HEALTH RELATED QUALITY OF LIFE IN CHILDREN WITH SICKLE CELL DISEASE AGED 8-18YEARS ATTENDING CLINIC AT MUHIMBILI NATIONAL HOSPITAL IN DAR ES SALAAM, TANZANIA

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MMed (Pediatrics and Child Health) Dissertation
Muhimbili University of Health and Allied Sciences
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By

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A Dissertation Submitted in Partial Fulfillment of the Requirements for the Degree of Master of Medicine (Paediatrics and Child Health) of Muhimbili University of Health and Allied Sciences.

Muhimbili University of Health and Allied Sciences
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CERTIFICATION

The undersigned certify that they have read and hereby recommend for acceptance by Muhimbili University of Health and Allied Sciences a dissertation entitled: **Health Related Quality of Life in children aged 8 to 18 years with sickle cell disease attending clinic at Muhimbili National Hospital in Dar es Salaam, Tanzania,** in (partial) fulfillment of the requirements for the degree of master of medicine (Paediatrics and Child Health) of the Muhimbili University of Health and Allied Sciences Dar es Salaam.

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Date

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Dr. Magdalena Lyimo.

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Date

DECLARATION AND COPYRIGHT

I, **Honesta John Kipasika**, declare that this dissertation is my own original work and that it has not been presented and will not be presented to any other University for similar or any other degree award.

Signature……**Date**……..

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DEDICATION

For my beloved father John Kipasika Lyimo and mother Theresia Paul Lekule, the foundation and support you have given me are immense. God bless you.

TABLE OF CONTENTS

LIST OF TABLES

LIST OF FIGURES

ACRONYMS

ANOVA	-	Analysis of Variance
AVN	-	Avascular necrosis
CHQ	-	Child Health Questionnaire
DM	-	Diabetes Melitius
HbSC	-	Sickle hemoglobin C disease
HbS beta .	-	Sickle cell beta thalassemia.
HIV	-	Human immunodeficiency virus
HgbSS	-	Hemoglobin SS
HPLC	-	High Performance Liquid Chromatography
HRQoL	-	Health related Quality of life
MNH	-	Muhimbili National Hospital
MUHAS	-	Muhimbili University of Health and Allied Sciences
PedsQL	-	Pediatric quality of life
QoL	-	Quality of life
SF-36	-	Short form 36
SCD	-	Sickle cell disease
WHO	-	World health organization

ABSTRACT

Background

Sickle cell disease (SCD) is prevalent in most of Africa countries, including Tanzania. In Dar es salaam, Tanzania approximately 17% of children entering hospital for any reason are carrier of sickle cell gene[3].

As a chronic illness and recurrent pain condition, SCD may cause a substantial burden on the daily functioning and wellbeing of children and their families. Less is known on HRQoL in children with SCD in Sub Saharan Africa particular in Tanzania.

Study Question; Do children with SCD in countries with limited resources like Tanzania have similar health related quality of life like children without SCD?

Broad Objective; To determine HRQoL in children with SCD aged 8-18 years compared to children without SCD in Dar es Salaam, Tanzania.

Methodology; This is descriptive comparative cross sectional study that involved children with SCD aged 8-18years.The patients under consideration were enrolled from the ongoing prospective SCD study at MNH.Comparison group were the random sample of siblings who do not have SCD obtained from the database in the same hospital in which the study population was obtained. The study used Generic PedsQL questionnaire for assessing HRQoL. Data entry was done using Epi Info and transferred to SPSS version 16 for analysis. The Independent sample t test and Analysis of variance (ANOVA) was applied to look for the association of HRQoL and factors such as socio demographic factors. Independent sample t test was also used to test for significant differences between children without SCD and the children with SCD. Association between HRQoL and hospitalization was tested using ANOVA. P - Value of less than 0.05 ($p < 0.05$) was considered significant.

Results; 100 children with SCD and 99 siblings without SCD aged 8-18 years and their parents were enrolled in this study. Females were 53% in SCD children and 63% in children without SCD. 45% of parents had primary education level and >85% were biological parents in both groups. Children with SCD and their parents scored lower HRQoL as compared to children without SCD, ($p<0.001$). Total mean score was 77.53 ± 12.9 in SCD children and 98.43 ± 3.5 in children without SCD. Children with SCD described their own HRQoL Significantly better than their parents perceived it to be. Lower age of the children and increased frequency of hospitalization had negative impact on HRQoL of these children.

Conclusion and Recommendations; Children with SCD and their parents perceived overall HRQoL and all subdomains to be lower than for children without SCD, therefore Assessment of HRQoL should be included in the guidelines of management of children with SCD as it will help to identify those who need psychosocial support and behavior intervention to promote appropriate functioning and to minimize activities restriction in order to improve their quality of life.

CHAPTER ONE

1. INTRODUCTION AND LITERATURE REVIEW

Sickle cell disease (SCD) is a major health care and societal problem that affects millions of people worldwide. It is a genetic disorder with an autosomal recessive pattern of inheritanceresults from the substitution of glutamic acid for valine at position 6 on the b-globin molecule that occurs predominantly in people of African ancestry[1].The disease is characterized by chronic haemolytic anaemia and vascular occlusion, causing recurrent painful episodes, irreversible organ damage, and neurocognitive deficits.

1.1.1EPIDEMIOLOGY OF SCD

More than 90% of those with SCD are born in Africa. The prevalence of the sickle cell trait ranges between 10 - 40% of the population in some parts of Africa[2].The estimated birth incidence of children with SCD in Tanzania is between 6 and 7 per 100 children. It is estimated that more than 50% of the children with SCA die before the age offive and over 5% of the infant mortality in Tanzania may be attributable to SCD[2].

Prevalence of HbSS in Tanzania is estimated to be 0.5% of all live births and prevalence of sickle cell trait up to 15% (Julie Makani unpublished). Other forms of SCD such as Hb SC and HbS βthalassemia are less common in Tanzania. In Dar es salaam, Tanzania approximately 17% of children entering hospital for any reason are carrier of sickle cell gene, among them four have homozygous form HbSS[3].

As a chronic health and recurrent pain condition, sickle cell disease (SCD) may cause a substantial burden on the daily functioning and wellbeing of children and their families. Children have to cope with disease exacerbations (e.g., recurrent pain) that may make it difficult for them to attend school, participate in sports, and play with friends[4].

1.1.2 QUALITY OF LIFE

1.1.2.1 Definition of Health Related Quality of Life.

World Health Organization (WHO) defines Health as the state of physical, mental, emotional, spiritual and social well being of an individual and not merely the absence of disease or infirmity. Quality of Life is individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns[6]. It is a broad ranging concept affected in a complex way by the person's physical health, psychological state, level of independence, social relationships, personal beliefs and their relationship to salient features of their environment[6].

Health-related quality of life (HRQoL) is a multi-dimensional concept that includes domains related to physical, mental, emotional and social functioning. It is a reflection of the way that patients perceive and react to their health status andto other nonmedical aspects of their lives. It goes beyond direct measures of population health, life expectancy and causes of death, and focuses on the impact health status has on quality of life. Clinicians and public health officials have used HRQoL and well-being to measure the effects of chronic illness, treatments, and short and long term disabilities[7].

1.1.2.2 Evaluation of Health Related Quality of Life

Evaluation of health related quality of life is important in determining the impact of chronic disease from the perspective of the child and parent. In clinical practice we rely on physiological evaluation which does not measure quality of life directly. HRQoL measures may be as simple as a single question like "how is your quality of life" to HRQoL instruments. These HRQoL instruments are questionnaires made up of numbers of items or questions which are added up to form domains and can be administered through different modes which include direct interviewer, telephone and self or surrogate responders[8].

There are generally two types of questionnaires used to measure HRQoL, generic and disease specific. AgenericHRQoLquestionnaire is not specific to a disease type and usually includes domains that measure physical functioning (e.g. can walk one block), emotional functioning (e.g. feel sad), social functioning (e.g. get along with friends),and school functioning (e.g. able to keep up with schoolwork)[9-10].

Ageneric HRQoL questionnaire can be utilized in healthy individuals and in those with chronic disease. It allows for comparison ofHRQoLacross different diseases. The Children's Health Questionnaire (CHQ)[11] and the Pediatric Quality of Life Inventory[9, 12–14](PedsQL) are commonly used generic HRQoL questionnaires forchildren. The SF-36is a commonly used generic questionnairefor adults[10].

Disease specific HRQoL instruments are comprised of questionsthat address specific areas of importance such as disease-relatedsymptoms (pain).

Health related quality of life questionnaires have been used in several pediatric SCD studieswhich have provided preliminaryevidence for validity and reliability[16-18].
In these studies, children with sickle cell disease andtheir caregiversreported more limited physical, psychological, and social well-being than healthy children.

1.1.2.3Pediatric Quality of Life Inventory;

The measurements of health related quality of life is a difficult process as health is dynamic, varying with age locations and beliefs. Currently there are several instruments which are used to measure quality of life in various disease conditions.

The PedsQL Measurement Model is a modular approach to measuring health-related quality of life (HRQoL) in children and adolescents. The PedsQL consists of brief, practical, generic core scales suitable for use with healthy school and community populations, as well as with pediatric populations with acute and chronic health conditions[14].It is designed to measure the core health dimensions delineated by WHO, including school functioning. It integrates both generic core scale and disease specific module into one measurement system.

Paediatric HRQoL measurement instruments must be sensitive to cognitive development. It must include both children self-report and parent proxy-report. The PedsQL Measurement Model consists of developmentally appropriate forms for children ages 2-4, 5-7, 8-12, and 13-18 years. Paediatric self-report is measured in children and adolescents ages 5-18 years, and parent proxy-report of child HRQoL is measured for children and adolescents ages 2-18 years [14].

The multidimensional PedsQL Generic Core Scales include the essential core domains for paediatric HRQoL measurement which are Physical Functioning (8 items), Emotional Functioning (5 items), Social Functioning (5 items) and School Functioning (5 items)[14].

1.1.3 Health Related Quality of Life and SCD

Health-related quality of life (HRQoL) may be impaired in sickle cell disease (SCD) due to morbid events,such as painful episodes, infections, anaemia, avascular necrosis,acute chest syndrome, splenic sequestration, stroke, or other organ system failures.

It is important to measure HRQoLin children with SCD in order to describe their healthprofile and functional status, and to evaluate their needs and those of their families.Regardless of this need, there are few data on HRQoL in children with SCD in developing countries.

Some studies have examined the influence of various determinants on quality of life in SCD including the role of socio-demographics, disease severity and the presence of complications. Most of those studies were carried out in developed countries.
A research by McLish et al[17] using SF 36 Questionnaire indicated that patients with SCD perceived overall HRQoL to be lower compared to the general population Dampier et al[18]found that children with SCD as well as their parents scored significantly lower on several HRQoL domains including; general physical, motor and independent daily functioning.

In a study by Kater et al[19] by using a multidimensional parent-completed HRQoL measure standardized in The Netherlands to compare HRQoL between a sample of children withSCD and health children. Results showed that children with SCD decreased independence in daily functioning and general physical limitations compared to healthy children.

In a study by Panepinto et al [20], 104 parents of children with sickle cell disease and 74 parents of children without disease completed the parent report of the PedsQL questionnaire, and 78 children with sickle cell disease and 40 children without disease completed the self report PedsQL questionnaire. Findings from this study revealed that children with sickle cell disease had worse HRQoL in all summary scores when compared to children without sickle cell disease even after considering the potential effect of family income on HRQoL. Children with sickle cell disease had worse HRQoL in the area of physical functioning as might be expected given that pain is a predominant symptom in children with sickle cell disease. Also social and school functioning where impaired compared to children without sickle cell disease.

Trzepaczet al[21]identified deteriorations in social and school competence for children with SCD, compared to healthy peers, but they did not find an association with disease severity as measured by sickle cell genotype. Panepinto et al[22] by using the CHQ–Child Form to 53 children with SCD and the CHQ–Parent Form to 95 parents/caretakers reported that children with SCD had significantly impaired HRQoL when compared with healthy children. Children self reported poorer functioning compared with healthy peers (mean difference) in physical functioning (-12.31), bodily pain (-12.27), general health perceptions (-11.93), and social functioning (-16.59) subscales.

In a qualitative study done by Taylor et al[23] in USA byusing focus group to asses HRQoL in 25 children and adolescent with SCD shows that children and adolescents with SCD report poor HRQoL in all domainsand fare worse in their HRQoL compared to controls on assessments of general physical, motor and independent daily functioning[24].

1.1.4 Health Related Quality of Life and social demographic characteristics.

Previous researches on chronic diseases show that HRQoL varies according to socio-demographic characteristics such as family income, educational status, marital status, occupational status, age, and gender, with the deprived groupsreporting lower HRQoL.It has been reported for many chronic conditions including cancer by Dapueto et al in South America[25]HIV infections by Akincigil et al [26],renal disease by Kimmel et al[27] and sickle cell disease[17-18, 20].

Several studies were done in developed countries to examined the influence of various determinants including the role of social demographics on HRQoL in SCD patients, results shows that older child age, female gender, lower income, and more severe disease were found to correlate with greater limitations in the physical health of children with SCD[17-18,22,32].

A study by Van den Tweel et al[29] in Amsterdam, Netherlandsshowed thatchildren of parents with low educational level perceived a significantly better HRQoL.This fact is not easy to explain, given that previous research mainly pointed out that high quality of life scores associated with high parental education, or that education had no effect at all[30-31]. Also, previous research by Varni et al[32] has shown that children with low socioeconomic status (SES) functioned worse than children from middle SES backgrounds.

Panepinto et al[20] usePedsQL Questionnaireto find out the impact of family income and SCD on the HRQoL of children in USA, a total of 104 children with sickle cell disease and 74 without disease participated in the study. After adjusting for family income, patient age, presence of co-morbidities and children with severe sickle cell disease had increased odds of poorer overall HRQoL (parent-proxy HRQoL report odds ratio [OR] 4.0) and physical HRQoL (parent-proxy report OR 5.67, child self-report OR 3.33) compared to children without sickle cell disease.

1.1.5 HRQOL in relation to associated SCDcomplications and frequency of hospitalization;

In patients with SCD, Health-related quality of life (HRQoL) may be impaired due to numerous physical symptoms and disease related complications.
Common symptoms and complications of SCD include recurrent pain, anemia, low exercise tolerance, lung problems, osteomyelitis, avascular necrosis, pulmonary hypertension, growth delay, strokes, priapism, delayed sexual maturation, and enuresis.[33-34]

These complications may require hospitalization thus affecting school attendance and normal play activities which have a negative impact on the HRQoL of these children.

Acute and chronic SCD complications may lead to predictable disease-related symptoms. For example, hemolysis causes variable degrees of chronic anemia that

contributes to fatigue, activity limitations, and vaso-occlusion associated with sickled erythrocytes cause acute and chronic pain[35].

Platt et al[4]showed that, among SCD complications, the frequency of pain events in childhood has been found to result in most significant morbidity and mortality in adulthood. Pain may start from the age of 6 to 9 months to some children, while in others pain does not occur until adolescence or early adulthood[5].

Patients who survive until adulthood, frequently experience significant organ system damage that may include stroke, pulmonary failure, pulmonary hypertension, renal failure, congestive heart failure, leg ulcers, and osteonecrosis of the femoral or humeral heads.

One of the major complications of HgbSS is stroke and its neurological sequelae. In children, this brain injury is usually ischemic in nature.

In 2002, Palermo et al[36] by using the Child Health Questionnaire (CHQ)—Parent Report to compare HRQoL of 58 children with SCD to a demographicallysimilar group of 120 healthy children found that HRQoL of children with SCD were negatively affected. Parents and caregiver of children with SCD reported more limited physical, psychological, and social functioning compared with healthy children. Older child age, female gender and more disease related complications predicted limitations in physical functioning and social wellbeing of these children.

Dampier et al [18]showed that the occurrence of acute and chronic complications requiring medical management was expected to have a negative impact on HRQoL and the most frequent SCD-related complications were the occurrence of vaso-occlusive pain and the presence of AVN of one or both shoulders/hips. The occurrence of an acute care visit for pain management was used as an example of a SCD acute complication and was associated with substantially diminished scores on virtually all parent- and child-reported scales of the PedsQL.

Mostafa et al[37]used SF-36 questionnaire to measure quality of life among children and adolescent with SCD in Saud Arabia, resultsshows that adolescents with SCD have a significant educational delay in terms of excessive failing and school retention compared tochildren and adolescents without SCD. 15% of adolescents with SCD demonstrated delay in the primary education compared to only 2% among adolescents without SCD, and 87.7% of adolescents with SCD in thepreparatory stage were delayed compared to 21.1% among adolescents without SCD. This delay wasattributed by the parents due to excessive absenteeism from schools in response to frequent hospitalization, emergency admissions, andappointments for checkups.

In a study done by Shapiro and colleagues [38] found that children with SCD who experienced frequent pain were absent from school on 21% of school days, with half of the absences occurring on days with reported pain. Also many children with SCD reduce their participation in peer activities [39] and experience disruptions in their sleep quality and sleep duration when experiencing painful events[40].

In 2005, Patel et al[41]by using multidimensional interview based questionnaire, assess the quality of life among 52 children with sickle cell disease and 12 children with sickle cell traits, results shows that all domains, physical, psychosocial and cognitive were affected[41].In SCD playing and mobility were most affected. There was feeling of sadness or disinterest and lack of support from teachers. The school attendance, vocational achievement perception, entertainment and participation in cultural activities were also affected.

1.2 STATEMENT OF PROBLEM

Sickle cell disease is a common genetic disorder in Tanzania. Recently Tanzania prevalence of HbSS is estimated to be 0.5% of all live births and prevalence of sickle cell trait up to 15% (Makani unpublished).

A number of disease-related factors have been found to have negative impact on HRQOL in children with SCD

Previous research in developed countries found that patients with SCD experienced a lower HRQoL compared to the general population[17-22, 24,37,41-42].

Very few studies have been done to determine HRQoL in children with SCD in developing countries particular in Sub Saharan Africa. No data are currently available on HRQoL in children with SCD in Tanzania.

This study has the interest of exploring further the concept of health related quality of life in children with SCD compared to children without SCD. Also it will determine the association of health related quality of life and social demography and number of hospitalization of these children.

1.3 RATIONALEOF THE STUDY

Health related quality of life (HRQoL) of children with SCD has not been well described despite the psychosocial and physical consequences associated with the disease, very little is known in developing countriesand no study has been done in Tanzania.

Assessing HRQoL will improve awareness and add knowledge in terms of addressing the effects of SCD on the health related quality of life of these children, it will alsohave future impact on public healthin the management of children with SCD in terms of addressing their needs to improve their quality of life as well as addressing their psychosocial needs as they grow up to adolescence and adulthood.

Moreover this study will enlighten on the role of SCD complications on impaired HRQoL and the importance of early diagnosis and intervention to prevent complications, those with psychological impairments will need a follow up by a psychiatrist thus improving the QoL and increase the life span of these children.

1.4RESEARCH QUESTION

Do children with SCD in countries with limited resources like Tanzania have similar health related quality of life like children without SCD?

1.5OBJECTIVES OF THE STUDY

1.5.1 Broad Objective

Assessment of health related quality of life in children aged 8 -18 years with sickle cell disease in comparison to that of children without SCD in Dar es Salaam, Tanzania.

1.5.2 Specific objectives

1.5.2.1. To determine HRQoL in terms of physical, emotional, social and school functioning of children with SCD.

1.5.2.2. To determine the HRQoL among children with SCD by demographic characteristics of the study population. (Age,sex, education of the parents/caregivers)

1.5.2.3. To determine the impact of hospitalization due to SCD related complications on HRQoL.

CHAPTER TWO

2. METHODOLOGY

2.1 Study Design

Descriptive,hospital basedcomparative cross sectional study.

2.2 Study population

2.2.1 Children with SCD

Children with SCD were enrolled from the study on prospective of SCD which is conducted at MNH.

This study was initiated in April 2004, it include active recruitment of newly diagnosed SCD patients and follow up of the current cohort. In this study eligible participants were those with ages ranging from 8-18 yrs. Study participants were scheduled for routine outpatient visits every 3- 6 months at which detailed clinical and laboratory data are recorded. Also all admissions to MNH of SCD patients and reasons for admissions are well documented. At each clinic attendance, patients are questioned about clinical complications and hospital admissions since last seen, and relevant hospital notes are periodically reviewed. Clinic records are therefore believed to be reasonably complete and comprehensive.

2.2.2 Comparison group.

These were the random sample of siblings who had no SCD obtained from the database in the same hospital in which the study population was obtained. These children were confirmed SCD negative by Hb electrophoresis and were not matched by age or sex.

2.3 Sample size

Sample size was determined by using one of the sample size estimation methods for PedsQL outcome. The formula chosen is the one for comparing two independent means using *t* test. Assuming the outcome is continuous variable and follows normal distribution,

$$n = 16\sigma^2/\Delta^2 + 1$$

Δ is the standardized difference between means of scores on PedsQL Children with and without SCD

n represents the required sample size per group

$\Delta = \mu_1 - \mu_2$

σ represents the standard deviation of the variable

Whereby μ_1 = mean score on PedsQL of children without SCD in Texas which is 86.9

μ_2 = mean score on PedsQL of children with SCD is 68.9

σ = standard deviation which is 12.5

Therefore, Δ =83.9 – 68.6/12.5

Substituting in the formulae for mean then the sample size, n will be

$$n = \frac{16(15.3)^2}{(12.5)^2} + 1$$

n =36 per group. (Minimum)

In this study I include 100 children and their parent / caretaker in each group.

This study was designed to have a power of 80% and confidence level of 95%.The ratio of two groups to be compared will be 1:1

2.4 Sampling method and randomization

SCD group;

Average of40 children with SCD attended SCD clinic per day (on Thursday and Friday)with maximum of 15 children aged 8-18. All eligible participantswere assigned numbers andthose with an odd number wererecruited into the study after obtaining assent and consent from the children and parents/caretakers. Maximum of 14 childrenwere recruited per week.

Comparison group;

There were 267 eligible siblings aged 8-18 years without SCD in the data base of the sickle cell study cohort, among them 173 had active contact mobile numbers. Simple computer generated randomization were done to this 173children, and a total of 100 children were obtained.

2.5 Recruitment of study subjects

SCD group;

Recruitment of children with SCD and their parents were conducted exclusively on Thursday and Friday during clinic hours after providing information regarding the study to the children and parents/caregiversafter which they were requested to sign an informedconsent form for participation in the study.

Comparison group;

Recruitment of comparison group was done on Wednesday after being informed two days earlier through the phone. Those who come to Muhimbili hospital, transport fee were refunded while those who did not come were followed to their homes but one of these children had travelled during the recruitment period hence we remain with 99 siblings. They were also requested to sign an informed consent for participation in the study.

2.5 Study area

Study was conducted at the SCD clinic outpatient department, Muhimbili National Hospital, Dar es Salaam, Tanzania. Dar es Salaam is the commercial city of Tanzania located along the East Coast of Indian Ocean. It has three municipalities namely; Ilala, Temeke and Kinondoni. MNH is one of the biggest hospitals in the country and serves both as referral and teaching hospital for MUHAS.

2.6 Study duration

This study was conducted for a total period of 14 months, from February 2012 to April 2013 which included proposal development, data collections, analysis, and dissemination of result findings.

2.7.1 Inclusion criteriain children with SCD

The eligible children for this study fulfilled the following criteria:

(1) A confirmed diagnosis of SCD.

(2) 8-18 years old.

2.7.2Exclusion criteria in children with SCD

(1) Children who were severely ill requiring admission were notincluded in the study

(2) Parents/guardian who did not grant informed consent to participate in the study.

(3)Children who did not assent.

(4) Other confirmed chronic illnesses i.e. DM, Asthma etc.

2.8.1Inclusion criteria in comparison group

(1) Children aged 8-18yrs.

(2) Confirmed sickle cell disease or trait negative.

2.8.2Exclusion criteria in the comparison group

(1)Children who were severely ill, who were not be able to respond to questions.

(2) Parents/guardian who did not sign written informed consent.

(3) Children who did not assent.

(4) Confirmed other chronic illness i.e. (DM, Asthma)

2.9Informed consent.

Written and signed consent were also be sought from parents of the study candidates as they are minors and considered incapable of giving consent. The participants were told about the purpose of the study and asked for their willingness to participate in the study. It was also made clear that acceptance or refusal to participate has no outward consequences and that they are free not to participate in the study. They were also assured of confidentiality. In addition, candidates were informed that no financial gain will be obtained by participating in this study. They were given the address/contacts of the principal investigator as well as the contacts of the director for research and publication committee from MUHAS

2.10Assent

An assent was also sought from all children aged 8-18 yrs.

2.11Research instruments

Pediatric Quality of Life Inventory tool was used as an instrument of obtaining data on health related quality of life of these children. This tool has the parallel children report and parental/caretaker proxy report.

2.11.1Paediatric Quality of Life Inventory questionnaire

The tool distinguished between healthy children and children with acute and chronic health conditions, also distinguished disease severity and chronic health condition.

It is one of the most widely used tool to assess the HRQoL in children, its advantages include availability, brevity and age specific version and consist of parallel forms for child and parent/caretaker. It has been shown recently to distinguish between healthy children from those with acute/chronic disease condition.

Scaling and Scoring of PedsQL Inventory

The Paediatric quality of life inventory is composed of23 items comprising 4 dimensions as stated earlier. Higher scores in the inventory indicate better health related quality of life. there are 5 point scale and 3 point scale.5 point scale ranges from 0(never), to 4 (almost always) and 3 point scale ranges from 0(not at all) 2(sometimes) and 3(a lot).These scores are transformed on a scale from 0 to 100.

The designed structured questionnaire included socio-demographic factors such as age, sex,education, employment of the parents/guardian and marital status. It also included the number of admissions in the past 12 months.

Scoring procedure:

Transforming score whereby items are reversed scored and linearly transformed to 0 to100 scale as 0=100,1=75,2=50,3=25 and 4=0 (so that higher PedsQL score indicate better HRQoL). Mean score is equal to sum of items over the number of items answered. Psychosocial health summary score is equal to sum of items over the number of items answered in the emotional, social and school functioning scale.

Physical health summary score is equal to the physical functioning scale score and total score is the sum of all items over the number of items answered on all the scale.

2.11.2Reliability and Validity of the Instrument

The pediatric quality of life inventory has been shown to have internal consistency reliabilities that exceed the standard of 0.7 for group comparisons. An alpha of 0.90 is usually recommended for individual patient analysis and this tool has been shown to approach this level inTotal Scale Score for self-report and proxy-report hence can be used in primary analysis of HRQoL outcome in clinical trials and other group comparisons.

The validity of the instrument was also analyzed using known group methods correlations with indicators of morbidity and illness burden, and factor analysis and has been shown to distinguish between healthy and pediatrics patients with acute or chronic ill condition and has been shown to relate to indicators of morbidity and illness burden and displayed a factor-derived solution largely consistent with the a priori conceptually-derived scales.

2.11.3 Cut offs scores for PedsQL

The established cut off scores meaning poor quality of life by using PedsQL generic instrument were 69.7 for child self report and 65.4 for parental proxy report in comparison with general population[43].There is no published data that grade the quality of life into severe, moderate and mild.

For children older than 8 years the recommended cut off scores for using total functioning to identify children with special health care needs were 78, for moderate were 76 and for major chronic conditions were 70[44].

2.11.4 Pre-testing of the PedsQL questionnaire

10% of the estimated sample size, which were 10 children with SCD and 10 children without sickle cell disease were used to pretest the questionnaire. The pre-test examining whether the tool could distinguish between two groups.

Pre-testing of the research instruments was done at Muhimbili National hospital.Pretesting is useful as it led to the reworking of unclear questions in the instrument.

2.11.5Data management and analysis:

The data were entered in Epi Info for storage and every day the data were cleaned to minimize errors and establish missing data which were then updated.

The data analysis was done by using SPSS version 16. The descriptions of socio-demographic characteristics were analyzed to establish means, standard deviations and frequency distributions.

The analysis of Variance (ANOVA) and independent t test was used to look for the association of QoL and factors such as socio demographic factors. Independent t test was used to test for significant differences between the children with sickle cell disease and children without SCD. T test were also used to test the significance differences between children with SCD and their parents while association between HRQoL and hospitalization were tested with ANOVA.

For statistical significant difference on ANOVA test a further Bonferroni alpha post hoc test with adjusted p- value was applied to detect the difference among groups.

Internal reliability was assessed using Cronbach's alpha for each of the four subscales of the PedsQL as well as for the summary and total scores.

The association was considered to be statistically significance with p value of< 0.05.

2.12 Ethical Clearance

Ethical clearance to conduct the study was obtained from the senate Research and Publication Committee of Muhimbili University and Allied Sciences Ethical Review Board. Permission to do the study was sought from MNH and Health Authorities. Assent to participate in the study was sought and obtained from the eligible study candidates.

2.13 Ethical consideration

Written informed consent was obtained from each parent/guardian of the child who met criteria before enrolling a child into the study. Description of the purpose of the study, its expected outcomes, potential benefits and risks, confidentiality of the information provided were clearly provided to ensure informed choice.

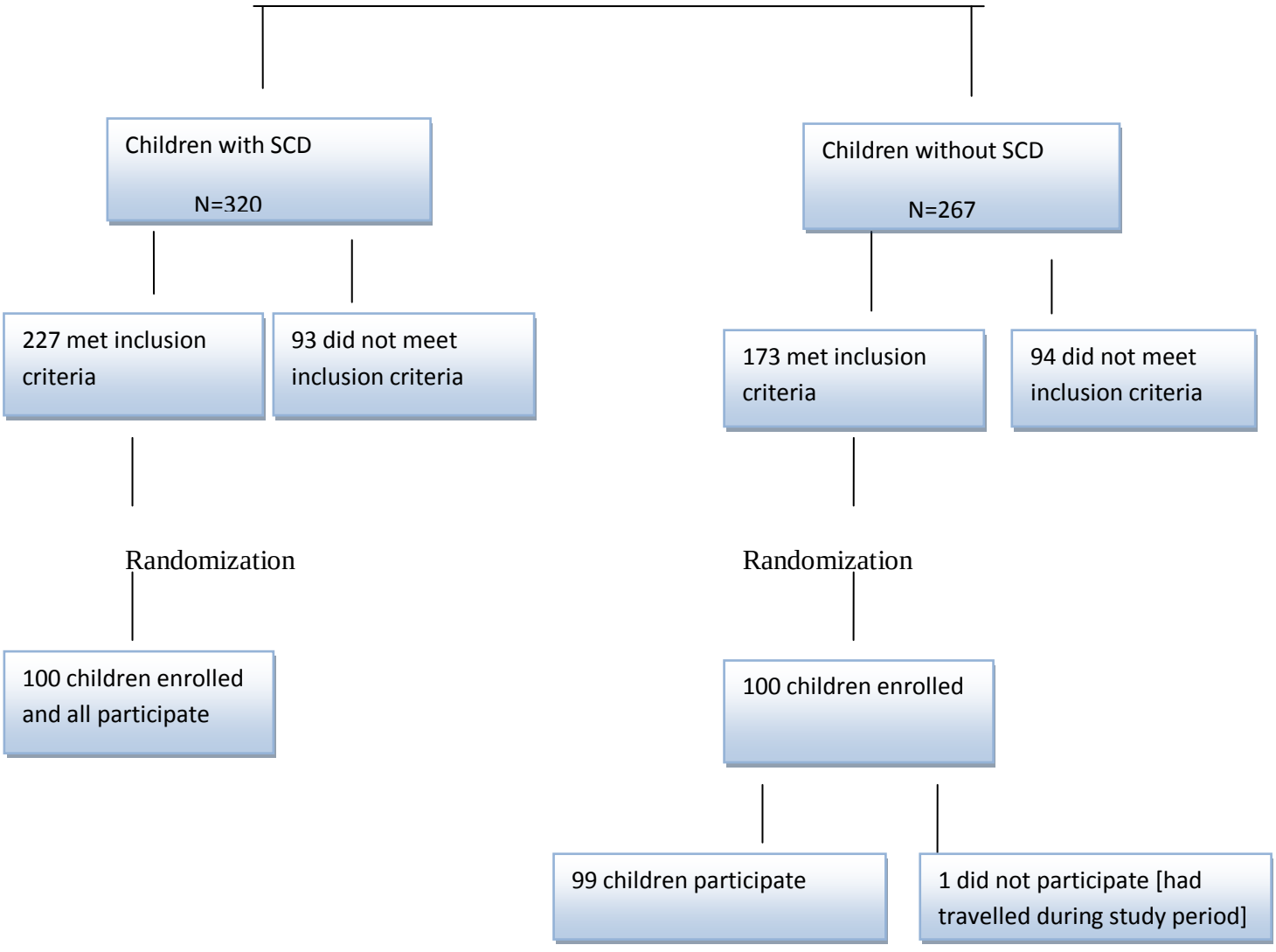
CHAPTER THREE

3. RESULTS

3.1. Study Participants.

This study involved a total of 100 children with SCDaged 8-18 years attending sickle cell clinic at MNH and a comparison group of 99 sickle cell free siblings selected randomly from the sibling database at the same clinic.

Figure 1: Participants flow chart



3.2. Social demographic characteristics of respondents;

Female were 53% in SCD group and 63.4% in comparison group and the age of the participants ranged from 8 to 18 with a mean{sd} age of 13.2 ± 2.87 and 13.25± 3.15 in SCD group and comparison group respectively

The majority of the participants in the SCD and comparison groups were in primary school ie 66(66%) and 61(61.61%) respectively. 4% of children with SCD do not attending school because of SCD related reasons.

The study also involvedparents /care takers of both the SCD group and the comparison group; among these 80% were females and 20 % of the care takers were males. Biological parents were 87% in the SCD group and 90.90% in the comparison group. The majority of these care takers i.e. 83% in the SCD group and 85.85% in the comparison group were married.

The care takers in both groups were of varied employment status, in the SCD only 46% were formally employed 37% unemployed and 17% self employed while in the comparison group 34.34% were formally employed , 52.52% unemployed and 13.13% self employed

Table 1: Social demographic characteristics of respondents

Variables	**SCD N=100 n(%)**	**Control N=99 n**(%)
Sex		
Male	47(47.00)	36(36.36)
Female	53 (53.00)	63(63.63)
Age		
8-12yrs	36(36.00)	41(41.41)
13-18yrs	64(64.00)	58(58.58)
mean age	13.20	13.25
Sd	2.87	3.15
Place of residence		
Ilala	31(31.00)	19(19.19)
Kinondoni	34(34.00)	33(33.33)
Temeke	35(35.00)	48(48.48)
Child level of education		
not school	4(4.0)	0(0.00)
primary school	66(66.0)	61(61.61)
secondary school	30(30.0)	38(38.38)
Parental employment status		
Employed	46(46.00)	34(34.34)
Unemployed	37(37.00)	52(52.52)
self employed	17(17.00)	13(13.13)
Parental education level		
No formal education	0(00.00)	3(03.03)
Primary	45(45.00)	44(44.44)
Secondary	23(23.00)	27(27.27)
Collage	32(32.00)	25(25.25)
Parental marital status		
Single	5(5.00)	4(4.04)
Married	83(83.00)	85(85.85)
Divorced	4(4.00)	3(3.03)
Cohabiting	1(1.00)	2(2.02)
Widowed	7(7.00)	5(5.05)
Relationship with the parents		
Biological Parent	87(87.00)	90(90.90)
Caretaker	10(10.00)	4(4.04)
Grandparent	2(2.00)	3(3.03)
Step parent	1(1.00)	2(2.02)

3.3. Disease characteristics of sickle cell sample

Among the SCD patients involved in the study, 63% had no history of hospitalization in the 12 months preceding the study while 25% were admitted less than 3 times and 12% were admitted more than three times in the 12 months period before the study.

The major reason cited for the reported hospitalization {n=37} was vasoocclusive pain crisis 59.46%, severe anemia 48.65%, malaria 27.03% and stroke 13.5%.

Regarding blood transfusion history in the prior 12 months in the SCD group 80% of the participants had not been transfused while 16% were transfused <3times and 4% were transfused more than three times. The reasons for the blood transfusions were in the majority severe anemia 85%.

Table 2: Disease characteristics of Sickle cell sample

Variable	No(%)
Diagnosis	**N**
SS	97 (97.00)
HbS βthalassemia	3 (0.3.00)
Hospitalization in prior 12 months (100)	
no hospitalization	63(63.00)
1-3 times	25 (25.00)
> 3 times	12 (12.00)
Reasons for hospitalization (n=37)	
Anemia	18 (48.65)
Chest problems	3 (08.11)
Painful episode	22 (59.46)
Stroke	5 (13.51)
Avascular necrosis	3 (08.11)
Chronic osteomyelitis	1 (02.70)
Malaria	10 (27.03)
Blood Transfusion in prior 12months (100)	
No transfusion	80 (80.00)
1-3 times	16 (16.00)
> 3 times	4 (04.00)
Reason for blood transfusion (n= 20)	
Severe Anemia	17 (85.00)
Stroke	3 (15.00)

3.4. Mean score of PedsQL scale betweenchildren with SCD and those without SCD

Children with SCD had a lower Mean score of 77.53 ± 12.9 compared to children without SCD 98.43 ± 3.5. Children with SCD score lower in all four parameters of PedsQL questionnaire as compared with children without SCD and it is highly statistically significant with P- value <0.001. Children with SCD scored the highest in emotion functioning 92.26 ± 9.31 and lowest in physical functioning 71.56 ± 16.3 followed by school functioning 73.69 ± 18.5.

From parent reports, total mean score was 97.27 ± 4.6 and 73.79 ±14.6 in children without SCD and in those with SCD respectively. Parents of SCD children scored lower in all parameters of PedsQL as compared to parents of children without SCD and this association is statistically significant {P<0.001}. The highest score was in emotional functioning 92.27 ± 9.3 and lower on physical functioning 67.34 ± 16.5.

Table 3:Mean score of PedsQL in children and Parents report between Children with SCD and those without SCD.

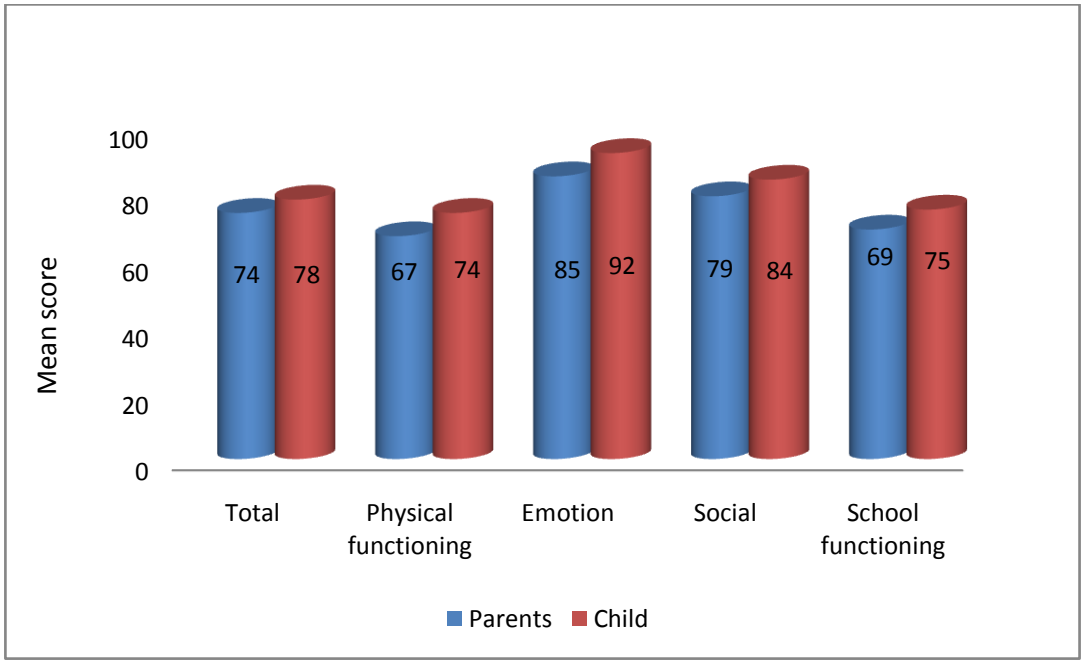
		Non SCD		SCD	
Scale	n	Mean(sd)	n	Mean(sd)	P-value
Child self report;					
Total Mean score	99	98.43 ± 3.5	96	77.53 ± 12.9	<0.001
Physical functioning	99	96.65 ± 6.4	100	71.56 ± 16.3	<0.001
psychosocial					
Functioning	99	98.99 ± 2.8	96	84.90 ± 12.2	<0.001
Emotional					
functioning	99	99.05 ± 3.2	100	92.26 ± 9.31	<0.001
Social functioning	99	98.91 ± 4.2	100	84.20 ± 13.7	<0.001
School functioning	99	98.71 ± 4.4	96	73.69 ± 18.5	<0.001
Parent report;					
Total Mean score	99	97.27 ± 4.6	96	73.79 ± 14.6	<0.001
Physical functioning	99	95.95 ± 6.8	100	67.34 ± 16.5	<0.001
Psychosocial					
functioning	99	97.83 ± 4.2	96	80.12 ± 13.0	<0.001
Emotional					
functioning	99	99.06 ± 3.2	100	92.27 ± 9.3	<0.001
Social functioning	99	99.46 ± 2.5	100	79.33 ± 13.1	<0.001
School functioning	99	94.95 ± 9.7	96	69.07 ± 22.8	<0.001

All scale 0 – 100. A high score indicates better health.

3.5. Mean score of PedsQL scale between children with SCD and their parents

There is a significant difference in mean scores between children with SCD and their parents. Children with SCD score higher compare to their parents with overall generic mean score of 78.83 ± 12.9 and 73.79 ± 14.6 respectively and this association was highly statistically significant on sample t test with P-value <0.001.

Figure 2: Mean score of PedsQL between children with SCD and their parents



All scale 0 – 100. A high score indicates better health. Sample t testP<0.001

3.6. Mean score of PedsQL Scale of children with SCD by age groups (n=100)

Children with SCD aged 13-18years had higher mean scores on PedsQL compared to those aged 8-12years in all scales in child report. This indicates that older age is associated with improved HRQOL as perceived by children. However difference in the mean score was only statistically significant for social functioning {P-value 0.028} and school functioning{P-value <0.001}

Table.4; Mean score of PedsQL scale of children with SCD by age groups {n=100}

		8-12years		13-18years	
Scale	n	Mean(sd)	n	Mean(sd)	P-value
		n=36		n=64	
Total Mean score	33	76.32 ± 13.9	63	84.40 ± 11.8	0.004
Physical functioning	36	66.99 ± 17.0	64	76.61 ± 15.1	0.05
Emotional functioning	36	90.79 ± 10.3	64	91.07 ± 9.9	0.06
Social functioning	36	79.79 ± 15.0	64	86.27 ± 12.7	0.028
School functioning	33	67.12 ± 22.3	63	83.62 ± 15.9	<0.001

All scale 0 – 100. A high score indicates better health

3.7. Mean score of PedsQL Scale of children with SCD by sex (n=100)

The results indicate that being male or female does not have impact on HRQoL in children with SCD as evidenced by overall total sores of 81.04 ± 15.1 and 82.77 ±11.0 in males and females respectively {P-value 0.517}. Females score higher in all subdomains of PedsQL except for emotion function however this association was not statistically significant.

Table 5: Mean score of PedsQL scale of the children with SCD by sex

		Male		Female	
Scale	n	Mean(sd)	n	Mean(sd)	P-value
		n=47		n=53	
Total Mean score	43	81.04 ± 15.1	53	82.77 ± 11.0	0.517
Physical functioning	47	73.07 ± 19.3	53	74.99 ± 13.1	0.85
Emotional functioning	47	94.04 ± 8.0	53	90.69 ± 10.1	0.54
Social functioning	47	83.26 ± 15.5	53	85.00 ± 12.1	0.79
School functioning	43	75.45 ± 22.7	53	81.38 ± 16.2	0.43

All scale 0 – 100. A high score indicates better health

3.8. Mean score of PedsQL scale in children with SCD by education level of parents.

Level of education of the parents does not have impact in children with SCD as perceived by both children and their parents in all parameters of Generic PedsQL scale.

Table 6: Mean score of PedsQL scale of the study population by parent education level

		primary education		Secondary education		Collage/university	
Scale	n	Mean(sd)	n	Mean(sd)	n	Mean (sd)	p-value
		n=45		n=23		n=33	
Child report;							
Total Mean score	41	83.30 ± 12.6	23	79.22 ± 13.2	32	82.22 ± 13.3	0.471
Physical functioning	45	75.48 ± 16.5	23	68.47 ± 16.3	32	74.51 ± 15.6	0.227
Emotional functioning	45	94.37 ± 7.2	23	90.14 ± 8.9	32	90.83 ± 11.5	0.12
Social functioning	45	86.37 ± 13.7	23	79.42 ± 13.9	32	84.58 ± 13.7	0.142
School functioning	41	78.41 ± 19.8	23	78.84 ± 19.6	32	78.95 ± 19.6	0.992
Parent report;							
Total Mean score	41	75.21 ± 14.4	23	71.54 ± 15.2	32	76.53 ± 14.6	0.451
Physical functioning	45	68.95 ± 16.3	23	61.82 ± 17.4	32	69.04 ± 15.8	0.193
Emotional functioning	45	85.62 ± 10.5	23	82.89 ± 12.0	32	84.16 ± 12.9	0.647
Social functioning	45	79.85 ± 13.3	23	75.94 ± 12.5	32	81.04 ± 13.2	0.344
School functioning	41	68.94 ± 23.7	23	65.50 ± 24.2	32	71.87 ± 20.8	0.592

All scale 0 – 100. A high score indicates better health.

3.9. Mean score of PedsQL Scale in children with SCD who were not hospitalized, hospitalized 1-3times and those who were hospitalized >3times in prior 12 months.

Children who were admitted more than 3 times in prior 12 month and their parents score lower on PedsQL scale as compared to those who were not admitted or admitted less than three times. The general trend is that there is decrease mean score with increase number of hospitalization, indicating lower HRQoL with increase hospitalization which is below the cut off scores established for PedsQL.

Table 7: Mean score of PedsQL Scale in children with SCD whowere not hospitalized, hospitalized 1-3times and those who were hospitalized >3times in prior 12 months

		Not hospitalized		1-3 times		> 3 times	
Scale	n	Mean(sd)	n	Mean(sd)	N	Mean (sd)	p-value
		n=63		n=25		n=12	
Child report;							
Total Mean score	61	88.04 ± 6.2	24	73.34 ± 9.9	12	61.33 ± 13.5	<0.001
Physical functioning	63	81.64 ± 10.2	25	64.87 ± 12.6	12	49.21 ± 16.1	<0.001
Emotional functioning	63	94.28 ± 6.8	25	90.93 ± 7.9	12	84.44 ± 16.8	<0.001
Social functioning	63	90.68 ± 9.2	25	77.60 ± 12.4	12	63.88 ± 10.4	<0.001
School functioning	61	90.05 ± 6.7	24	65.27 ± 16.3	12	47.77 ± 19.4	<0.001
							<0.001
Parent report;							<0.001
Total Mean score	61	83.05 ± 7.6	24	65.80 ± 11.3	12	51.54 ± 11.9	<0.001
Physical functioning	63	75.84 ± 9.8	25	58.75 ± 12.7	12	40.62 ± 14.3	<0.001
Emotional functioning	63	89.41 ± 6.4	25	78.40 ± 17.6	12	71.66 ± 16.1	<0.001
Social functioning	63	84.86 ± 10.4	25	73.86 ± 10.3	12	61.66 ± 10.6	<0.001
School functioning	61	82.88 ± 10.7	24	55.05 ± 16.7	12	32.22 ± 14.7	<0.001

All scale 0 – 100. A high score indicates better health

3.10. Bonferroni alpha Post hoc test of PedsQL Scale in children with SCD who were not hospitalized, hospitalized 1-3times and those who were hospitalized >3times in prior 12 months

Multiple comparison tests in children revealed significance difference between all groups except for emotion functioning were significance observed between those children who were admitted more than three times and those who were not admitted at all in prior 12 months by Bonferroni alpha Post hoc test with adjusted p value of <0.001. In parents post hoc test show statistically significant in all groups except in emotion functioning between children who were admitted one to two times and those admitted more than three times.

Table 8:Bonferroni alpha Post hoc test of PedsQL Scale in children with SCD who were not hospitalized, hospitalized 1-3times and those who were hospitalized >3times in prior. 12 months

		Child Report		Parent Report	
Scale	Comparison	Mean difference	P value	Mean difference	P value
Mean score	1−2	14.72	<0.001	17.24	<0.001
	1−3	27.73	<0.001	31.50	<0.001
	2−3	13.01	<0.001	14.25	<0.001
Physical functioning	1−2	16.77	<0.001	17.09	<0.001
	1−3	32.42	<0.001	35.21	<0.001
	2−3	15.65	<0.001	18.12	<0.001
Social functioning	1−2	13.08	<0.001	11.00	<0.001
	1−3	26.79	<0.001	23.20	<0.001
	2−3	13.71	0.01	12.20	<0.001
School functioning	1−2	24.77	<0.001	29.83	<0.001
	1−3	42.27	<0.001	50.66	<0.001
	2−3	17.50	<0.001	20.83	<0.001
Emotion functioning	1−2	3.35	0.33	11.07	<0.001
	1−3	9.84	<0.001	17.75	<0.001
	2−3	6.48	0.12	6.73	0.14

All scale 0 – 100. A high score indicates better health

1-Not hospitalized 2-Hospitalized 2-3 times 3- Hospitalized >3times

CHAPTER FOUR

4. DISCUSSION

4.1.1 Health related quality of life in SCD

In this study, PedsQL was used to evaluate Health Related Quality of Life {HRQoL} in 100 children aged 8-18years with Sickle cell disease and their parents/caretaker. HRQoL of these children was compared to arandom sample of 99 siblings who had no SCD obtained from the database in the same hospital in which the study population was obtained.

Children with SCD and their parents scored lower in each of the four subscales of the PedsQL as well as for the summary and total scores as compared to the children without SCD and their parents. This indicates that SCD is associated with lower health related quality of life as perceived by children themselves and their parents. This is in consistency with the study done by Dampier et al in USA by using the same tool (PedsQL Generic score scale) to asses Quality of life in children and adolescent with SCD. In their study,children and adolescent with SCD as well as their parents scored significantly lower on several HRQoL domains including; general physical, motor and independent daily functioning[18].

A research by McLish et al [17] in USA using SF 36 Questionnaire indicated that patients with SCD experience lower HRQoL compared to the general population. The same results were also found in other studies in USA by Panepinto et al[22] by using Child Health Questionnaire and Taylor et al[23]by using qualitative focus groups.The lower HRQoL among children with SCD may be explained by the disease itself, butmay also be attributed to the presence of numerous physical symptoms and disease related complications.

The overall HRQoL in this study as perceived by SCD children 77.53 ± 12.9 and their parent 73.79 ±14.6 were above the established PedsQL cut off scores of 69.7 in children

report and 65 in parent report[43]. This finding is in contrast with study done in Texas by Juanita et al[42] were Children with SCD and their parent scored69.10 ± 15.1 and 64.0 ± 17.7 respectively. This may be explained by the nature of the study population {children with SCD} where these children were enrolled from ongoing cohort where the level of care is high. Results could have been different if these children were from rural areas and were not involved in researches. These findings can also be explainedbycultural differences where level of social support, coping styles and perceptions of illness is different as compared to developed countries.

In this study t test was used to compare mean score between children with SCD and their parents. Results showed that children with SCD described their own HRQoL significantly better than their parents perceived it to be. Significant differences between parent and child ratings were observed in overall HRQoL and all subdomains {P <0.001}. This study is in congruency with the study done by Juanita et al[42] which used the same PedsQL questionnaire in children and adolescent with SCD.The same results were also obtained by Panepito et al in Chicago[20] and Palermo et alin Ohio using Child Health Questionnaire Parent Form (CHQ-PF50) in children with SCD[36].Possible reasons for child–parent discrepancies concerning health and well-beingmay relate to the patients themselves minimizing symptoms of distress to adapt to their illness[47] or parental distress may bias parents to over report child symptomatology[47-48].These findings can also be explained by the fact thatSCD children may not be involved fully in their evaluation and treatment or might be doing so because they do not want to be called lazy.

4.1.2 Health related Quality of Life and social demographic characteristics of SCD children.

In this study,females with SCD scored higher in all subdomains of PedsQL except for emotional functioning as compared to male gender, however this difference was not statistically significant (P- value 0.517). Some other studies in developed

countriessuggested that females have a poorer quality of life than their malecounterparts [17,18,22]. Other studies did not reveal gender difference[39].

With respect to age, the results shows that children with older age {13-18years}perceived themselves better than children aged 8-12 years with a total score of 84.40{11.8} and 76.32 {13.9} respectively {P-value 0.004}. In individual subdomainsthe significant difference was found in school and social functioning withP-value <0.001 and 0.028respectively.

The results of this study is in contrast with other studies done in developed countries were female and older age was found to correlate with greater limitations in physical and psychosocial health of children with SCD[17,18,22]. The difference in findings between this study and their studies might be due to cultural and social issues where most African children become self reliant early, so despite being sick they try to cope with general activities.

In this study, level of education of the parents had no impact in HRQoL in children with SCD as perceived in physical and psychosocial summary of child and parent report proxy {P-value 0.45 and 0.47 respectively}. Same findings were found in a study done by Palermo et al in USA on Parental report on HRQoL in children with SCD.Other studies on relationship between HRQOL and the parents education level revealed conflicting results, where a study done by Van den Tweel et al [29] in Amsterdam, Netherlands indicated that children of parents with low educational level perceived a significantly better HRQoL, while other previous research on children with chronic illness shows that high parental education associated with high quality of life scores or education had no effect on HRQoL[45,46]. In this study children with SCD were enrolled on ongoing cohort study where care and counselling is done to the children and their parents/ caretaker despite of education level, this means the awareness towards SCD Is equal to both parents.

4.1.3 Health related Quality of Life and frequency of hospitalization in children with SCD.

Significance difference in the mean score with respect to number of hospitalization was observed in this study. Children with increased frequency of hospitalization {> than 3 times} in prior 12 months scored worse in all subdomains as compared to children who were not admitted and those who were admitted less than 3 times. The overall mean score in children who were hospitalized more than three times was 61.33{13.5} in children report and 51.54{11.97} in parent report respectively, this results was below the cut off score established for PedsQL. Although it was not a primary study question, the study found that the common reasons for admission inchildren with SCD were vasoocclusive pain episode {59.46%} and severe anemia {48.65%}.Other studies by Mostafa et al[37] in Saud Arabiaand Shapiro et al[38] in USA had similar results where frequent hospital admissions were associated with lower scores in almost all subscales especially physical, role physical and emotional well-being domains andthe major reason for frequent hospitalization were vasoocclusive pain episode.This can be due to the fact that during the time of hospitalization, these children missed school and can't play with peers or participates in general activities at home which have negative impact in HRQOL.

4.2 STUDY LIMITATIONS

- This study was cross section, it evaluate the presence of association between the variables and not the temporal relationship.
- Generic rather than disease specific tool have been used this may not be able to detect significance differences of HRQoL and its correlates in children with SCD.
- These children enrolled from the ongoing cohort in the hospital with tertiary level of care which may bias our results with subsequent impact on their generalizability.

CHAPTER FIVE

5. CONCLUSION

- Children with SCD and their parents perceived overall HRQoL and all subdomains to be lower than for children without SCD.
- Ratings by parents were significant lower than those of their children in all subdomains of PedsQL questionnaire.
- Gender and parents education level had no impact on HRQoL of these children.
- Young age of the children and increase frequency of hospitalization due to disease related complications had negative impact on HRQoL.

5.1 RECOMMENDATIONS

- Assessment of HRQoL should be included in the guidelines of management of children with SCD as it will help to identify those who need psychosocial support and behavior intervention to promote appropriate functioning and to minimize activities restriction in order to improve their quality of life.

- Development of specific questionnaire which will be reliable, valid and cultural sensitive that will take into consideration nuances of SCD as a chronic illness with unique physical and psychosocial problems that differ significantly from other chronic disease.

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APPENDICES

Appendix i: Consent Form (English Version).

ID No……

Title: Health related quality of life in children aged 8-18 years with SCD attending SCD clinic at MNH in Dar es salaam, Tanzania.

To the Parents/ Guardians of ……..

Foreword

Greetings! I am…….. Working on this research project with the aim of assessing children aged 8-18 years with SCD on their health related quality of life.

Purpose of the Study

The study has a broad objective of assessing health related quality of life in children with SCD Moreover it aims at determining the association between health related quality of life outcome and frequency of hospitalization in prior 12 months. Further it explores socio-demographic characteristics that have profound effect on health related quality of life.

How to participate

The interviewer will give you/ask a questionnaire to complete some of the questions and other questions will be completed by the interviewer after signing at the end of this informed consent form.

Risks

We do not expect any harm during the course of your participation. Moreover there is no any medication or immunization provided so we do not expect any harm will happen to your child because of joining this study.

Confidentialiy

We would like to assure you that all the information that you will provide will remain confidential and will be used for research purpose only. No one will be allowed to see or go through your answers except the principle investigator only.

Consent

I have read and understood the explanation of the study. I accept for my child to be examined and participate in the study.

Signature of the Parent/Guardian .....

Relationship to the child .....

Date .....

For more information or clarification you may contact ;

Dr. Honesta 0754592201

Dr. Magdalena Lyimo 0756772662

Appendix ii: Assent Form (English Version)

ID No……

Title: Health related quality of life in children aged 8-18 years with SCD attending SCD clinic at MNH in Dar es salaam, Tanzania.

Foreword

Greetings! I am…….. Working on this research project with the aim of assessing children with SCD aged 8-18 years on their health related quality of life.

Purpose of the Study

The study has a broad objective of assessing health related quality of life in children with SCD Moreover it aims at determining the association between healthrelated quality of life outcome and frequency of hospitalization. Further it explores socio-demographic characteristics that have profound effect on health related quality of life.

How to participate

The interviewer will give you/ask a questionnaire to complete some of the questions and other questions will be completed by the interviewer after signing at the end of this informed consent form.

Risks

We do not expect any harm during the course of your participation. Moreover there is no any medication or immunization providedso we do not expect any harm to happen to you because of joining this study.

Confidentialiy

We would like to assure you that all the information that you will provide will remain confidential and will be used for research purpose only. No one will be allowed to see or go through your answers except the principle investigator only.

Subject's statement of assent

I have heard and understood the explanation of the study. My parent/guardian agrees that I can participate in this study. I have asked any questions I have about the study and my questions have been answered.

Signature of the child.....Date.....

Signature of the parent/guardian…….Date……

Signature of the investigator.....Date.....

For more information or clarification you may contact ;

Dr. Honesta 0754592201

Dr. Magdalena Lyimo 0756772662

Appendix iii: Consent Form (Swahili Version).

Namba ya fomu……

Hali ya maisha ya kiafya ya watoto wenye umri kati ya miaka 8-18 wanaoishi na sickle cell Dar es salaam, Tanzania

Kwa Mzazi/Mlezi wa ……..

Makubaliano ya kushiriki dodoso

Utangulizi

Habari! Jina langu ni……..Nafanya kazi katika huu utafiti wenye lengo la kuchunguza hali ya maisha ya kiafya ya watoto wenye umri kati ya miaka 8-18 wanaoishi na sickle cell, Dar es salaam, Tanzania**.**

Malengo ya utafiti

Utafiti huu una lengo kuu la kuangalia hali ya maisha ya watoto wenye sickle cell wenye umri kati ya miaka 8-18. Zaidi utafiti huu una lengo la kulinganisha hali ya maisha ya kiafya kwa watoto wenye sickle cell na watoto wasiokuwa na sickle cell. Pia una lengo la kuangalia Uhusiano wa hali ya maisha na mazingira anayoishi.

Jinsi ya kushiriki

Mtafiti atakuuliza maswali au atakupa fomu ya maswali kwa wewe kuijaza. Sehemu nyingine ya maswali itajazwa na mtafiti mwenyewe. Hii itafanyika mara baada ya wewe kukubali na kusaini mwisho wa fomu hii.

Madhara

Hatutegemei utafiti huu kuwa na madhara yoyote kwako au kwa mtoto wako. Hakuna dawa au kinga yoyote utakayopewa. Hivyo hakuna madhara yoyote yatayokupata wewe au mtoto wako kwa kushiriki katika utafiti huu.

Utunzaji wa Siri

Tunapenda kukuhakikishia kwamba,maelezo yote utakayotoa itakuwa siri na yatatumika kwa utafiti tu.Hakuna mtu yoyote atakaye ruhusiwa kusoma majibu yako isipokuwa mtafiti mkuu na wasaidizi wake tu.

Kukubali

Nimesoma na kuelewa madhumuni ya utafiti huu na nimekubali mimi na mtoto wangu kushiriki katika utafiti huu.

Sahihi ya Mzazi/Mlezi.....

Uhusiano na Mtoto.....

Tarehe.....

Kama una maswali kuhusu utafiti huu unaweza kuwasiliana na yoyote kati ya madaktari hawa;

Dr. Honesta 0754592201

Dr. Magdalena Lyimo 0756772662

AU endapo utakuwa na swali lolote kuhusu haki zako kama mshiriki katika utafiti huu wasiliana na:

Prof M. Aboud; Mkurugenzi wa kamati ya tafiti na matoleo chuoni. Chuo Kikuu Cha Afya na Sayansi za Tiba Muhimbili; S.L.P 65001 Dar Es Salaam.

Appendix iv: Child's Assent Form

(Toleo la Kiswahili)

Namba ya fomu……

Hali ya maisha ya kiafya ya watoto wenye umri kati ya miaka 8-18 wanaoishi na sickle cell Dar es salaam, Tanzania

Makubaliano ya kushiriki dodoso

Utangulizi

Habari! Jina langu ni……..Nafanya kazi katika huu utafiti wenye lengo la kuchunguza hali ya maisha ya kiafya ya watoto wenye umri kati ya miaka 8-18 wanaoishi na sickle cell, Dar es salaam, Tanzania**.**

Malengo ya utafiti

Utafiti huu una lengo kuu la kuangalia hali ya maisha ya watoto wenye sickle cell wenye umri kati ya miaka 8-18. Zaidi utafiti huu una lengo la kulinganisha hali ya maisha ya kiafya kwa watoto wenye sickle cell na watoto wasiokuwa na sickle cell. Pia una lengo la kuangalia Uhusiano wa hali ya maisha na mazingira anayoishi.

Jinsi ya kushiriki

Mtafiti atakuuliza maswali au atakupa fomu ya maswali kwa wewe kuijaza. Sehemu nyingine ya maswali itajazwa na mtafiti mwenyewe. Hii itafanyika mara baada ya wewe kukubali na kusaini mwisho wa fomu hii.

Madhara

Hatutegemei utafiti huu kuwa na madhara yoyote kwako. Hakuna dawa au kinga yoyote utakayopewa. Hivyo hakuna madhara yoyote yatayokupata kwa kushiriki katika utafiti huu.

Utunzaji wa Siri

Tunapenda kukuhakikishia kwamba,maelezo yote utakayotoa itakuwa siri na yatatumika kwa utafiti tu.Hakuna mtu yoyote atakaye ruhusiwa kusoma majibu yako isipokuwa mtafiti mkuu na wasaidizi wake tu.

Kukubali

Nimesikia maelezo ya utafiti.Maswali yangu yamejibiwa vema. Mzazi/Mlezi wangu amekubali nishiriki.

Sahihi ya mtoto.....Tarehe.....

Sahihi mzazi/mlezi…..……...Tarehe……

Sahihi ya mtafiti.....Tarehe.....

Kama una maswali kuhusu utafiti huu unaweza kuwasiliana na yoyote kati ya madaktari hawa;

Dr. Honesta 0754592201

Dr. Magdalena Lyimo 0756772662

AU endapo utakuwa na swali lolote kuhusu haki zako kama mshiriki katika utafiti huu wasiliana na:

Prof M. Aboud; Mkurugenzi wa kamati ya tafiti na matoleo chuoni. Chuo Kikuu Cha Afya na Sayansi za Tiba Muhimbili; S.L.P 65001 Dar Es Salaam.

Appendix v: Questionnaire (English Version)

HEALTH RELATED QUALITY OF LIFE IN CHILDREN AGED 8-12 YEARS WITH SCD IN DAR ES SALAAM, TANZANIA

I would like you to fill some of the few questions about yourself and your health. The information you will provide will help in assessing and following up of how children with SCD feel and able to do their usual activities.

(A) SOCIO-DEMOGRAPHIC CHARACTERISTICS

To be administered by an interviewer

Child full name……

Child gender ☐ Boy ☐ Girl

Child Age (Months)…….

Child Class……. ☐ Not attending Class

Residence……..

NAME OF THE PARENT/CARETAKER …….

Gender: ☐ Male ☐ Female

Relation to the child:

Biological ☐ Parent ☐ Grandparent ☐ Step Parent ☐

Other (specify)……. Caretaker ☐ Foster Parent ☐ Adoptive Parent ☐

LEVEL OF EDUCATON

No formal education ☐

Primary education ☐

Secondary education ☐

Collage education ☐

University education ☐

EMPLOYMENT STATUS OF PARENT/CARETAKER

Unemployed ☐

Employed ☐

Student ☐

Others……..

MARITAL STATUS OF THE PARENT/CARETAKER

Married ☐ Single ☐ Divorce ☐ Widowed ☐ Cohabiting ☐

Others……..

(B) THE PAEDIATRIC QUALITY OF LIFE INVENTORY

To be completed by CHILD for CHILD REPORT and PARENT for PARENT REPORT

DIRECTIONS

On the following page is a list of things that might be a problem for you.

Please tell us how much of a problem each one has been for you

during the past ONE month by circling:

0 if it is never a problem

1 if it is almost never a problem

2 if it is sometimes a problem

3 if it is often a problem

4 if it is almost always a problem

There are no rights or wrong answers.

If you do not understand a question, please ask for help.

In the past ONE month, how much of a problem has this been for you/your child

About My Health and Activities *(PROBLEMS WITH…)*	Never	Almost Never	Some-times	Often	Almost Always
1. It is hard for me to walk more than one block	0	1	2	3	4
2. It is hard for me to run	0	1	2	3	4
3. It is hard for me to do sports activity or exercise	0	1	2	3	4
4. It is hard for me to lift something heavy	0	1	2	3	4
5. It is hard for me to take a bath or shower by	0	1	2	3	4
6. It is hard for me to do chores around the house	0	1	2	3	4
7. I hurt or ache	0	1	2	3	4
8. I have low energy	0	1	2	3	4

About My Feelings *(PROBLEMS WITH…)*	Never	Almost Never	Some-times	Often	Almost Always
1. I feel afraid or scared	0	1	2	3	4
2. I feel sad or blue	0	1	2	3	4
3. I feel angry	0	1	2	3	4
4. I have trouble sleeping	0	1	2	3	4
5. I worry about what will happen to me	0	1	2	3	4

How I Get Along with Others *(PROBLEMS WITH…)*	Never	Almost Never	Some-times	Often	Almost Always
1. I have trouble getting along with other kids	0	1	2	3	4
2. Other kids do not want to be my friend	0	1	2	3	4
3. Other kids tease me	0	1	2	3	4
4. I cannot do things that other kids my age can do	0	1	2	3	4
5. It is hard to keep up when I play with other kids	0	1	2	3	4

About School *(PROBLEMS WITH…)*	Never	Almost Never	Some-times	Often	Almost Always
1. It is hard to pay attention in class	0	1	2	3	4
2. I forget things	0	1	2	3	4
3. I have trouble keeping up with my schoolwork	0	1	2	3	4
4. I miss school because of not feeling well	0	1	2	3	4
5. I miss school to go to the doctor or hospital	0	1	2	3	4

C. To be completed by an interviewer for children with SCD.

1. . Current symptoms

-Are you well today? Y/N

-Febrile illness Y/N

-Painful episode Y/N

-Symptoms of severe anemia Y/N

-Worsening of jaundice Y/N

-Cough Y/N

-DIB Y/N

-Other symptoms Y/N Specify……..

2. History of hospitalization in prior 12months;

a. 0……

b. 1-3……..

c. >3……

3. Reason for hospitalization

a. Severe anemia

b. Chest problems

c. Painful episode/ vasooclusive pain

d. Stroke

e. Avascular necrosis

f. Chronic osteomyelitis

g. Malaria

h. Leg ulcer

i. Others……..

3. History of blood transfusion in prior 12months

a. 0……

b. 1-3……

c. >3……

4. Reason for blood transfusion

a. Severe anemia

b. Stroke

c. Others… (specify)

TEEN REPORT (ages **13-18**)

Article I. **DIRECTIONS**

On the following page is a list of things that might be a problem for you.

Please tell us **how much of a problem** each one has been for you

during the **past ONE month** by circling:

0 if it is **never** a problem

1 if it is **almost never** a problem

2 if it is **sometimes** a problem

3 if it is **often** a problem

4 if it is **almost always** a problem

There are no right or wrong answers.

If you do not understand a question, please ask for help.

*In the past **ONE month**, how much of a **problem** has this been for you …*

About My Health and Activities *(PROBLEMS WITH…)*	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. It is hard for me to walk more than one	0	1	2	3	4
2. It is hard for me to run	0	1	2	3	4
3. It is hard for me to do sports activity or	0	1	2	3	4
4. It is hard for me to lift something heavy	0	1	2	3	4
5. It is hard for me to take a bath or shower by	0	1	2	3	4
6. It is hard for me to do chores around the	0	1	2	3	4
7. I hurt or ache	0	1	2	3	4
8. I have low energy	0	1	2	3	4

About My Feelings *(PROBLEMS WITH…)*	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. I feel afraid or scared	0	1	2	3	4
2. I feel sad or blue	0	1	2	3	4
3. I feel angry	0	1	2	3	4
4. I have trouble sleeping	0	1	2	3	4
5. I worry about what will happen to me	0	1	2	3	4

How I Get Along with Others *(PROBLEMS WITH…)*	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. I have trouble getting along with other teens	0	1	2	3	4
2. Other teens do not want to be my friend	0	1	2	3	4
3. Other teens tease me	0	1	2	3	4
4. I cannot do things that other teens my age can	0	1	2	3	4
5. It is hard to keep up with my peers	0	1	2	3	4

About School *(PROBLEMS WITH…)*	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. It is hard to pay attention in class	0	1	2	3	4
2. I forget things	0	1	2	3	4
3. I have trouble keeping up with my schoolwork	0	1	2	3	4
4. I miss school because of not feeling well	0	1	2	3	4
5. I miss school to go to the doctor or hospital	0	1	2	3	4

PARENT REPORT for **TEENS** (ages **13-18**)

Article II. DIRECTIONS

On the following page is a list of things that might be a problem for **your teen**.

Please tell us **how much of a problem** each one has been for **your teen**

during the **past ONE month** by circling:

0 if it is **never** a problem

1 if it is **almost never** a problem

2 if it is **sometimes** a problem

3 if it is **often** a problem

4 if it is **almost always** a problem

There are no right or wrong answers.

If you do not understand a question, please ask for help.

*In the past **ONE month,** how much of a **problem** has your teen had with.*

Physical Functioning ***(PROBLEMS WITH…)***	**Never**	**Almost Never**	**Some times**	**Often**	**Almost Always**
1. Walking more than one block	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in sports activity or exercise	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Taking a bath or shower by him or herself	0	1	2	3	4
6. Doing chores around the house	0	1	2	3	4
7. Having hurts or aches	0	1	2	3	4
8. Low energy level	0	1	2	3	4

Emotional Functioning ***(PROBLEMS WITH…)***	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad or blue	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying about what will happen to him or her	0	1	2	3	4

Social Functioning **(PROBLEMS WITH…)**	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. Getting along with other teens	0	1	2	3	4
2. Other teens not wanting to be his or her friend	0	1	2	3	4
3. Getting teased by other teens	0	1	2	3	4
Not able to do things that other teens his or her age can do	0	1	2	3	4
5. Keeping up with other teens	0	1	2	3	4

School Functioning **(PROBLEMS WITH…)**	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. Paying attention in class	0	1	2	3	4
2. Forgetting things	0	1	2	3	4
3. Keeping up with schoolwork	0	1	2	3	4
4. Missing school because of not feeling well	0	1	2	3	4
5. Missing school to go to the doctor or hospital	0	1	2	3	4

Appendix vi: Questionnaire (Swahili Version)

HALI YA MAISHA YA KIAFYA YA WATOTO WENYE UMRI KATI YA MIAKA 8-18 WENYE SICKLE CELL DAR ES SALAAM, TANZANIA

Ningependa kukuuliza maswali machache kuhusu wewe/mtoto wako mwenyewe na afya yako/yake.Taarifa utakazotoa zitasaidia katika kufatilia jinsi mtoto wako unavyojisikia na jinsi gani unaweza kufanya shughuli zako za kila siku.

SEHEMU YA KWANZA: TAARIFA BINAFSI

Sehemu hii ijazwe na msaili.

Jina kamili la motto ……

Jinsia ya mtoto ☐ Mme ☐ Mke

Umri wa mtoto (Miezi)…….

Darasa……. Haendi Shule

Mahali anapoishi

Nyumbani kwao ☐

Nyumba za kulea watoto ☐

Jina la Hospitali/kituo …….....

JINA LA MZAZI/MLEZI:

Jinsia: Mme ☐ Mke ☐

Uhusiano na mtoto:

Mzazi wa mtoto ☐ Mzazi wa kufikia ☐ Bibi/Babu ☐

Mzazi wa kisheria ☐ Mzazi wa pembeni ☐ Mlezi ☐ Mengineyo ☐ (jaza)……

ELIMU YA MZAZI/MLEZI

1.Sijasoma

2.Elimu ya msingi

3.Elimu ya sekondari ☐

4. Elimu ya chuo ☐

5. Elimu ya chuo kikuu ☐

KAZI YA MZAZI/MLEZI

Ameajiriwa ☐

Hajaajiriwa ☐

Mwanafunzi ☐

Mengineyo.....

HALI YA NDOA YA MZAZI/MLEZI

Ameolewa Pekee Ameachika

Mjane Anaishi bila ndoa Mengineyo.....

SEHEMU YA PILI: PAEDIATRIC QUALITY OF LIFE INVENTORY

IJAZWE NA MTOTO kwa fomu ya MTOTO, MAMA kwa fomu ya MAMA

MWONGOZO

Katika ukurasa ufuatao kuna orodha ya vitu ambavyo vinaweza kuwa tatizo kwako..

Tafadhali tueleze ni kwa kiasi gani kila tatizo linavyokuathiri kwa muda wa mwezi mmoja uliopita kwa kuzungushia moja wapo

:

0 Kama hakujawahi kuwa na tatizo

1 Kama tatizo la nadra sana

2 kama una tatizo

3 kama una tatizo mara kwa mara

4 kama muda wote ni tatizo

Hakuna majibu yaliyo ya sawa au yaliyo ya makosa Kama hauelewi swali, tafadhali uliza

Usaidizi

Kwa mwezi mmoja uliopita,ni kiasi gani ya shida imekupata …

kuhusu afya na mazoezi *(SHIDA NA…)*	Hakuna	Karibia hakuna	Wakati mwengine	Kila mara	Kwa wakati wote
1. Ni vigumu kwangu kutembea zaidi ya nyumba .	0	1	2	3	4
2. Ni vigumu kwangu kukimbia	0	1	2	3	4
3. Ni vigumu kwangu kufanya michezo au mazoezi	0	1	2	3	4
4. Ni vigumu kwangu kuinuwa kitu kizito	0	1	2	3	4
5. Ni vigumu kwangu kuoga mwenyewe	0	1	2	3	4
6. Ni vigumu kwangu kufanya shughuli katika	0	1	2	3	4
7. Naumia au nina maumivu	0	1	2	3	4
8. Sina nguvu	0	1	2	3	4

kuhusu hisia zangu *(SHIDA NA…)*	Hakuna	Karibia hakuna	Wakati mwengine	Kila mara	Kwa wakati wote
1. nina hisi uoga au kuhofia	0	1	2	3	4
2. nina huzuni	0	1	2	3	4
3. nina hisi hasira	0	1	2	3	4
4. ninapata shida kulala	0	1	2	3	4
5. nina hofu kuhusu kile kitakacho nipata	0	1	2	3	4

vile ninavyo elewana na wengine *(SHIDA NA…)*	Hakuna	Karibia hakuna	Wakati mwengine	Kala mara	Kwa wakati wote
1. Nina shida ya kuelewana na watoto wengine	0	1	2	3	4
2. Watoto wengine hawataki kuwa rafiki zangu	0	1	2	3	4
3. Watoto wengine wananichokoza	0	1	2	3	4
4. Siwezi kufanya vitu ambavyo watoto wenzangu wanafanya	0	1	2	3	4
5. Nashindwa kuendelea na mchezo wakati nacheza na watoto wenzangu	0	1	2	3	4

kuhusu shule *(SHIDA NA…)*	Hakuna	Karibia hakuna	Wakati mwengine	Kila mara	Kwa wakati wote
1. Ni vigumu kuelewa darasani	0	1	2	3	4
2. Nina sahau mambo	0	1	2	3	4
3 Nina shida na kufanya kazi za shule	0	1	2	3	4
4. Nakosa kwenda shule kwa sababu ya kuumwa	0	1	2	3	4
5. Nakosa kwenda shule kwa sababu naenda kwa dokta au hospitali	0	1	2	3	4