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Epidemiological aspect and quality of life of Children with Sickle cell disease

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Abstract

Background:*Sickle cell disease (SCD) is the most frequent genetic disease in the world**. Complications like a physical, emotional and social impact on the carrier's life that can compromise their quality of life (QOL). The study aimed to assess the quality of life and epidemiological aspect of children with sickle cell disease.**Methods:**It was across sectional study which conducted in outpatient Clinic and Pediatric Hematological Department in Zagazig university hospital Including30 children with sickle cell anemia and their parents who fulfill the following criteria, A- age ranges from 5 to18 years. B- free from any associated diseases. We used two tools, first was: Structure interview questionnaire sheets and second: was Pediatric Quality of Life Inventory that was used to assess children's quality of life. **Results:**Mean age of studied children 10.65±3.41 years,66.7% of them were males. Regarding to the level of education, it was found that 46.7% of children were in primary schools, 20% of them were in preparatory school and (33.3%) in secondary school. The birth order of children was the first for 40%, the second for 33% and the third and more for 26.7%. Most of children were in the rural area73.3% and 26.7% of them were in the urban. Affected members in the family with SCD occurred in 53.3%. 63.3% had history of positive consanguinity. Characteristics of children's mothers are presenting, it was clear the mean of age was35.93±6.03 years Ranged From 21 Years And 45 years. Concerning level of education, it was noticed that nearly one third of mothers (33.3%) were illiterate, 20% of them had basic education and 20% of them had diploma. While 26.7% of mothers had high education (University). It was seen that 76.6% were housewife and 23.3% of them were working. 66.7% was SCA,20% was SC trait, and 13.3%was β thalassemia. Concerning to the clinical data as reported by studied children, it was found that the onset of illness was in the first year for 66.7% of children, while it was in the second years for 30% of them. 73.3% of the studied children had pallor and jaundice appeared in 63.3% of them.10% of them had splenomegaly while 6.7% had hepatomegaly. Concerning operations 3.3% had splenectomy and 3.3% had cholecystectomy. VOC of studied patients, 40% had mild pain, 23.3% moderate pain and 36.6 had severe pain .26.6%. of pain affected abdomen, 53.4% taking NSAID, 63.3%of studied patient did not visit pain clinic. According to assessment of QOL we found that overall QOL 36.7% were poor and 63.3% were good.**Conclusion:** Sickle cell disease is a chronic disease which had a negative impact on QOL that include physical, emotional, social and school functioning. Our result reflected that all aspects of QOL were affected especiallyschool functioning.*

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I. Introduction:

Sickle cell disease (SCD) is the most frequent genetic disease in the world**,** with Sickle Cell Anemia (SCA) i.e. homozygous sickle cell disease or Hb SS, and to a lesser extent hemoglobin SC disease (SC), reaching the highest prevalence. It is estimated that over 300,000 children are born each year with a severe inherited hemoglobinopathy, over 80% of these in low –or middle –income countries, and approximately 220,000 newborns are affected by SCA**(1).**

The cause of this disease is a genetic mutation at position 6 of the beta chain of hemoglobin, the component of red blood cells that is responsible for transporting gases in the blood **(2).**

Sickle Cell Anemia is caused by homozygosis for the sickle gene, the RBCs have an abnormal crescent sickle shape that makes them sticky and rigid. The loss of red blood cell elasticity is central to the pathophysiology of SCA. Normal RBCs are quite elastic, which allows the cells to deform and pass through capillaries. In SCA, low oxygen tension promotes red blood cells sickling and repeated episodes of Sickling damage the cell membrane and decrease the cells elasticity. These cells fail to return to normal shape when normal oxygen tension is restored. As a consequence, these rigid blood cells are passing through narrow capillaries leading to vessel occlusion and ischemia. The actual anemia of the illness is caused by hemolysis and destruction of the red cells because of their shape. Although the bone marrow attempts to compensate by creating new red cells, It does not match the rate of destruction, healthy RBCs typically function for 90–120 days, but in SCA only last for 10–20 days. **(3).**

A distortion of the shape of red blood cells which start presenting a sickle shape and become more rigid can cause vaso-occlusive complications, reduced oxygenation, progressive tissue dehydration and reductions in organic functions **(4).**

People with sickle cell anemia present several signs and symptoms that can occur in different ways and at different severities; they occur jointly or separately. These include chronic anemia, painful crises, infection, fever, jaundice, splenic sequestration (retention of blood in the spleen), leg ulcerations, priapism and stroke **(5).**

These complications have a physical, emotional and social impact on the carrier's life that can compromise their quality of life (QOL). **(5).**

World Health Organization (WHO) defines QOL as "an 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" Evaluation of QOL by Recognition of Certain Events in people's life such as health status, cognitive function, daily activities, emotional well-being and family and social life **(6).**

The study aimed toassess the quality of life and epidemiological aspect of children with sickle cell disease.

II. Patients and Methods

A) Technical design

Site of the study: Outpatient Clinic of pediatric hematology unit of zagazig university hospital.

Time of the study: 6 months from September 2019 till March 2020.

Sample size:

The study was conducted on a sample of 30 children with sickle cell disease and their parents comprised the study subjects and full fill the following criteria:

Inclusion criteria:

a-age from 5-18 years.

b-both sexes.

c-Informed consent to enter the study.

Exclusion criteria:

a-age less than 5 years and more than 18 years.

b- Pateint with other hemolytic anemias.

B) Administrative design:

The study was presented to the (IRB) of zagazig faculty of medicine for its approval; informed consent was obtained from parents of every patient.

C) Operational design

Study design: Cross sectional study.

Tools of data collection

Two tools were used to collect the necessary data for this study.

Tool I: Structure interview questionnaire sheets.

It was included the demographic data of children; age, gender, level of education, birth order and residence.

Parents demographic data; age, level of education, occupation, and income.

The children's clinical data comprised the onset, duration and type of treatment of sickle cell disease such as blood transfusion: frequency and Side effect of blood transfusion, and compliance with chelating therapy.

Tool II: Pediatric Quality of Life Inventory TM Version 4.0 by **Varni et al.** [7]

Scoring system for assessment of quality of life[7]:

The Peds TM4.0 Generic Core Scales are comprised of parallel children self- report and parent proxy-report formats. The instructions ask how much a problem each item has been during the past 1 month.

- 0=I never have a problem.

- 1=I almost never have a problem.
- 2=I sometimes have a problem.
- 3=I often have a problem.
- 4=I almost always have a problem.

Items are reverse scored linearly transformed to a 0-100 scale as following

- 0=100(quality of life is very good=not affected QOL)
- 1=75(quality of life is good=mildly affected QOL)
- 2=50(quality of life is fair=moderately affected QOL)
- 3=25(quality of life is bad=severely affected QOL)
- 4=0(quality of life is very bad=severely affected QOL)
- This means that the higher the score indicate better HRQOL.

1) Methods

All patients were subjected to the following:

A-Complete history taking with special emphasis on:

B-Thorough clinical examination.

C-Laboratory investigation:

- Complete blood count. (CBC).
- Serum ferritin level.
- Hemoglobin Electrophoresis.

D- Specific investigation:

Every child and his mother were interviewed individually for 30-40 minutes to collect the necessary data .The researcher introduced her self and explained the purpose of the study briefly to them.

- Collect data using; Tool 1;Structure interview questionnaire sheets.
- And Tool II: Pediatric Quality of Life.
- Of all subjected patient and their parents.

Statistical Analysis: Data collected throughout history, basic clinical examination, laboratory investigations and outcome measures coded, entered and analyzed using Microsoft Excel software. Data were then imported into Statistical Package for the Social Sciences **(SPSS version 20.0)(**Statistical Package for the Social

Sciences) software for analysis. According to the type of data qualitative represent as number and percentage, quantitative continues group represent by mean ± SD.

III. Results:

Table (1): Demographic data distribution among studied patients. (N=30):

		Mean	Median (Range)	
Age		10.65±3.41	9.0(5-18)	
Weight		30.95±9.87	27.0(16-56)	
Height		128.23±20.13	124.0 (104-169)	
BMI		18.12±2.71	17.84 (14.42-24.3)	
			N=30	**100%**
Sex	●	Female	10	33.3
	●	Male	20	66.7
Level of education	●	Primary	14	46.7
	●	Preparatory	6	20.0
	●	Secondary	10	33.3
Birth order	●	1st	12	40.0
	●	2nd	10	33.3
	●	3rd and more	8	26.7
Residence	●	Rural	22	73.3
	●	Urban	8	26.7
Income	●	Insufficient	6	20.0

	● Sufficient	18	60.0
	● Sufficient & more	6	20.0
Similar condition	● No	14	46.7
	● Yes	16	53.3
consanguinity	● No	11	36.7
	● Yes	19	63.3

This table illustrates; mean age of studied children10.65±3.41years, 66.7% of them were males ,mean BMI of them is 18.12±2.71, 46.7%, of them in primary school,73.3% of studied children in rural area, 53.3% of studied group had similar condition in their family and consanguinity found in 63.3%.

Table (2): Characteristics of Children's Mothers:

	Mean=SD	**Median(range)**	
Mother's age	35.93±6.03	35.0 (21-45)	
		N=30	**100 %**
Mother's education	Illiterate	10	33.3
	primary	6	20.0
	secondary	6	20.0
	University	8	26.7
Mother's occupation	Working	7	23.3
	Housewife	23	76.6

This table illustrates; mother's age was distributed as 35.93±6.03years, 76.7% were housewives. Concerning level of education, it was noticed that 33.3% illiterate.

Table (3): Types of SCD among studied patients. (N=30):

Variant	N	%
SCA	20	66.7
SC trait	6	20
HbSβ^0 thalassemia	2	6.7
HbS/β^+ thalassemia	2	6.7

Concerning types ofSCD, 66.7% is SCA,

Table (4): Clinical presentation of SCD patients: (N=30):

		N=30	100%
Onset of disease	0-1year	20	66.7
	2-3year	9	30
	3year	1	3.3
Pallor	No	8	26.7
	Yes	22	73.3
Jaundice	No	19	63.3
	Yes	11	36.7
Enlarged liver	No	28	93.3
	Yes	2	6.7
Enlarged spleen	No	27	90.0
	Yes	3	10.0
Operations	splenectomy	1	3.3.0
	cholecystectomy	1	3.3.0
VOC Frequency	Less than3 per year	14	46.7

	3 or more per year	16	53.4

This table shows; 73.3% have pallor, 63.3% have jaundice and rarely manifested of enlarged liver6.7% and enlarged spleen (10%) respectively.

66.7% in disease appears in first years.

Table (5): Vaso Occlusive Crisis distribution among studied patients: (N=30):

		N =100	**100%**
Pain intensity	Mild	12	40.0
	Moderate	7	23.3
	Sever	11	36.6
Site	All body	4	13.4
	Abdomen	8	26.6
	Extremities	6	20.0
	Extremities + abdomen	7	23.3
	Chest	3	10.0
	Back	2	6.7
Treatment	Paracetamol	10	33.3
	NSAID	16	53.4
	Opiate analgesic	4	13.3
Frequency of pain clinic visit	No	19	63.3
	every week	5	16.7
	every month	5	16.7
	Every 3month	1	3.3

Table (5) shows VOC of studied patients, 40% had mild pain, 26.6%. of pain affected abdomen, 53.4% taking NSAID, 63.3%of studied patient did not visit pain clinic.

Table (6): Blood transfusion data of studied patients (N=30):

		N	%
Packed RBCs	+VE	15	50.0
	-VE	15	50.0
Frequency blood transfusion; n=15	Every3m	10	66.7
	Every6m	5	33.3
Received treatment	Hydroxyurea	30	100.0
	L-carnitine	30	100.0
	Folic acid	30	100.0
	Iron chelation	15	50.0
	Vit E	3	10.0
	L.A.Penicillin	1	3.3
Types iron chelating therapy; n=15	Deferasirox	9	60.0
	desferroxamin	6	40.0
Complication of blood transfusion; n=15	Rash-itching	4	26.7
	Incompatibility	2	13.3

Table (6) shows 50% of studied children had regular blood transfusion. 66.7% of children who received blood transfusion come to hospital once per 3 months for transfusion therapy, while 33.3% received blood transfusion once per 6 months. 100% of studied group take Hydroxyurea-carnitine and folic acid, 3.3% received long acting penicillin and 50% of studied patients on chelating therapy.40% of Patients who received blood transfusion had complications.

Table (7): Quality of life assessment distribution among studied group according to questionnaire:

		N	%
Physical aspect	**Good**	23	76.7

	Poor	7	23.3
Emotional aspect	**Good**	15	50.0
	Poor	15	50.0
Social aspect	**Good**	19	63.3
	Poor	11	36.7
School aspect	**Good**	14	46.7
	Poor	16	53.3
Total QOL	**Good**	19	63.3
	Poor	11	36.7

This table show;23.3% were poor regard physical aspect, 50% were poor regard emotional aspect, 36.7% were poor regard social aspect, 53.3% were poor regard school aspect and overall QOL 36.7% were poor and 63.3% were good.

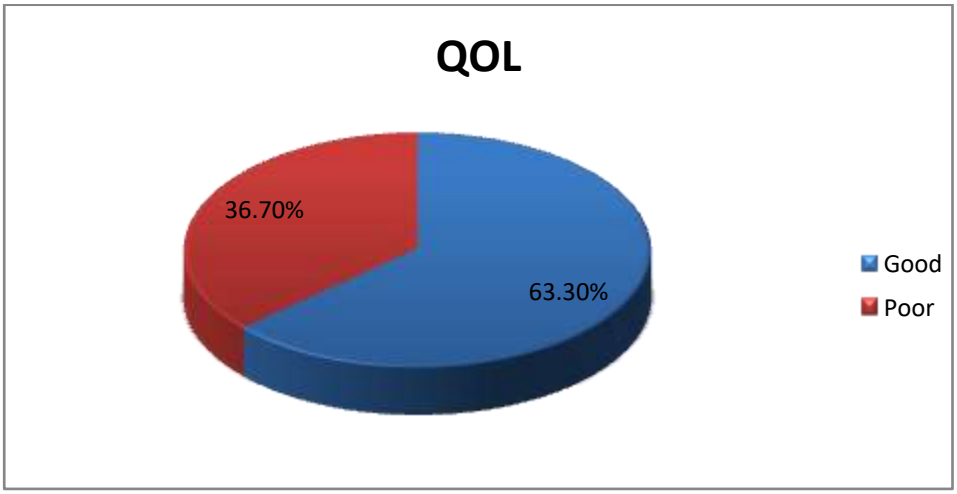

Fig. (1): Shows 63.3% of total QOL is good and 36.7% is poor

IV. Discussion

The mean age of SCD patients in our study is 10.65±3.41years and this is in agreement with **Patel and Pathan**[8] in Nagpur, India and**Sadarangani et al.,** [9]**.** They reported the mean age in their studies was to be 9.5 years.

Our study showed that males (66.7%) and females (33.3%).

Kamble and Chatruvedi[10] found that both sexes are equally affected in children, but significant differences in morbidity and mortality have been reported in adult with SCD**. Constantinou et al.,** [11]disagree with our study as female were76.7%. Our finding is supported by another study done in Macca region in Saudi Arabia that males are 67.5% and females are 32.5%.

Our study showed that 73.3% of studied group from rural area. Rural residence was a significant negative predictor for vitality and pain of Health-related quality of life (HRQOL) subscales. There are limited studies on rural/ urban differences in QOL among patients with SCD.

Telfair et al., [12] have shown that rural patients had lower health service access and utilization with lower physical functioning.

Consanguinity has a significant role in the continuity of SCA inheritance. Our study showed that 63.3% of the sickle cell patients have a positive history of consanguinity. Our results are in agreement with**Alhamdan et al.,** [13]he reported about 90% of high-risk relatives complete their marriage despite knowing the risk of disease inheritance. The percentage was higher (98%) in the study conducted by **Al-Sulaiman et al., in** [14].**Hazzazi, et al.,** [15] in Jizan region in Saudi Arabia found that 75.8% had positive history of consanguinity.

As regards to the personal characteristics of the studied mothers' patients, the current study revealed that mean patient mothers age, is 35.93±6.03 years This finding was supported by the finding of a similar study by **Ezenwosu et al.,** [16]**,** who reported that, the age of the studied patient mothers between 30 -39 years.

As regards to educational level of studied mothers' patients, our study showed that nearly 33.3%of mothers are illiterate, 20% basic, 20% had secondary education and 26.7 % had university education. **Tunde-Ayinmode.,** [17], found that quarter of mothers had secondary education. On the other hand, **Almutairi et al.,** [18], found nearly one third of mothers had bachelor education.

Our study revealed that, more than three quarters of studied mothers were housewife. This finding agreed with **Daak et al.,** [19] who found that three quarters of studied mothers were unemployed. On the other hand, this finding was disagreement with **Kayle et al.,** [20], who found that more than three quarters of studied mothers were employed.

Our study found that genotypes of sickle cell diseases are sickle cell anemia is 67%, 20% is sickle cell trait and 13% is sickle cell HbS/β^+ thalassemia. This finding of present study goes in line with the finding of [21]**.**

Matthew and Russell [22]found that, sickle cell pain is worse where its intensity or severity episodes vary widely in children It is involved the bone pain as in humerus, femur, and vertebrae that is presenting with acute tenderness, swelling, and fever. These will affect children's QOL whereas it hinders their (social interaction, communication skills and physical activity.

Montalembert et al., [23] stated that all children had pain and swelling in bone of legs and arms indicating vaso-occlusive phenomena.

According to the experienced pain in our patients, 40% of children had mild pain, 23.3% had moderate pain and36.6% had severe pain.13.4% of them were often feeling pain on their whole body. These results are in agreement with **Hamed,** [24] who found that children with SCD had sever and moderate pain (46% and 54% respectively).

In our study we used the WHOQOL-BREF criteria for measuring QOL in SCD and found that children with SCD scored positive 76.7% in physical activity, 50% in emotional aspect, 63.3%social aspect,46.4% in school aspect, and the total QOL assessment is 63.3%.

Concerning physical aspect of QOL in our study, our children had no problem with self-care followed by usual activities and mobility such as take shower, do chores around the house, and to do sports. This findings are not in agreement with study of[21;25].

V. Conclusion:

Sickle cell disease is a chronic disease which had a negative impact on QOL that include physical, emotional, social and school functioning. Our result reflected that all aspects of QOL were affected especiallyschool functioning.

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