.9	11	20	4.0	4.9
5.0 – 5.9	24	44	5.0	5.9
6.0 – 6.9	14	58	6.0	6.9
7.0 – 7.9	9	67	7.0	7.9
8.0 – 8.9	8	75	8.0	8.9
9.0 – 9.9	7	82	9.0	9.9
10.0 – 10.9	6	88	10.0	10.9
11.0 – 11.9	3	91	11.0	11.9
12.0 – 12.9	5	96	12.0	12.9
Total	96			

Source: Bio data 2021

place SCT prevalence at 13.3% and this illustrates the risk of having children with SCD If both partners have SCT and don't test before marriage.**Average haemoglobin levels among sicklers aged 1-15 years.**

Determining the average haemoglobin levels among the sicklers aged 1-15 years at LH was among the objectives of this study. Analysis and interpretation of data revealed that the minimum Hb concentration was 3.1 g/dl and the maximum Hb was 12.6 g/dl. The average Hb concentration was 6.8 g/dl with the modal Hb being 5.6 g/dl and the median of 6.2 g/dl. Children that had their hemoglobin concentration below 5g/dl were 20. According to age groups, 1 – 5 years of age were 13 (65%) and 6 – 10 years were 7 (35%).

The average Hb was 6.8 g/dl which agrees with the study findings (children's research,2021) that persons with SCD their Hb concentration ranges between 6 – 11 g/dl.

This study revealed the modal Hb concentration as 5.6 g/dl and 20 of these children had their Hb below 5g/dl with the 1-5 years age group most affected with 65% having the severe form of anemia that is life-threatening and this could be because of the sudden onset of complications among sicklers that cause RBC count to drop. This also agrees with the study carried out by Mudathir (2019) as it estimated that 75 – 85% of children born with SCD in Africa, the mortality rates for those under age 5 ranges from 50% to 80%.

Health supervision and management of sickle cell disease by the guardians/parents

The objective for this was to find out how effective the health supervision and management of the sicklers aged 1-15 years aged by their guardians/parents. Data analysis and interpretation revealed that 73(76.04%) never had proper guide tool books and were using exercise books for monitoring their children's medication and management not only but also most guardians 68(70.9%) that were residing in rural areas found it difficult accessing the health facility in times of crisis for proper management and these were the challenges identified. This is probably because of finical constraints among the parents and insufficient fund support from the government.

These findings agree with (Joylene,2015) as revealed that family medical homes are essential to the care of children, adults, and families affected by SCD as many don't have regular or convenient access to comprehensive SCD centers that support them. Also, Brous-seau in 2018 showed in his study emphasizes the importance of regular medical care.

Guardians expressed the need of using local drinks like hibiscus and beetroot as they helped to reduce the physical signs of anemia and this agrees with the study findings for the non-pharmacological management revealed by Ademola, 2015.

6 Conclusions

This study specifically sought to determine the prevalence, average hemoglobin level of sicklers aged 1-15 years of age, and the health supervision and management by their guardians. This study

established that the prevalence was 11.5% with the average Hb level being 6.8 g/dl and modal Hb of 5.6 g/dl out of the 96 children enrolled in the study. The health supervision and management of sicklers by their guardians were still poor.

Generally, there is still a big burden of SCD management among children aged 1-15 years at LH in the Luwero district.

Recommendations

Government of Uganda through the Ministry of Health

Set aside funds and other resources towards massive awareness creation encouraging premarital testing for SC among couples to curb the increase in the number of children with SCD and fill the huge information gap that exists concerning the matter.

Sickle cell clinic and health workers at LH in Luwero district

Create more awareness by having outreaches to the rural areas and educating them about SCD encouraging testing of couples at the facility free of charge as facilitated by the government. The facility should make a proper budget of the funds provided by the government and other supportive organizations and ensure that the guardians have proper guide tool books for the management and supervision of their children and proper follow-up of their medication and progress.

7 Limitation of the study

Limited finances for conducting the study in as far as transport, typing, printing and photocopying is concerned. However, the researcher got financial support from the parents and used it within affordable costs.

The unwillingness of some participants to release information due to fear. The researcher tried to explain the purpose of the study for beneficiary purposes in the future.

Time for the study to acquire the required information. However, this limitation was passed over with questionnaires that was time saving.

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Abbreviations and Acronyms

ASSC: Acute splenic sequestration crisis
Hb: Hemoglobin
HbA: Normal adult hemoglobin
HbS: Sickle hemoglobin
LH: Luwero Hospital
MOH: Ministry of Health
NHLBI: National Heart, Lung and blood institute
PROPS: Prophylactic oral penicillin
SCA: Sickle cell anemia
SCD: Sickle cell disease
SOPs: Standard operating procedures
VOC: Vaso-occlusive crisis

8.1

8.1.1 O: World Health Organization

RBC: Red blood cell
SC: Sickle cell
g/dl: Grams per deciliter

Operational definitions.

Anemia: This is the condition in which their reduction one lacks enough red blood cells to carry adequate oxygen to the body

ASSC: This is when sickled red blood cells get trapped in the spleen causing the spleen to enlarge.

Morbidity: This refers to having a disease or symptom of a disease or the amount of disease within a population.

Mortality: This refers to the number of deaths in a certain group of people or the death rate in a certain period.

Prevalence: This is the proportion of persons in a population who have a particular disease or attribute at a specified point in time or over a specified period

SCD: This is a group of disorders that affect hemoglobin molecule that delivers oxygen to all the cells throughout the body. The defect hemoglobin is called hemoglobin S which can distort red blood cells into a sickle or crescent shape.

VOC: These are painful situations that occur when the flow of blood is blocked to an area because the sickled-shaped cells have become stuck in the blood vessel most often in the legs, arms, and chest.

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Table 5. References:

1) Ademola Samson, Adewayin. Anemia 2015. Management of sickle cell disease. A review for physician education in Nigeria (sub Saharan Africa)https://doi.org/10.1155/2015/791498PMid:25667774 PMCid:PMC4312619

2) Assaf P. Oron, Dennis Choa. (2020) Caring for Africa's sickle cell children. Will we rise to the challenge? BMC medicine 18, Article number 92(2020).https://doi.org/10.1186/s12916-020-01557-2PMid:32340612 PMCid:PMC7187492

3) Broussea David c. Panepinls Julie A, 2018 Medical college of Wisconsin. Implementation of evidence based care for the acute treatment of sickle cell disease.

4) Children's research 2021, 262 Danny Thomas Place, Memphis TN 38105 2021. St Jude children's research Hospital. http://www.stjude.org

5) G Ndeezi, (2016) Burden of sickle cell trait and disease. Sickle cell surveillance study (US3): a cross sectional studyhttps://doi.org/10.1016/S2214-109X(15)00288-0

6) Gulendam karadag, Zeynep Gungarmus and Zeynep Olcar, (2018): Journal of caring sciences. Experiences and problems encountered by families of children with sickle cell anemiahttps://doi.org/10.151 71/jcs.2018.020PMid:30283756 PMCid:PMC6163154

7) Joylene John- Sowa, Barbara P Yawn, MD, MSc, George R. Buchanan, MD, Araba N. Afenyi - Annan et al (2014) Management of sickle cell disease. Summary of the 2014 evidence based report by expert panel

8) NHLBI (2022) What Is Sickle Cell Disease? https://www.nhlbi.nih.gov/health/sickle-cell-disease

9) Odunvbun. M.E, A.R. (2015). Newborn screening for sickle cell disease in Nigerian Hospital. Public Health 11 - 16.

10) Olarewaju Sunday Olakunle, Enwerem Kenneth, Adebimpe Wasu Olakekan, Olugbeng-Bello (2013), Pan Africa medical Journal 15 (1),

11) Rudan Igor, Wastnedge Elizabeth Waters, Donald Patel, Smruti Morrison Kathleen Goh Mei, (2018). JGH Journal of Global Health, J Glob Health 2047-29782047-2986 Edinburgh University Global Health society jogh-08-021103 10. 7189/jogh.08. 021103 Article. The global burden of sickle cell disease in children under five

12) Steinberg MH (2016). Sickle cell disease and other hemoglobinopathies. In L Goldman, A shafereds, Goldman- Cecil Medicine, 25th edition vol. 2

13) The global burden of sickle cell disease in children under five by E. Wastnedge, 2018