Research Article

Quality of life assessment for children with sickle cell disease (SCD) in Mecca region

Emad Abdulaziz Alharbi[1], Roa'a Faiz Alamri[2], Ebtisam Saud AlJerayan[1], Hadeel Suliman Salawati[2], Shroog Mohammad Al-Mjershi[2]

[1]Medical Intern Graduate, Umm-Alqura University, Mecca, Saudi Arabia.
[2]Nursing Student, Umm-Alqurra University, Mecca, Saudi Arabia.
Correspondence to: Emad Abdulaziz Alharbi, E-mail: dremadz91@gmail.com

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Abstract

Background: Sickle cell disease (SCD) is an autosomal recessive condition characterized by production of abnormal hemoglobin S and is related to high morbidity and mortality. It also affects the quality of life (QOL) of patients and their relatives/caregivers.

Objective: To assess QOL for children with sickle cell anemia.

Materials and Methods: This descriptive study was conducted in pediatric hematological departments in selected hospitals at Makkah Al-Mukarramah. A purposive sample composed of 40 pediatric children with SCD with their mothers was involved in this study according to specific criteria. Different questionnaires were used for data collection.

Result: The most commonly affected age group was of 10–12 years, with boys more involved in the disease. It was found that most of the children always found hard to take a shower, do chores around the house, and do sports. It was also found that children with sickle cell anemia always felt afraid or scared. It was clear that the majority of the children with SCD were always facing problems such as other children did not want to be their friends, having trouble getting along with kids, and other kids tease them. Children always have trouble keeping up with the schoolwork. Overall, all the parameters of QOL are affected in the children with SCD, particularly physical and social well-being.

Conclusion: Most of the subcategories of the QOL, and physical, social, emotional, and school well-being were affected by SCD. There must be adequate measures included in standard treatment guidelines for SCD to improve QOL in children with SCD.

KEY WORDS: Sickle cell disease, quality of life, children, physical well-being, emotional well-being

Introduction

Sickle cell disease (SCD) is an autosomal recessive genetic disorder. It is characterized by a chronic hemolytic anemia that leads to painful crises and chronic inflammation. It also leads to oxidative stress, decreased oxygen affinity of sickled hemoglobin, disordered sleep-induced fatigue, and cognitive impairment.[1] SCD is a serious health problem and chronic hereditary disorder, which affects the red blood cells. This disease occurs predominantly in people who live in parts of the world where malaria has long been endemic. Sickle cell anemia (SCA) exists among Nigerians, Afro-Americans, and Caucasians, living in Greece, Italy, Turkey, Iran, and North Africa.[2] SCD is a common disease present throughout in Saudi Arabia, with a high prevalence in the Eastern and Southern regions. The prevalence of SCD in the various regions is reported to be as follows—Qatif (Eastern region): 0.170, Gizan (Southern region): 0.103, Mekka (Western region): 0.025, and Alula (Northern region): 0.081. In Madina, the estimated prevalence of sickle cell homozygosity (Hb SS) is 0.01,

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and that of the carrier state (Hb AS) is 0.087.[2] In Khaiber, a neighboring area, the gene frequency of Hb S is 0.239.2.[2] SCD was detected in 108 of 45.682 children and adolescents, with the prevalence of 24 per 10.000. The regional distribution of SCD showed Eastern region dominance with prevalence of 145 per 10.000, followed by the Southern region with a prevalence of 24 per 10.000, Western region 12 per 10.000, and Central region six per 10.000.[3]

Onset of SCD occurs during the first year of life. Chronic hemolysis produces jaundice, pigment (calcium bilirubinate) gallstones, splenomegaly (early in life), and poorly healing ulcers over the lower tibia. Life-threatening episode of hemolytic crisis may trigger by viral or other infection or by folic acid deficiency causing reduced erythropoiesis. Acute painful episodes occur owing to acute vaso-occlusion by clusters of sickled red cells. This can be provoked by infection, dehydration, or hypoxia. Vertebrae and long bones are generally involved in acute episodes. The episode lasts from hours to days and may produce low-grade fever. Acute vaso-occlusion may be present as a life-threatening event such as cerebrovascular stroke. Splenic sequestration, liver congestion, lung and heart injuries, leg ulcers, and aseptic and bone infarcts may also occur.[4]

The aims of therapy for the SCD include the management and prevention of the acute manifestations and to block the clustering the red blood cells; last, but not least, to educate the patients and their family members regarding the disease. There is no single line of treatment defined to combat the anemia. Children and their parents may be adversely affected as the disease cannot be managed effectively, which may lead to disharmony in the family, the deterioration of the quality of life (QOL), loss of energy, and increasing care burden and despair.[5]

The WHO has defined the health as a state of complete physical, mental, and social well-being and not merely absence of disease or infirmity.[6] QOL is a crucial feature of mental and social well-being that is frequently overlooked. It is the individuals' opinions of their life in the perspective of their culture and value systems to which they belong and in connection with their goals, expectations, standards, and concerns.[6] SCD is a disease that can affect the QOL not only of patients but also their caregivers. Assessment of QOL in these children is significant to know how and to what level their QOL is affected. This study was undertaken to assess QOL in children with SCD.

Materials and Methods

This descriptive study was conducted in pediatric hematological departments. A purposive sample composed of 40 children with SCD with their mothers which was fulfilling the following inclusion and exclusion criteria was included.

Inclusive Criteria

- Children with SCA.
- Children aged 6–12 years.

Exclusive Criteria

- Children with SCA and other disease such as thalassemia.
- Children with splenectomy.

Tools of Data Collection

Three tools were used for data collection as follows:

1. *Interviewing Questionnaire*
   It consisted of two parts:
   - First part: Caregivers' sociodemographic characteristics: including their age, gender, educational level, etc.
   - Second part: children' sociodemographic characteristics: including age, gender, school level, etc.

2. *Pediatric QOL Inventory (PedsQL)*
   It was adopted by Varni.[7] It was used to assess children QOL.

3. *Pain Scale*
   It was classified into two parts:
   - First part: Pediatric Pain Questionnaire adopted by Varni[7] used to assess pain for children with SCA and identify locations on their body.
   - Second part: Wong-Baker FACES Pain Rating Scale[8] was used to assess pain intensity among children

The actual field work was carried out throughout 4 months from the beginning of August to the end of November 2014. The nature and purpose of the study were explained to caregivers and their children. Written informed consents were taken from patients and their relatives/caregivers before enrolling them into the study. Ethical approval was taken before conducting the study.

Result

This study showed that children of 10–12 years of age (38%) were most affected when compared with other age groups [Figure 1]. Boys (67.5%) were more affected when compared with the girls [Figure 2]. Table 1 shows QOL for children with SCD regarding their physical health. It was found that most of the children (72.5%, 70.0%, and 67.5%) found it always hard to take a shower, do chores around the house, to do sports, respectively. With regard to QOL for children with SCD regarding to their mental health, it was found that that 72.5% of children with SCA always felt afraid or scared [Table 2]. It was clear that the majority of the children with SCD were always facing problems such as other children did not want to be their friends, having trouble getting along with kids, and other kids tease them (97.5%, 92.5%, and 92.5%, respectively) [Table 3]. If we analyzed the school well-being of the children with SCD, it was found that 72.5% children always have trouble keeping up with the schoolwork [Table 4]. Overall, all the parameters of QOL are affected in the children with SCD, particularly physical and social well-being [Table 5].

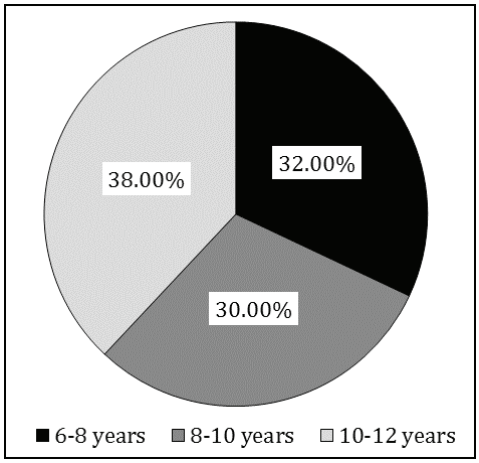

Figure 1: Distribution of children with sickle cell according to age.

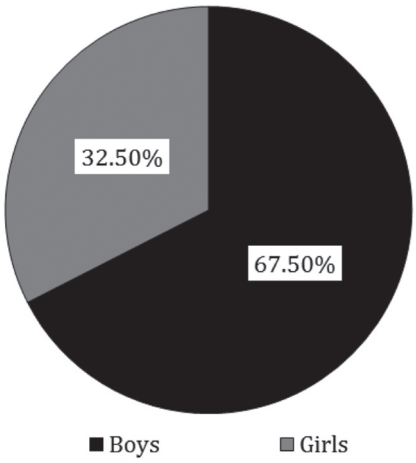

Figure 2: Percentage distribution of children with SCA according to gender.

Table 1: Distribution of the study sample according to QOL (physical health)

Statements	SCA children						χ^2	*p*
	Always		Sometimes		Never			
	n = 40	%	*n* = 40	%	*n* = 40	%		
It is hard for me to walk	26	65	10	25	4	10	19.40	0.000**
It is hard for me to run	22	55	11	27.5	7	17.5	9.05	0.011*
It is hard for me to do sports activity	27	67.5	5	12.5	8	20	21.35	0.000**
It is hard for me to life something heavy	23	57.5	8	20	9	22	10.55	0.005**
It is hard for me to take a shower by myself	29	72.5	4	10	7	17.5	27.95	0.000**
It is hard for me to do chores around the house	28	70	5	12.5	7	17.5	24.35	0.000**
I hurt or ache	15	37.5	19	47.5	6	15	6.65	0.036*
I have low energy	11	27.5	19	47.5	10	25	3.65	0.161

*$p < 0.05$; ** $p < 0.001$.

Table 2: Distribution of the study sample according to QOL (emotional well-being)

Statements	SCA children						χ^2	*p*
	Always		Sometimes		Never			
	n = 40	%	*n* = 40	%	*n* = 40	%		
I feel afraid or scared	29	72.5	8	20	3	7.5	28.55	0.000**
I feel sad	18	45	16	40	6	15	6.20	0.045*
I feel angry	22	55	14	35	4	10	12.20	0.002**
I have trouble sleeping	22	55	13	32.5	5	12.5	10.85	0.004**
Worry about what will happen to me	20	50	14	35	6	15	22.20	0.000**

*$p < 0.05$; ** $p < 0.001$.

Table 3: Distribution of the study sample according to QOL (social well-being)

Statements	SCA children						χ^2	*p*
	Always		Sometimes		Never			
	n = 40	%	*n* = 40	%	*n* = 40	%		
I have trouble getting along with other kids	37	92.5	1	2.5	2	5	63.05	0.000*
Other kids do not want to be my friend	39	97.5	0	0	1	2.5	36.10	0.000*
Other kids tease me	37	92.5	1	2.5	2	5	63.05	0.000*
I cannot do things that other kids at my age can do	24	60	9	22.5	7	17.5	12.80	0.002*
It is hard to keep up when I play with other kids	27	67.5	10	25	3	7.5	12.95	0.002*

*$p < 0.05$; ** $p < 0.001$.

Table 4: Distribution of the study sample according to QOL (social well-being)

Statements	SCA children						χ^2	*p*
	Always		Sometimes		Never			
	n = 40	%	*n* = 40	%	*n* = 40	%		
It is hard to pay attention in class	27	667.5	10	25	3	7.5	43.4	0.000**
I forget things	22	55	14	35	4	10	29.0	0.000**
I have trouble keeping up with my schoolwork	29	72.5	6	15	5	12.5	49.0	0.000**
I miss school because of not feeling well	9	22.5	10	25	21	52.5	18.2	0.000**
I miss school to go to the doctor or hospital	5	12.5	4	10	31	77.5	70.5	0.000**

*$p < 0.05$; ** $p < 0.001$.

Table 5: Distribution of study sample according to total QOL

Statements	SCA children						χ^2	*p*
	Always		Sometimes		Never			
	n = 40	%	*n* = 40	%	*n* = 40	%		
Physical well-being	32	80	5	12.5	3	7.5	26.30	0.010*
Social well-being	32	80	7	17.5	1	2.5	56.60	0.000**
Emotional well-being	28	70	7	17.5	5	12.5	20.50	0.015*
School well-being	16	40	15	37.5	9	22.5	22.15	0.014*

*$p < 0.05$; ** $p < 0.001$.

Discussion

Children with SCD require specialized medical care to obtain good QOL. Nurses use their potential skills for healing children with their families in managements; therefore, this study was carried out on 40 children and their mothers to assess QOL for children with SCA

This study revealed that nearly equal percentage of one-third of the SCD children aged from 6 years to less than 8, from 8 years to less than 10 years, and from 10 years to 12 years, respectively was involved in the study. This finding was supported by another study done by Sadarangani et al.[9] in Killifish, Kenya. In addition to this, another study done by Patel and Pathan[10] in Nagpur, India, reported the mean age of SCD children involved in his study was 9.5 years.

Boys were more affected than girls in this study. These findings were in contradiction to study done by Constantinou et al.[11] In another study carried out by Kamble and Chatruvedi[12] found that both the gender are equally affected in children, but significant gender differences in morbidity and mortality have been reported in adults with SCD.

On analysis of QOL for physical health subcategories for children with SCD, it was found that most of the children always felt hard to take a shower, do chores around the house, and to do sports. These findings were supported by the study done by Elsayed and El-Gawad SMEA.[13] Similar findings were observed in a study carried out by Asnani et al.[14] This study has used the WHOQOL-BREF criteria for measuring QOL in SCD and found that children with SCA scored less in physical activity and daily functioning independently.[14] Physical functioning was observed significantly poor in SCD patients. This is the reason for more medical consultations for SCD cases when compared with patients of other chronic diseases.[15]

With regard to QOL for children with SCD regarding their mental health, it was found that that 72.5% of children with SCA always felt afraid or scared. Similar results have been observed in different studies as well.[13,16] According to the study of Bhagat et al.,[15] role limitation owing to emotional problems and fatigability were significantly higher in SCD patients.[15]

In our study, it was clear that the majority of the children with SCD always faced issues such as other children did not want to be their friends, having trouble getting along with kids, and other kids tease them. Again these findings are very well supported by other studies.[13,16]

Concerning school achievements of children with SCA, the results of this study found that more than half of the study sample was hardly being attention in class and the children forgot things. Approximately, three quarters of children with SCA had trouble keeping up with their schoolwork. Half of the study sample never missed school because of not feeling well, and above three quarters never missed school to go to the doctor or hospital. There were statistically significant differences in all school health subitems. These findings were supported by Sadarangani et al.,[9] who stated that excessive absence of children with SCA is owing to the complications of the disease and, sometimes, affects a child's ability to meet up with class work regarding all social well-being subitems.[9]

Overall general health of SCD children was poor in our study. This study demanded the requirement of palliative care for physical health and functioning. Addressing social domains of QOL (emotional support and social dysfunction), counseling, and rehabilitative services are recommended to maintain and improve QOL.

Conclusion

It was concluded from this study that SCD affected the QOL of children with it. Most of the subcategories of the QOL, physical, social, emotional, and school well-being were affected by SCD. There must be adequate measure included in standard treatment guidelines for SCD to improve QOL in children with SCD.

Recommendations

- Future research studies needed with larger samples of SCA and on other age groups.
- Improving awareness of parents about importance of premarital examination and genetic counseling services.
- Developing educational program for parent with their children with SCA regarding risks factors, precipitating factors, and prevention of sickle cell crises.
- Improving awareness of SCD and its clinical manifestations for parents, which help to improve QOL of children

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