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Assessing Parental Knowledge on Sickle Cell Disease: A Phenomenological Study

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Abstract
Sickle cell disease (SCD) is a genetic disease affecting millions of people in the world, making the disease a global problem. It is an inherited blood disorder, resulting in morbidity and mortality among disease sufferers. Consequently, the incidence of Sickle Cell Disease in Ghana has increased dramatically over the last decade affecting 2% of newborns in the Kumasi Metropolis yearly. While the overwhelming majority of people born with the disease still come from Africa, least developed services exist for people living with the disease. Efforts to coordinate knowledge among health care givers and parental/ community care can significantly improve health and well-being of individuals with the disease. The purpose of this qualitative study was to explore parents' knowledge on SCD at Komfo Anokye Teaching Hospital (KATH). It involved 10 parents who were purposively sampled and interviewed from the Sickle Cell Clinic at KATH. Data was analysed using thematic content analysis**.** The study revealed that there is a gap in knowledge on Sickle cell disease and crisis and thus recommend the intensification of education as it plays a vital role in giving optimal care and preventing complications.
Keywords: Sickle cell disease, knowledge, education

1. Background

In 1910, Dr. James Herrick, a Chicago physician, was the first American to formally report and identify elongated, sickle shaped haemoglobin in an anaemic Grenadian student's blood smear. Herrick coined the now familiar term "sickle cell" (NIH, 2007). Sickle Cell Disease(SCD) affects millions of people throughout the world and it is found to be the most common blood disorder among families whose ancestors came from Sub Saharan Africa, South America, Cuba, Central America, Saudi Arabia, India and the Mediterranean Regions (Creary, Williamson, & Kulkarni, 2007).

Statistics from the Sickle Cell Disease Association of America (SCDAA) during the 41st Annual Convention in September 2013 shows that globally, 5,476,407 people have Sickle Cell Trait (AS) and 312,302 people have Sickle Cell Anaemia (SS). Sub Saharan Africa takes about 75.5% of the total number which represents 3,580,207 for Sickle Cell Trait (AS) and 235,681 have Sickle Cell Anaemia. In Ghana, in the year 2010, about 5,815 people had Sickle Cell Disease and there is an estimated 32.3% increase globally, 46.0% increase in the Sub-Sahara regions and 17.9% increase in Ghana by the year 2050 (Piel-Simon, et al., 2013).

Although a lot of seminars and workshops have been organized by the Kumasi Metropolitan Sickle Cell Unit in conjunction with the Sickle Cell Foundation of Ghana, to help reduce the increasing rate of the disease by putting in place measures such as newborn screening for sickle cell disease, tracking and referral of affected babies to commence comprehensive care at the KATH Clinic to promote the longevity of sufferers and also educate parents and students on SCD. It was therefore worth the attempt to determine Parents' knowledge on SCD and pain management to improve quality of life.

2. Methods

2.1 Study Design and Setting

The study was an exploratory study, which employed qualitative techniques in obtaining information from 10 selected parents/caregivers from October 2016 to April 2017 who met the following inclusion criteria; A primary caregiver of a child or children with SCD;

A caregiver with children between the ages of 2 to 18 years with regular attendanceat the SCD clinic for not less than two years;

A caregiver whose child was diagnosed through the new born screening for sickle cell disease program. The study was conducted at KATH popularly known as "Gee". The hospital was named in honour and memory of the legendary and powerful fetish priest Okomfo Anokye. It is located in Bantama, Kumasi in the Ashanti Region. It is bounded on the east by Bantama, Kejetia to the South, West by the 4BN of Infantry Uaddara Barracks and on the north by the Central Police Barracks. It houses the famous Komfo Anokye Sword. The hospital is a teaching hospital and the referral point for other health facilities in and outside the region has numerous facilities. The hospital is divided into clinical directorates which consists of Child Health, Diagnostic,

Anaesthesia and Intensive Care Unit, Dental, Eye, Ear, Nose and Throat, Medicine, Obstetrics and Gynaecology, Oncology, Polyclinic, Surgery, Accident and Emergency Department, Pharmacy and Physiotherapy and the Non Clinical directorate which comprises of Domestic Services, Security, Supply Chain Management, Research and Development and Technical services. The Hospital can boast of about 3615 staffs including medical officers, nurses, auxiliary staffs and paramedics.

2.2 Study Population and Sample

A total of 10 parents with children diagnosed with Sickle Cell Disease were enrolled and interviewed for the study using the purposive sampling technique. The inclusion criteria for selection were; being the primary caregiver of a Sickle Cell Disease child or children, caregivers with children between the ages of 2 to 18 years and have been attending the sickle cell disease clinic for not less than two years and was diagnosed through the new born screening for sickle cell disease program. All other mothers who did not meet these criteria were excluded from the study.

2.3 Data Collection and Analysis

A qualitative tool was used for measuring data collected. Phenomenological approach was the type of qualitative research method used in order to determine the respondents lived experiences. Indepth interviewing was used in data collection. A face to face interview with semi structured questions comprising of open ended questions was used to collect data. The interview guidelines were analysed and all necessary omissions and additions were made with the help of our supervisor to get rid of all ambiguity and repetitions.

Interviews were audio taped and field notes were taken during the interview. Participants' responses were later played over and over and data was transcribed verbatim. Data was collected on the 6th of February, 2017 from 7:30 am to 3:00 pm and each interview lasted for about 30 minutes. Recorders and all other documents used were kept under lock and key after. Thematic Content Analysis was used in analysing the data collected.

2.4 Ethical Consideration

Ethical clearance was sought from the Committee on Human Research, Publications and Ethics, School of Medical Sciences, KNUST. After ethical clearance was given, informed consent was obtained from each participant without coercion and the rationale for the study was explained to them. They were also informed that the interview will be audio taped to help the researchers to later produce a write up and present a verbatim transcription of data collected. Each participant was given a pseudonym as no real names were used in data reporting.

3. Results

The table shows that, 8 of the study respondents were females whiles 2 of the respondents were males. The age of respondents interviewed fell between the ages of 30 and 50 years however majority of the study respondents were between the ages of 30 and 35 years. Also, 7 of the respondents were married whereas 3 of them were widowed. A little above half of them had no knowledge on their status whereas, the remaining few were Sickle Cell Carriers (AS) or had Sickle Cell Anaemia (SS) and all respondents knew their children's Sickle cell status.

Table 1. Background Characteristics of Respondents

Variables	Frequency N=10	Percentage (%)
Sex		
- Male	2	20.0
- Female	8	80.0
Age, years		
- 30-35	6	60.0
- 36-41	2	20.0
- 42 and above	2	20.0
Marital Status		
- Married	7	70.0
- Widowed	3	30.0
Parity		
- 1	2	20.0
- 2	2	20.0
- 3	3	30.0
- 4	3	30.0
Number of Children Affected		
- 1	8	80.0
- 2	1	10.0
- 3	1	10.0
Genotype of Respondents Children (all children of respondents not equated to N)		
- AS	2	15.0
- SS	5	39.0
- SC	6	46.0
Educational Background		
- Primary	2	20.0
- Junior High School	3	40.0
- Senior High School	4	30.0
- Tertiary	1	10.0

3 of the respondents had parity of 3 and 4 respectively whereas a significant number of respondents (8) said they had only a child affected. Majority (4) of the respondents had attained their senior high school education.

3.1, Aetiology and Inheritance Pattern

In assessing parents' knowledge on the cause and pattern of inheritance, it was revealed that, a little over half (6) of the respondents understood Sickle Cell Disease as blood related disorder out of which only two (2) of the respondents were able to explain the inheritance pattern. A few of the respondents (2) could not identify Sickle Cell Disease as a blood disorder neither could they explain the inheritance pattern whereas the other respondents (2) could not identify Sickle cell disease as a blood disorder but were able to explain the inheritance pattern. All respondents had knowledge on sickle cell disease based on the genotype of their children; other types of sickle cell disease were unknown to them. All respondents interchanged sickling test for blood group test.

Madam R1O2 uttered;

It is a blood disorder which occurs when you and your husband are carriers or have sickle cell disease and you both pass the Haemoglobin S to your child.

Madam P1G1 added;

What I know about sickle cell disease is that, it is a blood disorder which occurs as a result of sexual intercourse between two individuals.

Mr. M1I9 commented;

I know it is a condition that is transmitted through the blood but I do not know how and why.

Madam T1A10 echoed;

I thought it occurred when you do not eat well during pregnancy but when I brought my daughter here, I was told that I have SC and my husband has AS that is why we gave birth to a child with SC.

Madam D1A6 added;

I only know of SS and AS because my child has SS and my husband and I have AS.

3.2 Prevention

On prevention of crisis and sickle cell disease. A few of the respondents (3) advised in undergoing genetic counselling and sickling test to ascertain their sickle cell status before getting married while the rest gave varying answers ranging from abstinence from conceiving, administration of drugs and managing the condition with the help of a health service provider and not getting married as methods of prevention. About half of them (4) believed the disease can neither be prevented nor cured but can only be managed.

Madam D1A6 supplied;

In preventing the condition, I will advise anyone who wants to marry to undergo "blood group" testing and if the results are the same (meaning both spouse have AS) then they should not marry. Thus if one is AS he or she will need a partner with AA so they can have a child with AS; which is at least normal.

Mr. M1I9 divulged;

The only way to prevent the condition is to avoid any blood relations such as blood transfusion.

Madam R1O2 also commented;

It can be prevented by protecting the child from insect bites, putting on clothing that will protect child from cold weather and managing their diet well especially with foods that will boost blood production because they frequently have anaemia. I also prevent them from taking cold water because I know that is what brings about the crisis as well as cold weather".

Madam F1A3 echoed;

I believe there is no cure for the condition but it can only be managed.

Madam P1G1 added;

The only way to prevent the condition is to not give birth.

Madam Y1A5 supplied;

The only thing that can be done is to manage the condition from deteriorating because regardless of what you do, it will occur.

3.3 Pain Episodes and Management

All 10 respondents confirmed that their children have experienced Sickle Cell Crisis. Although a few of them (3) knew their children had pain episodes, they were unaware of the term 'Crisis'. All the 10 respondents identified joint pains mostly in the hands and legs which sometimes are severe enough to cause temporary paralysis as a symptom of crisis. While a handful of them (4) also identified swollen hands, legs, periorbital oedema, fever, crying and jaundice as signs and symptoms of the crisis. Furthermore, most respondents (8) attributed the crisis to be triggered by low temperatures while a few others (2) attributed diet, medications, insect bites and stress or physical exhaustion as the precipitating factor for the crisis.

On management of the crisis, majority of the respondents admitted to immersing the affected part in warm water, after which they apply ointment and administer the drugs that were prescribed during the clinic visit. Only 2 of the respondents rushed the child to the hospital immediately the crisis begins. In the quest of finding out the treatment regimen received at the hospital, all of the respondents reported that the drugs; penicillin V, folic acid and zincovit are given but only a few (2) of the respondents knew why each drug was given. None of the respondents had heard about hydroxyurea. 4 of the respondents' children have been hospitalized but only 3 were hemotransfused.

Madam R1B4 lamented*;*

Mostly, my eldest child Philip complains of leg and hand pains and it sometimes severe that he finds it difficult to walk. It occurs when he takes in cold drinks, plays a lot or when the weather changes. She went on to say "*When it happens I immerse the leg in warm water and administer penicillin V and apply any other ointment at the affected part. He sometimes also complains of pains in the hands and I provide the same treatment.*

Mr. M1I9 added;

My child sometimes complains of hand and leg pains and there are times when her legs and hands become swollen. She also experiences periorbital edema. It mostly occurs when I bathed her with cold water or she takes in anything cold and this makes her cry a lot. When it occurs, I apply ointments to the affected part.

Madam R1O2 also commented;

Sickle cell crisis is when the child experiences joint pains, fever and yellowish coloration of the eyes (jaundice). When the child experiences the crisis, I give him the drugs that were prescribed during our previous clinic visit and if it still persists, I rush the child to the hospital.

Madam S1B7 added;

They have been giving him penicillin V and folic acid. The folic acid is given for energy but I don't know why penicillin V is given also. He has been administered once at KATH and twice at Tech Hospital. I have never heard of hydroxyurea either.

Mr. K1A8 supplied;

She is given injections whenever I bring her to the hospital and I am told it relieves her of the pains she goes

through and folic acid. No, I have not heard of hydroxyurea but I once heard there are other drugs that can help my child but are very expensive.

4. Discussion

In a study conducted by Treadwell et al., (2006) that surveyed 282 people from northern California about their exposure to and knowledge of sickle cell disease and sickle cell traits. Interestingly, 68% of those interviewed in this study responded correctly to knowledge questions about Sickle Cell Disease and only 15% were aware of their own trait status. The study conducted by Treadwell is in resonance with this study which revealed that a little above half of the respondents (6) had no knowledge on their status whereas, 3 were Sickle Cell Carriers (AS) and 1 had Sickle Cell Anaemia (SS).

This study revealed that only 30% of the respondents had knowledge on genetic counselling whereas 60% of the respondents had no knowledge on their status or family history. This is similar to the study conducted by Prabhakar (2009), which revealed that there is lack of knowledge regarding carrier status, family history and genetic counselling. Only 20% of this study's respondents understood the inheritance pattern of the disease and this corroborates with a study by Boyd et al., 2005 which concluded that out of the 162 women who met the eligibility criteria for the study, only 9.3% understood the inheritance pattern of Sickle Cell Disease, and 11% were unaware of their carrier status.

In a study by Marlowe and Chicella on the precipitating factors of Sickle Cell Crisis, it revealed that "unfortunately, the underlying mechanism responsible for the onset of pain episodes remains ambiguous to researchers and experts who study Sickle Cell Disease. Researchers believe that the onset of pain episodes can be provoked by changes in altitude, temperature, physical and emotional stress, menstruation, infection, fatigue, and dehydration" (Marlowe & Chicella, 2002). This study also reported that, most parents outlined low temperatures, diet, medications, insect bites, stress or physical exhaustion as provoking factors to pain onset. Dehydration and menstruation were not mentioned because most of the respondents had children below the ages of 10 and only a few of the children were females**.**

Although hydroxyurea was identified in a pilot study by the National Heart, Lung, and Blood Institute (NHLBI) with participants who ranged from 6 to 24 months of age that the drug is well tolerated in younger children, most respondents in this study reported immersing the affected part in warm water, applying ointment and administering penicillin V, folic acid and zincovit as the treatment regimen for Sickle Cell Crisis and had never received hydroxyurea.

A clinical trial study, which evaluated the efficacy of transfusions among patients with some Sickle Cell Disease complications, showed that repeated transfusions reduce the risk of recurrent strokes in children with Sickle Cell Disease. A study by Sebastiani, Ramoni, Nolan, Baldwin, & Steinberg in 2005 also showed that 50% of children with Sickle Cell Disease, who had suffered strokes and had not received transfusions, would suffer strokes within three years compared to 10% of Sickle Cell Disease individuals who had received transfusions. This is contradicted in the current study because only 3 of the children of the respondents have been hemotransfused and neither of them together with the remaining children who have never been hemotransfused have experienced stroke. And this can be attributed to the fact that only a few of the respondents sent their children to the hospital when they experienced crisis. Hydroxyurea and Transfusion were not widely used according to the study.

4.1 Limitations

Results of the study will not be generalizable because participants were all from only one clinic.

5. Conclusion

In an illness like sickle cell disease, parental understanding of the disease and its causes has both physical and psychological value. At a physical level, knowledge can be crucial for optimal self-management of the disease in order to reduce long term complications, while psychologically it can help improve psychosocial adjustment and adaptive coping. But, while knowledge can be beneficial, it can be equally detrimental, especially when the information is incorrect. The study revealed a major gap in knowledge on Sickle Cell Disease, its etiologic, inheritance pattern, respondents and children's status, cause, signs and symptoms, management and prevention of the condition. Less than half of the respondents were able to correctly respond to most of the questions asked on Knowledge on Sickle Cell Disease and Sickle Cell Crisis.

Competing Interest

The authors affirm that they have no competing interest.

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Author's Contribution

The study was conceived and planned by all authors. ADA and EJEN collected the data under the supervision of AAS. All authors were involved in the analysis of data and interpretation of study findings. ADA and AAS wrote the first draft of the manuscript. Review and revision of the manuscript was critically done by all authors.

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