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ORIGINAL ARTICLE

Premarital genetic screening and care of Tanzanian children with sickle cell disease: a qualitative study on parents' views and experiences

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Abstract

Genetic testing and counselling is one of the approaches to reduce the high birth rate of individuals with sickle cell disease (SCD). A descriptive phenomenological approach was used to explore parents' views on premarital genetic screening for sickle cell trait and their experiences in the care of Tanzanian children with SCD using a face-to-face in-depth interview. The study was conducted at sickle cell clinic at tertiary hospital in Dar es Salaam region between June and August 2020. The study found that most of the parents with SCD children knew about genetic testing and counselling after the diagnosis of their children's SCD status. Major approaches employed in managing SCD crises were supportive, preventive, and symptomatic. Parents expressed a heavy burden related to caretaking due to the lack of financial support and stigmatization. These affected their children's quality of care and management. In conclusion, participants expressed a preference for premarital genetic testing, where others insisted it becomes compulsory. In addition, there should be an adequate clinic for early screening, accessible therapeutic support and long-term follow up for children with SCD. Support to poor families with individual with SCD through national health assurance scheme and free provision of preventive medications such as hydroxyurea, is recommended.

Keywords Sickle cell disease · Premarital screening · Dar es Salaam · Children

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Introduction

Sickle cell disease is am inherited condition, caused by a mutation in the β globin gene that affects haemoglobin, the molecule in red blood cells that delivers oxygen to cells throughout the body Brousse et al. 2014. Prevention of the disease through carrier identification and genetic counselling is one of the approaches to reduce the impact of the disease in the low-income countries including Tanzania where the condition is most prevalent Emmanuel et al., 2015.

In Cyprus, Greece and Italy, screening for thalassemia is standard practice where most at-risk couples are identified and offered early diagnosis in the first pregnancy; service majority use and produce healthy offspring Board (2005). Cyprus made premarital screening compulsory by law in 1980 followed by compulsory prenatal diagnosis in 1984. Birth rates showed a sharp decrease of one in every 2–3 years, in contrast to an average of 18–20 cases per year before the implementation of the "thalassaemia prevention programme"Bozkurt 2007.

This indicates the effectiveness of compulsory premarital screening and counselling as a prevention mechanism for genetic diseases. Premarital screening programmes have become widely accepted and highly valued in preventive health care so much that many countries have made them compulsory Rahman et al. 2015.

In Tanzania, the number of individuals with genetic disorders is high with sickle cell disease (SCD) having an estimated birth prevalence of 7/1000 resulting in a high disease burden Makani et al., 2011. Children with SCD require frequent visits to the hospital for routine medical care and management of complications. Treatment available to manage SCD is usually symptomatic and preventive rather than curative Ali and Razeq 2017. Premarital testing for human immunodeficiency virus (HIV) has allowed married individuals to make informed decisions which have contributed to decreasing the disease burden in Tanzania UNAIDS 2018. Couples who undergo testing can be informed about genetic risks and provided options for reducing risk, including prenatal diagnosis Rahman et al. 2015.

This study explored the views of SCD parents towards compulsory premarital SCD testing and their experience in the care and management of their children with SCD in Tanzania.

Materials and methods

Study design, area and period

A descriptive phenomenological approach was used to explore parents' views on premarital genetic screening for sickle cell trait and their lived experiences in raising a SCD child (27). The study was conducted in Dar es Salaam region between June and August 2020. Participants were selected from the paediatric sickle cell clinic at Muhimbili National Hospital (MNH). MNH is a National Referral Hospital and University Teaching Hospital with 1,500-bed facility, attending 1,000 to 1,200 outpatients per day, admitting 1,000 to 1,200 inpatients per week. MNH conducts weekly sickle cell clinics.

Study population

The study targeted parents of SCD children who were receiving care at the sickle cell clinic in the paediatric department at MNH. The study included biological parents with the age above of 18 years caring for a child with SCD whose children were hospitalized at the sickle cell clinic at the time the study was conducted.

Sample size and sampling technique

The study participants were purposively selected based on fulfilment of eligibility criteria and have the ability to provide information useful to the study (28). Ten biological parents of children with SCD were recruited into the study. This sample size was reached after attaining the saturation point during data collection.

Data collection method

A face-to-face in-depth interview was the selected method for data collection. A total of 10 participants were interviewed by two members of the study team, with each interview taking 15–20 min. This rationale for this number stemmed from the goal that the interviews should include enough participants to yield diversity in the information provided until the point of data saturation was reached. After each interview, data gathered were transcribed to allow the researcher to refine the interview guide.

Data collection procedure

An interview guide was developed based on an extensive review of the literature and objectives of the study Brousse et al. 2014, Emmanuel et al., 2015, Rahman et al. 2015. It included demographic data and open-ended questions to allow considerable probing and flexibility, allowing respondents to talk and freely express their feelings and perceptions towards the issue under study. Participants were identified by ID codes to maintain anonymity during the analysis and presentation of findings.

The study aimed to explore the views of parents towards compulsory pre-marital genetic testing and experience of therapeutic management of their children with SCD. Specific objectives were as follows: (i) to explore the views towards compulsory pre-marital genetic testing and counselling for SCD among parents of SCD children in Dar es Salaam, (ii) to describe the experience of therapeutic management among parents of SCD children in Dar es Salaam and (iii) to explore the preferred method of intervention for SCD among parents of children with SCD in Dar es Salaam. The interviews were conducted at the clinic in a consultation room for the convenience of the parents whose children were hospitalized. The interviews were conducted between June and August of 2020.

The participants discussed three questions developed from the research questions. The language used to conduct the interview was Kiswahili (National Language) as all patients could freely express themselves in Kiswahili. The interviews took 10–15 min each and were audiotaped with permission from participants. Data was gathered over a period of 3 weeks.

Data management

Data gathered was transcribed verbatim, translated into English and then back-translated and compared to notes recorded to ensure the trustworthiness of the data. All names were removed from the transcript so that participants were not identified. Pseudonyms were given to the participants based on their sex and numerical entry into the study such as FP1 whereby 'F' stood for female, 'P' for parent and '1' for the first parent interviewed. Similarly, MP3 implied the third parent interviewed who was a male participant. Soft copies of the transcribed data were safely stored in a computer.

Data analysis

Raw data gathered from the field was analysed immediately, following the interview. Data gathered from participants were transcribed verbatim and analysed using thematic analysis Lopez and Whitehead 2013. Repeated active reading assisted in familiarization and identification of patterns and meanings embedded in the data, noting down initial ideas. Fundamental segments that were interesting in the data were outlined systematically and used to develop initial codes, collecting specific aspects of the data that was related to it. Using tables, all the relevant codes were collated and merged into broader overarching themes.

The themes were defined and clarified to achieve coherence and to identify the story in the raw data that the theme conveyed. This brought to light the essence of the theme in relation to the raw data. The story within and across the themes was used to produce a report by analysing the selected extract, which was clear and compelling, in relation to the research questions. Analysis was done by two members of the study team who did not conduct the actual interviews. In the event of discrepancies, discussion between the researchers and verification with interviewers took place until consensus was reached. Data analysis was done using NVivo software (TETRA Insights, USA).

Trustworthiness was ensured by thick description of the findings so as can be applicable to other contexts, circumstances and situations. The descriptions of the findings were based on participants' responses. Lastly, participants' feedback was collected after reviewing the qualitative data to verify if their response was captured and filled in any gaps.

Results

Demographic characteristics of the participants

A total of ten biological parents, who were the biological parents of the SCD children, took part in the study. The participants included seven mothers and three fathers who were Tanzanians and, at the time of the interview, lived in Dar es Salaam. The ages of the parents ranged from 29 to 56 years with four of them below 40 years and six above 40 years. All ten of the parents were married and living with their spouses.

The parent's educational level varied; two of the parents had tertiary education, three had completed secondary school, three had secondary school education and two were basic school leavers. Furthermore, three of the parents interviewed were self-employed entrepreneurs, three were office employees, one was a peasant, one had retired and two were unemployed. In terms of age of child's sickle cell diagnosis, three of the ten SCD children were diagnosed when under 1 year old, two were 2 years old, three were 3 years old and two were 4 years old. All the participants as well as their spouses were sickle cell carriers and did not have SCD (Table 1).

Theme organization

In general, three major themes emerged from the subjective recount of parents' experiences. All themes, that is views on compulsory premarital testing, the experience of raising a child with SCD and preferred SCD intervention, emerged contextually from the data gathered and were in line with the objectives of the study. In all eight subthemes were identified. All the themes and subthemes are described in the ensuing sections supported with specific quotes from the participants (Table 2).

Table 1 Social demographic data of the participants

Variable	Category	Frequency
Age	21–30	2
	31–40	2
	41–50	4
	51–60	2
Gender	Female	7
	Male	3
Relationship status	Married	10
Employment Status	Employed	3
	Unemployed	3
	Self-employed	4
Education Level	Primary Education	2
	Some Secondary education	3
	Complete Secondary education	3
	Tertiary Education	2
Age of child's diagnosis	Birth–1 year	3
	1–3 years	5
	3–5 years	2

Table 2 Table showing the themes and subthemes arising from thematic content analysis of the data collected

Themes	Subthemes
Premarital testing and counselling for sickle cell trait in parents	• Perceptions of SCD • Awareness of SCD premarital genetic testing • Views on compulsory premarital testing
Experience in raising a child with SCD	• Stigmatization • Finances • Home management • Hospital management
Preferred method of intervention	• Premarital testing and therapeutic management

Views on compulsory premarital testing

Perceptions of SCD The parents of SCD children perceived SCD differently. For most, it was until when their children experienced a crisis that they heard about sickle cell and, even then, had to visit a variety of health facilities before getting a diagnosis and education on the disease. One mother recall:

I first heard of sickle cell when my child was diagnosed. Even on diagnosis I did not fully understand what sickle cell is until when my child started attending the Paediatric sickle cell clinic at Muhimbili National Hospital… Sickle cell is a lifelong problem and is inherited. Both my partner and I are sickle cell carriers. I also understood that we were troubled at first because we were uninformed. Sickle cell is not a communicable disease. I'm not sure if my child got sickle cell from my husband or me, but I've been told that it could have been from grandparents who had the disease on both sides of the family. My husband and I have never been tested. All my other children are sickle cell free. **FP1, age: 41 years.**

One parent believed that sickle cell disease was hereditary and passed on from one or both parents.

Sickle cell as far as I know in Kiswahili, they say it is Selimundu, which is caused by heredity from one or both parents. I don't think it is communicable. **MP3, age: 32 years.**

Another parent narrated on how she lost her son due to limited knowledge on SCD:

My older child had sickle cell disease but died. At the time I did not know the proper care for a child with the disease. I did not insist on him taking medication such as the folic acid. In Iringa, when they discovered that he had sickle cell disease, they gave us medication and sent us home. When the medication was over, I saw no importance of getting more medication because I did not know they were crucial. When crises began again, I could not manage them and eventually he died. **FP2, age: 42 years.**

From the parents' narratives, it was found that their perceptions about SCD before them receiving education at clinics were influenced by community beliefs about SCD patients. Some perceptions were associated with witchcraft and others with HIV/AIDS. One mother narrated her experience with her community as follows:

Most areas do not understand what sickle cell disease is. They categorize sickle cell patients with HIV/AIDS patients. My parents in law did not understand what sickle cell disease is and categorized the child as HIV/AIDS infected. This made them reluctant to assist me with childcare. This is also the understanding of many in my village which lead to my child being stigmatized. Care in small health facilities is almost none as they only help with a case of Fever. **FP9, age: 35 years.**

Another mother explained:

People around me are not as aware. You find that on telling someone my child has sickle cell, they ask you whose side of the family has sickle cell. So, they assume that sickle cell can only come from one side, but it is something that I, as an educated person, understand can come from both sides. **FP5, age: 42 years.**

Awareness of premarital genetic testing

All participants reported that counselling after diagnosis of the child's sickle cell status was their main source of information on genetic testing.

My husband and I have never been tested. I did not know about testing until I came for hospital care for my other child. That is when we were educated on the importance of premarital genetic testing… Genetic testing is important because it makes you aware of your status. I prefer it be premarital so that everyone knows what they are getting into it. **FP2: 42 years.**

One parent mentioned finding about genetic testing when looking for a resolution to a conflict between her and her husband on the actual source of their dead child's sickle cell gene.

> My second child died of sickle cell disease. At first, my husband did not believe that the disease was partly because he contributed to the sickle cell gene. He denied being the real father of the baby. To reconcile we went to Muhimbili National Hospital for consultation. We were tested for sickle cell disease and found out we were both carriers. We have a third child who is a carrier, a fourth child who is normal and a fifth child who has sickle cell disease. **FP5, age: 42 years**

Compulsory premarital genetic testing A consistent theme was that participants had witnessed crises experienced by their children. These experiences created a strong desire to look after their children's future health as well as hope other children do not get to experience these crises, with premarital genetic testing viewed as a key mechanism of prevention. They supported the premarital genetic testing of partners who planned on having families, asserting that preventative measures would protect parents from the challenges of caregiving for SCD children and protect the child's future health should carrier parents choose to have children after testing.

Seeing the challenges my child is facing, I would prefer that the government make premarital genetic testing available and compulsory. These children suffer terribly. When my child faces a crisis, it is so intense that I worry if she will make it to the next day. I would prefer that the government took initiatives to make it compulsory. **FP1, age: 41 years** .

The one half of the participants did not think it was appropriate to make premarital testing compulsory rather if appropriately educated people would choose to go for testing on their own. One father explained as follows:

It should not be compulsory. People should be educated enough to understand the implications their traits may have on their children. I would get tested if I knew then what I know now. But others would be reluctant to get tested by force for the fear of stigmatization. I think, if properly educated on the disease and its implications, many would voluntarily get tested. But some people are just stubborn, so maybe it could be emphasized before marriage. **MP10, age: 50 years.**

Experience raising a child with SCD

Crisis self-management The parents indicated that the major challenge on them concerning their children's health is when their children go into crises. All the parents expressed their desire of having a crises-free life for their children. They disclosed that seeing their children in such pain regularly made them feel they terrible. This experience influenced the opinion of the parents towards premarital genetic testing.

On how they managed their children's crises at home, most parents mentioned the use of analgesics, ibuprofen and acetaminophen, for the management of pain. One mother remarked:

At home my child is sick with joint swelling, I give him fluids and massage him with a towel soaked in warm water. For the pain, I give him analgesics usually Panadol… **FP5, age: 42 years.**

Another mother shared:

> When my child experiences anaemia she experiences joint pain. I try to give her vegetables, dates and roselle fruit juice that boost blood production and other blood food supplements. **FP6 age: 32 years**

Several parents mentioned only managing mild pain at home. Any other complication was to be dealt with at the hospital. One mother remarked:

When he has problems such as pain, we were suddenly advised not to stay with him and as often as possible to take him to the nearest hospital so that he can get immediate treatment… If it's a mild fever I can give him pain medication. But when he feels severe pain and I have noticed swelling on joints of his body, I often take him to the hospital for his safety. *FP4, age: 48 years.*

Hospital care Most parents complained of the difficulty in diagnosing SCD in children due to the limited facilities.

Discovering the child's problem was very difficult. We went to many hospitals; including Mwananyamala Regional Referral Hospital, twice, before being referred to Muhimbili National Hospital where they conducted intensive investigations to see what was bothering my child and discovered it was sickle cell disease. So, after the doctors found out that the child was suffering from sickle cell disease, we were given a brief seminar on the disease, and they began the child's treatment. **FP4, age: 48 years** .

Severe pain from the vaso-occlusive crisis, anaemia and joint pains and swelling of the limbs were some of the commonly mentioned complications that children took to the hospital faced. All parents disclosed that their children were managed on penicillin, folic acid and hydroxyurea.

If there is a problem and it happens that the child is sick with a high fever, the joints are swollen and sore, we immediately go to the health centre to get care. We only give

prescribed medication to our child. Usually, it is penicillin V, folic acid, hydroxyurea and pain medication which I was told is for preventing severe pain and anaemia. **MP3, age: 32 years.**

Most parents narrated similar methods of intervention for crises and pain.

My son is currently taking hydroxyurea as well as folic acid which he is given at the hospital every time he goes for treatment… If it is found that he has a certain problem, perhaps tested for malaria or any other problem, he takes the appropriate medication but the medication he is currently taking, even in his normal state, is the two types of drugs I mentioned. **FP4, age: 48 years.**

Some children suffer from anaemia frequently and require blood transfusions.

My son, after discovering he has sickle cell, the first year was not a big deal. But during the second year, there was a lot of trouble. He was hospitalized in September and received blood transfusions, and again in October. He was also hospitalized in November for yet another transfusion. In December, we rested but January required another transfusion again. So, we do not rest at home. **FP5, age: 42 years.**

Most of the parents were content with the attitudes of the doctors, nurses at the outpatient department and pharmacists. However, some complained about the attitude of nurses at the inpatient department:

Some nurses are not very supportive, and it is discouraging. My son may have anaemia, haemoglobin level 3.2 g/dL, and the nurse would treat it like it is normal and not an urgent situation. Or they may tell you that the children do not grow up to healthy individuals which I find to be very discouraging. **FP9, age: 35 years.**

Some parents complained of long waiting time during routine care at the outpatient paediatric sickle cell clinic and long queues and congestion at the laboratory as a laboratory test was required on each visit to the Sickle Cell Clinic and they often have to wait for hours in a queue.

The government should help in making life easier for us because when we go to the hospital, we spend a lot of time, the cycles and procedures become more frequent in the hospital where we may have to spend more than half a workday since the clinic is on a weekday. **MP3, age: 32 years.**

Financial burden The unpredictable nature of crisis occurrence prevented parents from carrying out planned activities. Frequent stay at the hospital prevented some mothers from attending work fulltime or taking their children to school. One mother described her experience as follows:

With this challenge, I cannot work for somebody. I try to work for myself, but my kiosk (small shop) is often closed, and I lose customers because they go elsewhere. The moment I want to start afresh, the child will fall sick, and I have to massage his joints or take him to the hospital. So, we have to depend on my husband's income which sometimes is not enough. **FP9, age: 32 years.**

Describing their experiences, most parents were grateful that the government provided health insurance as the major factor that would have burdened them was the cost of medicine and laboratory services. However, some still complained about the insurance services not being adequate and providing incomplete financial support.

My insurance did not cover hydroxyurea for the first year which required off pocket expenditure where I was spending about 1500/= Tshs per tablet. When you multiply this to the amount required for the child, it becomes very expensive! So sometimes, my son went without medicine for some weeks. **FP5, age: 42 years.**

Others complained that some of the prescribed medication was not covered by insurance and discouraged pharmacological equivalents were given instead.

My son usually used Panadol when in pain, but I was advised by the doctor to shift to ibuprofen. However, ibuprofen is no longer covered for by National health insurance so I have to buy it with my own money and when I cannot, due to other health expenditures, I use the Panadol they give us. **MP10, age:50 years** .

Most parents pleaded with the government to receive more consideration in the National Health Insurance Scheme.

I am very grateful that the government leadership has done something basic for us, first to provide us with adequate education and to provide children with insurance. However, I urge the government to continue with the support for these children by taking full financial responsibility for hospital care. We, parents of children with Sickle Cell, still need to provide good food, home care and most of us have very low incomes as childcare interferes with our businesses and jobs. **FP4, age: 48 years.**

Stigmatization Parents reported receiving support from their partners, nuclear and extended families, a few community members and also from healthcare professionals. Support was either instrumental, or emotional. Instrumental support was expected and obtained from spouses who provided financial support.

My family understands what sickle cell is and have been very supportive towards my wife and me. They cooperate in caring for the child when my wife and I need assistance. **MP3, age: 32 years.**

> The father currently has no problem. On clinic days, we leave at five in the morning he brings us here and later he goes to work. **FP5, age: 42 years**

However, in some narrations, it was evident that few parents faced discrimination towards themselves and their children. This stigmatization was largely influenced by the

communities' perception surrounding the disease. One mother expressed her concerns as follows:

The people we live with are aware but at school, some stigmatize him. The teachers show co-operation by educating children who do not understand. For example, my son has red eyes. His colleagues often avoid him and run away from him for the fear of contracting what he has. So, teachers try to tell and reprimand them not to stigmatize their partner because he has a problem and is sick. **FP4, age: 48 years.**

Another mother expressed her concern as follows:

> …when someone sees my child like this they say 'that child is weak, give him blood transfusions'. We give it to him, but one does not understand that the blood of a person with sickle cell is short-lived that is, it is formed and dies in a shorter time than a normal person. Similarly, when a person sees that he is out with his teammates playing, they say, 'don't play with him for he is sick' and they treat him as if he is not a normal person. **FP5, age: 42 years.**

Preferred method of intervention All parents were in support of more effort being placed on preventive measures that is premarital testing and counselling for genetic diseases. For example:

> I would prefer the government to put more effort in prevention of sickle cell disease to assist early diagnosis of sickled children should the parents decided to have children after knowing their sickle status. **FP6, age: 32 years**

Most parents emphasized on the priority being compulsory premarital testing over therapeutic management to avoid loss and pain in their families. As explained by one father:

> I think we should be tested before marriage and it should be compulsory. This is where the government should put more effort. I have lost my eldest child to sickle cell and other parents here have similar stories. If we know our status beforehand, the emotional turmoil that comes with loss of our children. **MP7, age: 56 years**

Another patient articulated the link between compulsory premarital genetic testing and the cost of childcare:

> They say that prevention is better than cure. It is therefore best to insist that couples test before marriage. From my point of view, I think it should be compulsory. When couples want to get married, they prioritize getting tested for HIV because they understand its implications. Making premarital genetic testing compulsory allows partners to be aware of their genetic status and the implications it may have economically and socially should they have a sickle cell disease child. **MP3, age: 32 years**

Two parents expressed their desire that equal effort be put towards prevention and management of cases. As one mother explains it:

I think equal effort should be placed on both sides. Children with sickle cell disease need better healthcare and better medication to improve their lives. The burden of sickle cell can be reduced if people know their status before they decide to have children. There is less shock on learning the child's status as you have been counselled before. So, I think both are important. **FP8, age: 29 years.**

While the primary concern of these parents was to avoid any long-term harm from lack of treatment in childhood, they also wanted to protect and maintain normality and protect childhood itself from medicalization. For example:

> …I would ask is for the government to help us with these children. They need to have good food but also to study in a good educational environment. Now you find a lot of us parents have little money, and we can't get to our goals of making sure those kids are well and educated in a good environment. So, I am asking if it is possible for the Government to set up a special unit for children with sickle cell problems, especially to raise them academically and to get money to buy them food that is suitable for their health. **FP4, age: 48 years.**

Discussion

This study aimed at exploring parents' views and experience of raising children with SCD. All the participants in the study were the biological parents of the children. The majority parents were mothers of the children with SCD except for three who were fathers. The findings highlight the role of a Tanzanian woman in the family as a caregiver, thus explaining why majority the participants were mothers. Consistent with these findings, a study carried out in Cameroon on the psychosocial burden of SCD on parents found that women were the main informal caregivers of children with SCD since they placed a high value on childbearing and caring for their children especially when they are ill Wonkam et al. 2014.

The study found that all participants knew SCD although their understanding of it varied. The majority of the parents knew that genes could transmit hereditary diseases and a fair number of them had moderate information about sickle cell disease. Some of the understanding was affected by social and cultural background. Some previously held the belief that there were supernatural explanations to the cause of

the disease in addition to genetics or the hereditary factors. It was observed that their beliefs were highly influenced by their communities' beliefs about SCD. Parents disclosed that their communities had varied views concerning the cause of the disease, treatment and lifespan. In some of the situations, male parents and the community believe that SCD in not a genetic disorder that is inherited from both parents but only from female parents (unpublished data). This has negatively affected the childcare from both parents.

Most of the participants knew about genetic testing as they had received counselling upon diagnoses of their children. However, prior to counselling, all parents reported not to have known about premarital genetic testing. A similar study conducted in Saudi Arabia found that most participants had some knowledge about hereditary diseases but had inadequate knowledge about premarital genetic testing Isah et al. 2016. This is an indication of the need for premarital genetic testing educational programmes as well as genetic counselling programmes targeting youth at the marriage age groups.

Premarital genetic testing is carried out for several reasons: to prevent disease transmission to offspring, to ensure health of their partner, to explore alternative means of having offspring and to provide counselling and advice to allow informed decision making between partners. Melaibari et al. 2017. A key finding was that all of the parents were supportive of premarital genetic testing for partners who wanted to start families but only half thought it should be compulsory. This is similar to previous studies in Saudi Arabia of attitude towards premarital genetic testing which found that the majority of people had a positive attitude towards screening for genetic diseases but were against it being compulsory because of fear of stigmatization and religious implications Moussa et al. 2018.

Concerning the introduction of compulsory premarital genetic testing before marriage, it was seen that the major conditions that made parents support the idea were empathy for the children's experiences that were burdened with frequent episodes of severe pain from the vaso-occlusive crisis, frequent hospitalization, delayed growth and development of their children, as well as the financial burden parents experience with children with SCD.

Most of the parents were found to manage crises at home as per the standard treatment guideline of Tanzania where non-steroidal anti-inflammatory drugs such as ibuprofen and acetaminophen were used. Other parents gave supportive treatment such as massage to help with the swelling of the joints. Parents often managed crisis first at home with over-the-counter drugs before sending their children to the hospital when their conditions became serious. This is congruent with findings of a study which revealed that the perception of parents about vaso-occlusive crisis influences the degree of threat associated with the disease and the urgency with which care is sought Pantaleao et al. 2019.

Moreover, the study found that parents who failed to manage crises at home depended on hospital care. It was found that the major indications for hospitalization in the children with SCD were anaemia and pain. The type of treatment given was consistent to most of the children. Acute infection is one of the most common causes of admission to hospital inpatients under 10 years hence penicillin prophylaxis and folic acid. Hydroxyurea is the only drug that reduces the frequency of acute vaso-occlusive complications Brousse et al. 2014. Children who were managed on hydroxyurea had less pain crises. However, the financial burden and inability of the national health insurance scheme to cover all required medication affected compliance of some children to medication hence increase in the frequency of crises.

The parents expressed hardship in the caring for children with SCD concerning the responsibility of preventing complications and caring for their children during a crisis. Parents felt that their children with SCD were not very marginalized by the surrounding community but required more support in care and finances.

Lastly, attitudes of medical staff and waiting time were the issues relating to the quality of care that posed a problem to parents. The parents reported that due to frequent hospitalization and routine medical check-up, they were always asking permission to stay out of work or had to leave their businesses. This led to even loss of job and a lower income. This finding is congruent with findings of a study in Cameroon which revealed that parents providing care to their children with SCD compromised their loyalty to their jobs and prevented them from giving out their best in their various jobs Wonkam et al. 2014.

Study limitations

This study has revealed that there is a need for further research on the experiences of parents of children with SCD to provide a deeper understanding of the phenomenon. This study involved parents who were attending sickle cell clinic at Muhimbili National Hospital, Dar es Salaam. Dar es Salaam is an urban business city with many health centres and opportunities. Further studies can be carried out in in health facility in rural areas and at community level. Communities' perceptions about SCD and their influence on their health-seeking behaviour can also be explored to provide an adequate understanding of the phenomenon.

Conclusions

The study showed that most of the children with SCD were diagnosed with the disease after suffering from complications of the disease. Therefore, there is the need to introduce premarital genetic testing as well as neonatal screening for SCD at all major health facilities as well as extend it to other healthcare facilities and incorporate it into prenatal care. This will allow for early diagnosis of the disease and early management with prophylaxis to reduce mortality and co-morbidities associated with the disease.

Moreover, the study revealed that parents caring for children with SCD developed an understanding of SCD after diagnosis of children. Awareness about genetic testing and counselling was limited. This indicates the need to intensify education on genetic counselling and testing for SCD. Additionally, there should be an adequate clinic for early screening, accessible therapeutic support and long-term follow-up for children with SCD. All interviewed parents were positive about genetic screening for sickle cell traits, where some insisting it becomes compulsory.

Regarding their experiences in raising SCD children, financial burden and inability of the national health insurance scheme to cover all required medication affected compliance of some children to medication hence increase in the frequency of crises. Parents recommended that preventive medications hydroxyurea should be made free to all families with children who have SCD in order to improve the outcomes of therapeutic care given to children with SCD.

Abbreviations *FP*: Father parent; *MP*: Mother parent; *SCD*: Sickle cell disease

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Author contribution EK participated in conceptualization, study design, data collection and analysis and manuscript drafting. RM and MK participated in manuscript revision and data interpretation. FS participated in conceptualization and designing the study. BJN, WPM, DM, GS and AIM participated in revising the manuscript. GMB participated in conceptualization, study design and drafting of the manuscript. All authors have read and approved the final version of this manuscript.

Data availability The datasets generated and/or analysed during the current study are available from the corresponding author on reasonable request.

Declarations

Conflict of interest The authors declare no competing interests.

Ethical approval Ethical clearance to conduct this study was obtained from Muhimbili University of Health Science (MUHAS), ethical review board, and the permission to conduct the research was obtained at MNH and presented to the head of paediatric department at MNH where the study was conducted. Participants were requested a written ethical consent before their enrolment. Confidentiality was ensured by using codes during data analysis and presentation.

References

Ali RMA, Razeq NMA (2017) The lived experience of parents of children with sickle cell disease: a qualitative study. Open J Nurs 07(11):1348–1364

World Health Organization. Control of Genetic Diseases (2005) https://www.apps.who.int/gb/archive/pdf_files/EB116/B116_3-en.pdf. Accessed 19 July 2021

Bozkurt G (2007) Results from the North Cyprus Thalassemia Prevention Program. Hemoglobin 31(2):257–264

Brousse V, Makani J, Rees DC (2014) Management of sickle cell disease in the community. BMJ 348(March):1–9

Colorafi KJ, Evans B (2016) Qualitative descriptive methods in health science research. Heal Environ Res Des J 9(4):16–25

Emmanuel OA, Andy E, Luka GD, Do A, Esther om, Oyedele Emmanuel CA (2015) Awareness and acceptance of premarital genotype screening among youths in a Nigerian community. Int J Med Heal Res 1(11):17–21. http://www.medicalsjournals.com

Farr BC (2008) Designing qualitative research. Transform an Int J Holist Mission Stud 25(2–3):165–166

Isah BA, Musa Y, Mohammed UK, Mto I, Kj A, Yunusa EU (2016) Knowledge and attitude regarding premarital screening for sickle cell disease among students of State School of Nursing Sokoto. Ann Int Med Dent Res 2:29–34

Lopez V, Whitehead D (2013) Sampling data and data collection in qualitative research. https://www.scholar.google.com/scholar?hl=en&as_sdt=0%2C5&q=Lopez+V%2C+Whitehead+D+%282013%29+Sampling+data+and+data+collection+in+qualitative+research.+In+2013.+p.+123–40&btnG=. Accessed 19 July 2021

Makani J, Cox SE, Soka D, Komba AN, Oruo J, Mwamtemi H, et al (2011) Mortality in Sickle Cell Anemia in Africa: A Prospective Cohort Study in Tanzania. PLoS ONE 6(2):e14699. https://doi.org/10.1371/journal.pone.0014699

Melaibari MA, Shilbayeh SAR, Kabli AO (2017) University students' knowledge, attitudes, and practices towards the national premarital screening program of Saudi Arabia. J Egypt Public Health Assoc 92(1):36–43

Moussa S, Al-Zaylai F, Al-Shammari B, Al-Malaq KA, Rashed Al-Shammari S, Al-Shammari TF (2018) Knowledge and attitude towards premarital screening and genetic counseling program among female university students, Hail region. Int J Med Heal Res 4(1):1–6

Pantaleao A, Di Placido J, Guite JW, Zempsky WT (2019) Caregiver factors related to emergency department utilization for youth with sickle cell disease. Child Heal Care [Internet] 48(1):59–74. https://doi.org/10.1080/02739615.2018.1454838

Rahman MM, Naznin L, Giti S, Islam MS, Khatun N (2015) Premarital health screening - a review and update. J Armed Forces Med Coll Bangladesh 10(1):103–109

Isangula K, Holmes A, Brownie S (2015) HIV Testing for PMTCT in Tanzania: Time to move from 'Voluntary' to 'Mandatory'?. Adv Soc Sci Res J 2(2):131–138. Available at: https://www.ecommons.aku.edu/eastafrica_fhs_sonam/90. Accessed 19 July 2021

Wonkam A, Mba CZ, Mbanya D, Ngogang J, Ramesar R, Angwafo FF (2014) Psychosocial burden of sickle cell disease on parents with an affected child in cameroon. J Genet Couns 23(2):192–201