Case report

FRUSTRATIONS OF PARENTS/GUARDIANS OF CHILDREN LIVING WITH SICKLE CELL DISEASE; CASE REPORTS.

Comment [P1]: Please specify the study
This is a case report or case series??

ABSTRACT

Aim: To bring to light the frustrations and problems parents/guardians of children living with Sickle Cell Disease go through.

Comment [P2]: Please reframe the aim of manuscript
Use another word instead of to light

Presentation of Reports: We present series of reports of parents and guardians whose children are less than fourteen (14) years of age and have sickle cell disease (SCD). In two of such reports the parents wished that their wards were dead to relieve them (the parents) of the challenges they go through to take care of the children. In the third report, the husband divorced the wife, blaming her for the plight of their two children; and the final one presents a couple who contemplated withdrawing their wards from school, so they would not have to spend money to educate them and lose them eventually to death.

Comment [P3]: Specify the study
If you are describing more than one report than this study is Case series

Discussion: The reports give clear indications of the challenges guardians with children living with SCD go through. The disease which is hereditary affects the haemoglobin of the red blood cells, making the cells sickle-shaped. The clinical hallmarks of frequent infection, bone-pain crisis, anaemia, fatigue, organ failure and many other symptoms associated with the condition lead to frequent hospitalization. This is where the frustrations of the parents come from. With proper management of the condition and education to the guardians, SCD patients can go through normal life and the guardians can also have relative peace of mind to support such children.

Conclusion: Individuals who are carriers of the sickle cell traits should be discouraged from getting married to other carriers; and if they do, they should be cautioned about the challenges they stand to face if they decide to have children. By doing this, perpetuation of this condition and its attendant problems may be reduced. Newborn babies should be tested for the condition and proper management given from the onset to minimize crisis.

Keywords: Sickle cell disease, red blood cells, bone-pain crisis, haemoglobin.

1.0 INTRODUCTION

Sickle cell disease (SCD) is a disturbing condition that comes about as a result of inherited recessive gene in the haemoglobin of the red blood cells (RBC). Substitution of valine (an amino acid) for glutamic acid at position six on the beta globin chain of the haemoglobin (the component of the RBC that carries oxygen from the lungs to other body tissues) is the cause of this genetic abnormality (1). The normal RBCs are spherical in shape; however, in the case of people with SCD the red cells are shaped like crescents or sickles (hence the name sickle cell).

The condition is characterized by frequent infection, bone-pain crisis, anaemia, fatigue, organ failure and many more. Severity of signs and symptoms varies from individual to individual.

The disease is carried as an *autosomal recessive trait*; meaning that two copies of the abnormal gene (one from the father and the other from the mother) must be present for manifestation of the condition. A person who inherits haemoglobin S from one parent and normal haemoglobin A from the other parent will have the *sickle cell trait*, and such an individual usually will have no symptoms of SCD.

Comment [P4]: Please add burden of disease

In instances where there is frequent bone-pain crisis and infections, sufferers, especially children are often hospitalized, and this can leave parents devastated. In most parts of Africa where guardians or individuals are left to bear the full cost of treatment and medication for all diseases, people with low income and/or large families are left distressed when their scarce resources are channeled to secure the well-being of their sick child(ren). This has prompted some parents to wish that their children suffering from SCD were dead so that they (parents) could have 'relief' from the burden of having to care for and spend their little resources on constant hospitalization of the sick.

Many disease conditions are not properly managed if the affected persons cannot pay for the cost of treatment. This somehow accounts for the large dependence of many people in sub-urban and rural areas of Africa depending on traditional or herbal remedies for treatment and management of various diseases, including SCD. The disease is managed with various interventions at the hospitals and in the homes.

Comment [P5]: Write name of some interventions

These case studies throw light on the frustrations of some families with children suffering from SCD and highlights the need to educate people on how the disease could be eradicated or lowered drastically.

2.0 CASE PRESENTATION

2.1 Report One

It was a fine Tuesday morning around the hour of 11.15GMT at a major Teaching and Referral Hospital in Ghana. A middle-aged woman around the age of 38years was angry with *herself*, walking briskly and talking audibly to no one in particular, and making gestures. She was

oblivious of the environment and surrounding people. One could see worry and misery around her, but somehow, everyone was busy going about their own activities.

The young woman was confronted gently and asked what the problem was. When she eventually calmed down she lamented that her daughter had been hospitalized for the third time in two months, and coldly stated, without any sign of remorse, that she wished that she (the daughter) would die for her (the mother) to have her *peace* of mind. This sounded a bit strange; for a biological mother to wish her daughter dead. She was assured of assistance to help her overcome whatever challenges she was going through, and encouraged to explain further what she meant by her statement.

Her issue was that she and her husband had five children; they had however lost two of them to SCD. The previous deaths came through situations similar to what their daughter was currently going through. They would spend all their resources and still lose the child. They were fed-up with the recurrent losses, and so she preferred that their daughter would just die at that moment, however she knew this would not happen. They would borrow money from friends and relatives for treatment but eventually the child would die and they (the parents) would have to struggle to pay back their debt.

The woman was counselled but no amount of empathy, encouragement, assurances, would change her wish, especially as she had buried two children already. Upon further assurances that many have survived similar crisis, she reluctantly accepted and seemed a bit regretful for her outburst. This was a clear case of frustration, hopelessness and dejection.

2.2 Report Two

In a similar presentation to the first report, a woman brought her thirteen-year old daughter to a pharmacy for her routine medication. In the full glare of the daughter, other clients and the staff of the pharmacy, the woman said she had been spending so much of her resources to manage the SCD the child was suffering from but there had not been much improvement. Unfortunately, the daughter too would not die and end it all. Upon hearing this the child started weeping bitterly, but her mother felt no remorse for her utterance. She kept repeating them, and when she was even cautioned, she boldly stated that she meant every word. The woman was separated briefly

from the daughter and counselled by well-meaning people. At this juncture, she poured out her frustrations and the challenges she had been through and continued to go through because of the recurrent episodes of sickle cell crisis her daughter experienced.

According to her, she lost her job because of frequent absenteeism from work. She asked, '*how do you think I would wish that my only child should die if not because I have been stretched beyond my limit? Look at her stature at thirteen years of age, there is absolutely no hope. She cannot make it in life and the earlier she departs this world (dies), the better.*' She continued to say that she had lost interest in giving birth. Asked if that was how she had been treating the daughter at home, she said no, though she had harboured that wish (of death for her) for a long while.

Upon education and reference to many instances where people have lived normal life for long even after initial struggles with the disease, she smiled amidst her tears, and asked if there was any hope for the daughter. When she was answered in the affirmative, she seemed relieved.

The daughter who was so distressed was also called and counselled and encouraged that she was not going to die; she only needed to take her medications, eat properly and follow hospital appointments judiciously. Asked whether the mother had ever mentioned the death wish before, she answered in the negative. She was assured that the mother loved her, and it was out of frustration that she said what she did, so she (the daughter) should not habour bitterness against her mother.

The mother openly apologized to the daughter when she was brought to her, and they reconciled, after she (the daughter) professed her forgiveness. The required medication was dispensed, and then the mother and daughter left the pharmacy.

2.3 Report Three

A young woman was left devastated following a divorce from her husband, who accused her of being responsible for the plight of their two children, both of whom suffered from SCD. Following the divorce, the man left for an unknown destination leaving the woman alone to care for the children. Unable to sustain the pressure of care alone, the woman sank into depression and vented her anger on the two children aged nine and six years respectively. It was the timely intervention of a psychologist that partially saved the situation. Even with the intervention, the

burden of constant visit to the hospital for management of the disease made her wish she had not given birth at all.

2.4 Report Four

Two of the three children of a couple have been diagnosed with SCD. The affected children both girls, aged eleven and nine years have had frequency crisis from the disease. According to the mother, there were instances the teacher of one of the children had asked her not to allow the girl to come to school for a period of time so she could recover. The mother obliged, believing that it was in the best interest of the daughter, but it negatively affected the child's academic performance. The situation was exacerbated by the *misconception* that most SCD patients do not grow beyond the teen-age. According to the mother she succeeded in convincing her husband to withdraw the affected children from school and concentrate on the third who was healthy. As a result, the affected children stayed at home for almost a whole academic year.

It was when support and education came from friends and relatives that the couple reluctantly enrolled them back to school.

3.0 DISCUSSION

SCD is a genetic disorder in which the RBC contain an abnormal form of the oxygen-carrying protein, haemoglobin S (HbS). The abnormally shaped haemoglobin makes them form clumps as they stick together in blood vessels, thereby blocking or reducing the flow of circulating blood through the body tissues. When such genes are inherited from both parents (i.e. homozygotes), the individual develops SCD. When one inherits the gene from only one of the parents the person will have the sickle-cell trait (SCT), becoming a 'carrier' without exhibiting the symptoms associated with SCD. The condition is relatively common in Africa, especially sub-Saharan Africa where it is estimated that about 80% of the annual birth of SCD patients are found (2).

SCD presents many symptoms that may start as early as a few months from birth. The major ones which are excruciating pains, infections and anaemia, normally result in what is termed the 'sickle cell crisis'. Several acute conditions such as vaso-occlusive or acute painful crisis, aplastic crisis, splenic sequestration crisis, hyper-haemolytic crisis, hepatic crisis, dactylitis, acute chest syndrome are associated with the sickle cell crisis. Other acute complications include

pneumonia, meningitis, sepsis and osteomyelitis, stroke, avascular necrosis, priapism, and venous thromboembolism (3).

The episode of pain is one of the distressing symptoms of SCD which results from blockade of blood vessels to some parts of the body such as the limbs, pelvis, stomach, ribs and sternum. On the average the pain can persist for about a week. Painful incidents can be triggered by dehydration, stress and *unfavourable* weather. Further, SCD patients are prone to several infections, from mild ones (such as common cold) to more serious one such as pneumonia and meningitis. A drop in the number of RBCs leads to anaemia which can, in itself, cause other symptoms such as rapid heartbeat, headaches, dizziness and sometimes fainting.

There are many social aspects of the SCD for affected persons and their families. Social life, education, employment prospects are all affected in one way or the other. Such situations are worsened by general lack of understanding in the healthcare, education and social care systems (4). Children with SCD may not have the support of their teachers or care-givers; they are sometimes made to feel that they are the cause of their conditions. If care-givers and teachers could understand, for example, that such children need to take in a lot of fluid (water) to keep them hydrated and require less exercise than others, they can encourage affected children to observe same. And if it happens that because of keeping hydrated children must get out of class often to urinate, a teacher should not embarrass them. Teachers should also understand that SCD children may not always be able to attend school; their absenteeism may increase because of frequent hospital visitations, hospital admissions and hospital re-admissions.

Treatment of SCD has always been a complex one. The goal of therapy has been to control the symptoms and manage complications associated with the disease. Maakaron (2020) has indicated some management and preventive strategies to control the disease (5). These include management of vaso-occlusive crisis, chronic pain syndrome, chronic haemolytic anaemia, as well as management of the complications and the various organ damage syndromes associated with the disease. Additionally, prevention and treatment of infections, prevention of stroke, and detection and treatment of pulmonary hypertension must be encouraged as other strategies to help control the disease.

Clinical actions required include enhancement of preventive care, management of common acute and chronic complications of the disease; and initiation and monitoring of the two SCD-specific disease modifying therapies of hydroxyurea and chronic blood transfusion therapy (6).

In Africa, and elsewhere where there is heavy reliance on herbal treatment for management of many diseases, some people have resorted to the use of some herbal remedies in the management of SCD. Medicinal plants such as *Carica papaya* (7) and *Fagara zanthoxyloides* (8) have been used to manage SCD in traditional settings.

Frustrations that parents and/or guardians of children living with SCD go through emanate mostly from frequent hospital admissions and re-admissions, cost of management of the disease. When such children are sick or admitted at the hospital, the guardian may have to take time off work for catering purposes; and this may affect the financial position of the individual as they may not earn enough money due to absence from work. Where the caregiver even goes to work when the ward is sick, they may be late, and may want to leave early from work. Additionally, the parents may not have stable minds to work productively. On the part of the affected child frequent absenteeism from school may affect academic performance, and this may in turn affect the parent emotionally. Families having children with SCD may go through challenges such as anxiety, divorce and emotional disturbances; and they may show less commitment to social activities in the community (9). Furthermore, there is demonstrated psychological and social disturbances among sufferers of SCD and their families (10).

Complete management of the disease therefore requires that it is looked at holistically from social, psychological, behavioural and medical perspectives. Sufferer of the disease must as much as possible keep calm and stay in optimum *temperatures*, as extremes of temperature may precipitate crisis. Unnecessary disputes and arguments with people who may not understand what they (the sufferers) are going through should not be encouraged. The population as a whole need to understand sickle cell disease so that they can empathize with sufferers and their caregivers. And there must be proper counselling of potential couples who are carriers of the sickle cell traits. If individuals know their sickling status and are properly informed in terms of education on SCD, carriers would be careful in the choice of potential partners who are also carriers of the disease. Discouraging marriage between potential couples who are carriers of the

disease remains the surest way of eliminating or reducing the incidence of sickle cell disease in the population.

4.0 CONCLUSION

Sickle cell disease is a hereditary condition, and individuals who suffer from it go through a lot of pain and psychological trauma. Guardians of children with the disease may go through a lot of financial difficulties, and may suffer from loss of productive working hours, social and emotional torture. They sometimes wish that their affected children are dead.

As the disease is hereditary, arising from homologous recessive genes, the surest way of preventing the condition is that, carriers of the sickle cell traits should be discouraged from getting married to another carrier; and if they do, they should be aware of the potential problems their children are likely to have if they give birth.

Newborn babies should be screened for the condition and proper management given from onset to minimize crisis. By doing this, guardians of affected children may have a little respite.

CONSENT

The guardians used in the case reports consented to the interactions being used for academic purposes.

ETHICAL APPROVAL

Not applicable

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