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Original Research Paper

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HEALTH RELATED QUALITY OF LIFE OF CHILDREN WITH SICKLE CELL DISEASE IN VIDARBHA REGION OF MAHARASHTRA, INDIA

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ABSTRACT

Introduction: Sickle cell disease (SCD) is a chronic hemolytic anemia and most common manifestation is acute painful crises. Health related Quality of life (HRQL) is an important feature of mental and social well-being that is frequently overlooked during assessment of morbidity of chronic diseases like sickle cell disease. This study is planned to assess quality of life of children with sickle cell disease (SCD) in Vidarbha region of Maharashtra, India.

Methods: This was a cross sectional study conducted in the Government Medical College and Hospital, Akola, Maharashtra, India. Total 50 children with SCD in age group of 6-12 years were enrolled. Pediatric Quality of life inventory was used to assess children HRQL. It includes physical well-being, social well-being, emotional well-being and school well-being.

Results: Out of 50 enrolled subjects, 30 were male and 20 were female. In physical well being, most common item (60%) was "It is hard for me to run", in emotional well-being it was "I feel angry", in social well-being it was "I cannot do things that other kids at my age can do" and in school well-being it was "I forget things" and "I miss school because of not feeling well". In 48% of subjects HRQL was affected as we observed "Always" choice.

Conclusion: Health related Quality of life of children with SCD is affected in all subcategories. Along with treatment of SCD, appropriate interventions should be done to prevent, diagnose and treat impairment in HRQL.

KEYWORDS : quality of life, scale, sickle cell disease.

Introduction

Sickle cell disease (SCD) is an autosomal recessive genetic disorder. It is characterized by a chronic hemolytic anemia and most common manifestation is acute painful crises. [1] SCD is a lifelong chronic disease. This disease occurs predominantly in people who live in parts of the world where malaria is endemic. Sickle cell anemia (SCA) exists among Nigerians, Afro-Americans, and Caucasians.

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. [2] Health-related quality of life (HRQL) is defined as the patient's appraisal of how his/her well being and level of functioning, compared to the perceived ideal, are affected by individual health. [3] Quality of life (QOL) is an important feature of mental and social well-being that is frequently overlooked during assessment of morbidity of chronic diseases like sickle cell disease.

Sickle cell disease can affect the HRQL not only of patients but also of their caregivers. Assessment of HRQL in these children is significant to know how and to what level their HRQL is affected. In India, sickle cell disease (SCD) is common in Vidarbha, Chhattisgarh, Madhya Pradesh, Orissa, Gujarat, Tamil Nadu and Andhra Pradesh. Contrary to the earlier belief, it is commonly seen in non tribal population in central India. [4] Many patients of SCD in central India have relatively severe manifestations. This may result from lower frequencies of alpha thalassemia and more frequent severe sickle cell-beta (+) thalassemia. [5] SCD is common in Vidarbha region of Maharashtra [6] and no recent study from this region revealed about HRQL in SCD.

Hence, this study is planned to assess health related quality of life of children with sickle cell disease (SCD) in Vidarbha region of Maharashtra, India.

Methods:

This was a cross sectional study conducted from July 2017 to October 2017 in the Department of Pediatrics, Government Medical College and Hospital, Akola, Maharashtra, India. Children with SCD in age group of 6-12 years attending pediatrics outdoor patient department were enrolled in the study. Diagnosis of SCD was done by high performance liquid chromatography. Children with sickle cell trait, SCD with other chronic disease, psychiatry disorder and intellectual disability were excluded. Total 50 children (30 male and 20 female) were enrolled in the study after written informed consent from parent. Institutional ethical committee approval was taken before start of study.

Two tools were used for data collection in the study as follows:

1. Interviewing Questionnaire: Children sociodemographic characteristics like Age, gender and school level
2. Pediatric QOL Inventory (PedsQL): It was adopted by Varni.et al [7] and was used to assess children HRQL. It includes physical well-being, social well-being, emotional well-being and school well-being.

Choices available for questionnaire were always, sometimes and never. Questionnaire was translated into Marathi and again translated into English. Numerical data presented as mean and percentages.

Result:

Out of 50 enrolled subjects, 30 were male and 20 were female. In physical well being, most common item was "It is hard for me to run" in 60% of subjects and least common was "It is hard for me to take a shower by myself". [Table 1] In emotional well-being, most common item was "I feel angry" in 54% of subjects and least common was "I have trouble sleeping". [Table 2] In social well-being, most common item was "I cannot do things that other kids at my age can do" in 54% of subjects and least common was "I have trouble getting along with other kids". [Table 3] In school well-being, most common item was "I forget things" and "I miss school because of not feeling well" in 52% of subjects. [Table 4] In 48% of subjects HRQL was affected as we observed "Always" choice. [Table 5]

Discussion:

Sickle cell disease is a chronic hemolytic anemia. Morbidity and mortality due to SCD is described in recent studies from central India. But, there are very few studies describing about HRQL in SCD. Many physicians and medical researchers have begun to focus on more patient-centered methods of disease assessment by incorporating measurement of HRQL. A study from central India by Patel AB et al [8] used various QOL scales like Patrik and Erickson, EUROQOL and WHO QOL BREF to develop QOL for SCD children. In our study, we used Pediatric QOL inventory by Varni et al as it includes physical well-being, social well-being, emotional well-

being and school well-being. A recent study from Saudi Arabia [9] used Pediatric QOL inventory by Varni et al and found that QOL of SCD is affected in all subcategories of QOL. We also found similar observations. A recent case control study from India by Bhagat VM et al [10] used WHO SF- 36 Questionnaire and found that QOL in SCD was lower than controls who were suffering from other chronic illness. In most of the studies, QOL is obtained from child self-report. However, recent study by Constantinou C et al [11] used child self-reports, parent-proxy reports, and health care professional-proxy reports. A study from Nigeria [12] reported psychosocial problems in patients of SCD and their mothers also. So, interventions to improve HRQL should target to complete family of SCD patient. Relation of adherence to hydroxyurea and HRQL has been studied and found low HRQL in SCD patients with poor adherence to hydroxyurea. [13] Health related QOL can be assessed in infants with SCD by asking questionnaire to parents, as described in recent study. [14] All subcategories of HRQL scores need to be more accessible and understandable to healthcare providers and have to be considered during health education, counseling and treatment. Along with treatment of SCD children, interventions should be done to prevent, diagnose and treat impairments in HRQL.

This was the cross sectional study and changes in HRQL over time was not studied. We have not taken into consideration the severity of SCD, as this may have impact on HRQL. Future study should find out changes in HRQL with change in health and validity of Pediatric QOL Inventory scale.

Conclusion:

Health related Quality of life of children with SCD is affected in all subcategories like physical well-being, social well-being, emotional well-being and school well-being. In 48% of subjects HRQL was affected as we observed "Always" choice. Along with treatment of SCD, appropriate interventions should be done to prevent, diagnose and treat impairment in HRQL.

Table 1: Distribution of the subjects according to QOL (physical well being)

Statements	Sickle cell anemia children					
	Always		Sometimes		Never	
	n=50	%	n=50	%	n=50	%
It is hard for me to walk	21	42	16	32	13	26
It is hard for me to run	30	60	10	20	10	20
It is hard for me to do sports activity	26	52	12	24	12	24
It is hard for me to lift something heavy	24	48	15	30	11	22
It is hard for me to take a shower by myself	18	36	18	36	12	24
It is hard for me to do chores around the house	22	44	16	32	12	24
I hurt or ache	22	44	15	30	13	26
I have low energy	28	56	12	24	10	20
Mean	24	48	14	28	12	24

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

Statements	Sickle cell anemia children					
	Always		Sometimes		Never	
	n=50	%	n=50	%	n=50	%
I feel afraid or scared	21	52	16	32	13	26
I feel sad	24	48	15	30	11	22
I feel angry	27	54	14	28	9	18
I have trouble sleeping	20	40	15	30	15	30
Worry about what will happen to me	22	44	17	34	11	22
Mean	23	46	15	30	12	24

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

Statements	Sickle cell anemia children					
	Always		Sometimes		Never	
	n=50	%	n=50	%	n=50	%
I have trouble getting along with other kids	23	46	17	34	10	20
Other kids do not want to be my friend	25	50	16	32	9	18
Other kids tease me	26	52	14	28	10	20
I cannot do things that other kids at my age can do	27	54	13	26	10	20
It is hard to keep up when I play with other kids	24	48	17	34	9	18
Mean	25	50	15	30	10	20

Table 4: Distribution of the subjects according to QOL (school well-being)

Statements	Sickle cell anemia children					
	Always		Sometimes		Never	
	n=50	%	n=50	%	n=50	%
It is hard to pay attention in class	24	48	15	30	11	22
I forget things	26	52	14	28	10	20
I have trouble keeping up with my schoolwork	21	42	15	30	14	28
I miss school because of not feeling well	26	52	12	24	12	24
I miss school to go to the doctor or hospital	18	36	15	30	17	34
Mean	23	46	14	28	13	26

Table 5: Distribution of subjects according to total QOL

Statements	Sickle cell anemia children					
	Always		Sometimes		Never	
	n=50	%	n=50	1%	n=50	%
Physical well-being	24	48	14	28	12	24
Social well-being	23	46	15	30	12	24
Emotional well-being	25	50	15	30	10	20
School well-being	23	46	14	28	13	26
Mean	24	48	14	28	12	24

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