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Assessment of Health-Related Quality of Life and Muscular Strength in Children with Beta Thalassemia

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ABSTRACT
Background: Thalassemia is an inherited blood disorder that requires regular blood transfusions and chelation therapy. These treatments can lead to restrictions in physical activities, social participation, and muscle strength. Quality of life, as measured by individuals' own perceptions of their well-being and other aspects of life, can be significantly affected by thalassemia.
Objective: To assess quality of life related to health, muscular strength and pain in children with beta thalassemia.
Methods: We studied 60 children with beta thalassemia, aged 1 to 15 years. The quality of health-related life, including physical, emotional, and social performance, was assessed. Muscle strength was evaluated for all children using a standardized questionnaire. Both patients and their parents completed the questionnaire at baseline.
Results: The study results showed that a significant number of participants (86.6%) had a family history of thalassemia. The majority (76.6%) had hemoglobin levels above 65%, and 88.3% required splenectomy. Most participants (83.3%) received chelation therapy, and 81.6% underwent monthly blood transfusions. Regarding psychological effects, 61.6% were affected, with 63.3% experiencing difficulty breathing and pallor. In terms of quality of life, 98.3% suffered from physical functioning issues, 78.3% from emotional problems, 73% from social difficulties, and 70% from pain and muscle weakness.
Conclusion: The study concluded that thalassemia, a chronic disease, has a negative impact on the health-related quality of life (HRQoL) and muscle strength of children in different age groups.

Keywords: Quality of life, Muscular strength, Children with beta thalassemia.

Background

Thalassemia is an inherited blood disorder which is genetic production of hemoglobin, a molecule found inside all Red Blood Cells (RBCs) that transports oxygen throughout the human body [1]. Thalassemia is a global public health problem affecting both developing and developed countries with major consequences for human health as well as social and economic development. As per the World Health Organization (WHO) database on anemia globally, anemia affects 1.62 billion people (95%), which corresponds to 24.8% of the population [2]. Beta-thalassemia is an inherited hemoglobin disorder in the beta-globin chain that results in chronic hemolytic anemia. Two major forms of beta-thalassemia have been identified according to their clinical severity. Thalassemia major manifests with severe anemia in the first year of life in newborns, while thalassemia intermediate (TI) Involves clinically asymptomatic mild of anemia. In developing and underdeveloped countries, patients with TM die in childhood and adolescence due to the lack [3], Thalassemia on physical health can lead to physical deformity growth retardation, and delayed puberty. Its impact on physical appearance, e.g., bones deformities and short stature, also con- tributes to a poor self-image. Severe complications such as heart failure, cardiac arrhythmia, liver disease, endocrine complications, and infections are common among thalassemia patients. These problems do not only affect patients' physical functioning but also their emotional functioning, social functioning and school functioning, leading to Impaired health-related quality of life (HRQOL) of the patients [4]. Children with thalassemia are less active than their healthy peers and generally have decreased muscle strength and flexibility. Pain has become increasingly common and an emergent complication of thalassemia. The exact mechanism of pain In thalassemia has not yet been clarified; however, Iron overload, low hemoglobin level, and low bone mass have been suggested as possible causes. The most frequent sites of pain were the lower back (82%), the leg (56%), head (48%), and mid-back 47%. [5]. Thalassemia is an autosomal recessive congenital disease. Deficiencies in globin chain synthesis may lead to severe anemia requiring regular blood transfusions and iron chelation therapy starting at an early age. Despite the advances made in treatment over the past decades, many patients with beta- thalassemia major, especially

those living in developing countries, do not have access to conventional and/or innovative treatment approaches life such as education, free-time, physical activities, skills, capabilities and family adjustment, the effects of which often result in anxiety, isolation and depression [6], Hence, the primary aim of this study was to assess quality of life related to health, muscular strength and pain in children with beta thalassemia.

METHODS AND MATERIALS

A Descriptive Cross-Sectional Study of beta thalassemia in thalassemia Children at Al-Kut Hospital Women and Children. This study aimed to investigate the prevalence and characteristics of beta thalassemia among children attending Al-Kut Hospital Women and Children. A non-probability purposive sampling technique was used to select 60 thalassemia children as participants. A questionnaire was developed by the investigator based on a review of relevant literature and previous studies. Data collection was conducted using the questionnaire, and the collected data were analyzed using SPSS version 26. Ethical approval for this study was obtained from the higher health institute in Wasit.

RESULTS

Distribution according to the ages of children with thalassemia, they accounted for 40%, fall between the category of 11 to 15 years, the males constituted slightly more than the females, 53.3% and 4637% for females, 78.8% were enrolled in schools, and 21.2% were not enrolled in schools, while the most of them were in housing 65% lived in the urban, and the remaining 35% lived in the rural **(Table 1).**

Table 1: Distribution of the Children According to their Socio-demographic characteristics.

Socio-demographic characteristics Variables		Frequency	Percent
Agegroupbyyears	1 – 5	14	23.3
	5 – 10	22	36.7
	11 – 15	24	40.0
Sex	Male	32	53.3
	Female	28	46.7
Residence	Urban	39	65.0
	Rural	21	35.0
Total		60	100.0
Education	Attendingschool	26	78.8
	Non–attendingschool	7	21.2
Total		33	100.0

Clinical of the thalassemia characteristics of the participants in the study made clear, most of them have had a family history of injury thalassemia reality 86.6% and the percentage of haemoglobin was mostly more than 65% , 76.6% are those who needed to surgical interference to the splenectomy, The majority have had the regularity of the transfer blood by 88.3% recurrence of blood transfusions and found no difference between the blood transfusion within two weeks or every three weeks, and the reality of 51.6% to 48.3% , Most of them used chelation therapy, 83.3% fell. As for the psychological state of the children, 61.6% were affected, and 63.3% were suffering from difficulty breathing and pallor of the face, and most of them by 81.6% were affected by blood transfusions every month**(Table 2).**

Table 2: Clinical characteristics of the thalassemia children.

Clinicalcharacteristics		Frequency	Percent
Family history of thalassemia	Yes	52	%86.6
	No	8	%13.3
Hb	<7g/dl	1	%1.6
	7–9g/dl	20	%33.3
	>9g/dl	39	%65
History of splenectomy	Yes	46	%76.6
	No	14	%23.3
Blood transfusion	Regular	53	%88.3
	Irregular	7	%11.6

Frequency of transfusion	Every2weeks	31	%51.6
	Every3week	29	%48.3
Useofchelationtherapy	Yes	50	%83.3
	No	10	%16.6
Psychical Health	Yes	37	%61.6
	No	23	%38.3
Dyspnea and face Pallor	Yes	38	%63.3
	No	22	%36.6
Effect transfusion blood monthly	Yes	49	%81.6
	No	11	%18.3
Total		60	100.0

The quality of life for children evaluate according by domains following was mostly suffering from physical functioning were 98.3% and emotional state affected by the reality of 78.3% as well as social functioning affected were 73% and finally Pain and strong muscle were 70% suffered m showed that in **(Table 3 and** Figure 1).

Table 3: over view of studies evaluating of (HRQOL) in pediatric patients with thalassemia.

HRQOL domains		Frequency	Percent
Physical functioning	No	1	1.7
	Yes	59	98.3
Emotional functioning	No	13	21.7
	Yes	47	78.3
Social functioning	No	16	26.7
	Yes	44	73.3
Pain and strong muscle	No	18	30.0
	Yes	42	70.0

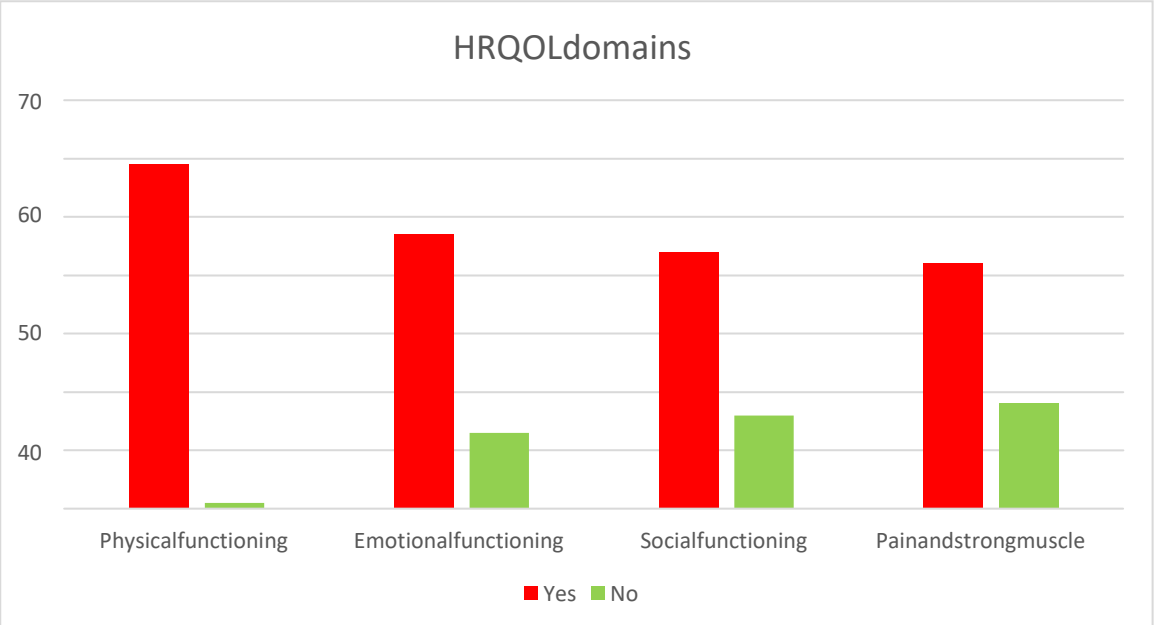
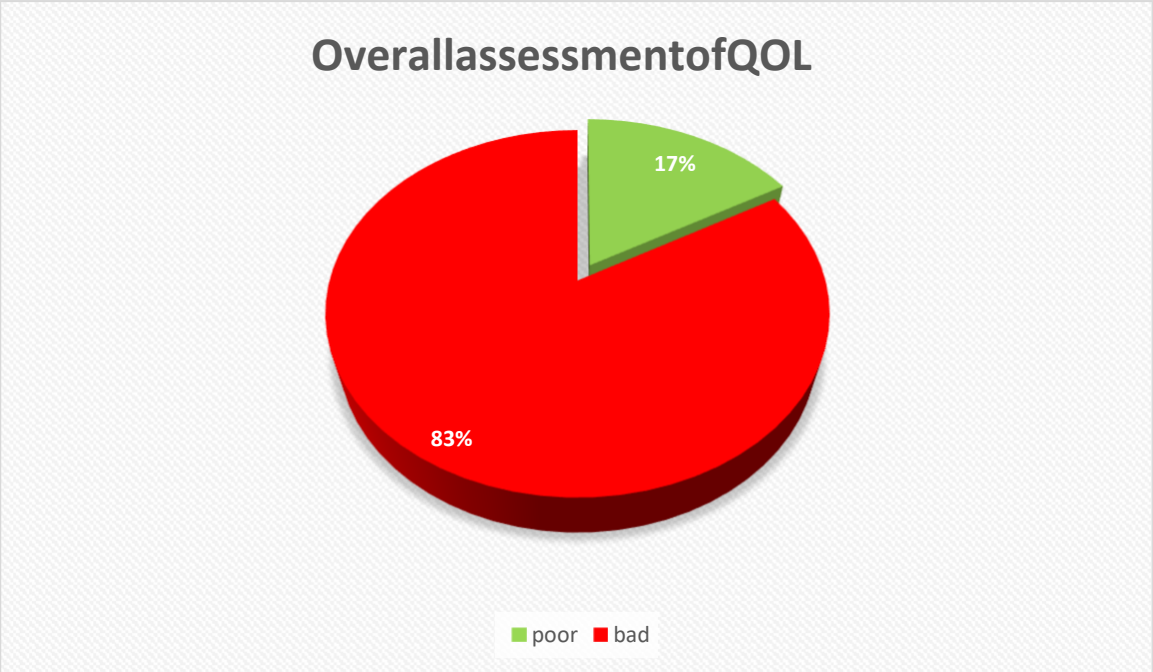


Figure 1: over view of studies evaluating of (HRQOL) in pediatric patients with thalassemia.

Appeared the final assessment of thalassemia quality of life was 83% whose bad affected and 17 whose lives were poor of quality of life **(Figure 2).**



The relationship between the final assessments shows the QOL and age groups of the participant it in the bad QOL were getting worse as their age increased. 100.0 affected by bad QOL in age 44-15 years ,note that between the ages from 5 to 10% they were affected by 81.8%, their QOL was bad at the ages of 5 to 10 years .and 57.1 at the age of less than five years, and it. Showed a very high relationship between their QOL and age groups of children with thalassemia **(Table 4).**

Table 4: the relationship between overall assessment of QOL and age groups.

			Overall assessment ofQOL		Total	P.Value
			Poor	Bad		
Agegroup	1 - 5	F.	6	8	14	0.003
		%	42.9%	57.1%	100.0%	
	5 - 10	F.	4	18	22	
		%	18.2%	81.8%	100.0%	
	11 - 15	F.	0	24	24	
		%	0.0%	100.0%	100.0%	
Total		F.	10	50	60	
		%	16.7%	83.3%	100.0%	

There are no significant relationship between the QOL and the Sex. If they are male or female, their bad life is noticed. The same for males and females 87.5 vs. 18.6. This indicates that there is no relationship or significant difference and that the QOL affects all Sex equally **(Table 5).**

Table 5: the relationship between overall assessment of QOL and Sex.

			Overall assessment of QOL		Total	P.Value
			Poor	Bad		
Sex	Male	F.	4	28	32	0.355
		%	12.5%	87.5%	100.0%	
	Female	F.	6	22	28	
		%	21.4%	78.6%	100.0%	
Total		F.	10	50	60	
		%	16.7%	83.3%	100.0%	

School age children consisted of 33 children of whom 26 had completed their education and the remaining 7 did not complete their education, 84% of those who completed or did not complete were affected by their bad QOL

and there is no non significance between those who completed their education or did not complete and Overall assessment of QOL **(Table 6).**

Table 6: the relationship between overall assessment of QOL and Education level.

			Overall assessment of QOL		Total	P.Value
			Poor	Bad		
Educationlevel	Educated	F.	4	22	26	0.943
		%	15.4%	84.6%	100.0%	
	Uneducated	F.	1	6	7	
		%	14.3%	85.7%	100.0%	
Total		F.	5	28	33	
		%	15.2%	84.8%	100.0%	

Quality of life and their relationship to Residence was almost equal reality 87.2% and 76.2% were living between the urban and rural had bad QOL, so there are no significant between the two variables **(Table 7).**

Table 7: the relationship between overall assessment of QOL and Residence.

			Overall assessment of QOL		Total	P.Value
			Poor	Bad		
Residence	Urban	F.	5	34	39	0.276
		%	12.8%	87.2%	100.0%	
	Rural	F.	5	16	21	
		%	23.8%	76.2%	100.0%	
Total		F.	10	50	60	
		%	16.7%	83.3%	100.0%	

Quality of life and their relationship to Frequency of transfusion was almost equal reality 80.6% and 86.2% were between Every 2 weeks and 3 week had bad QOL, so there are no significant between the two variables **(Table 8).**

DISCUSSION

The most of our participants aged between 11-15 years and all of them have a bad QOL score where the about half of age group of 1-5 years have a bad QOL score. The symptoms of beta thalassemia major occur when an infant is between 6 and 24 months include poor growth and development, our results agree with study done by [7], entitled(Thalassemia beta. The early onset of illness resulting in accumulation of tissue iron and it is the leading cause of morbidity and mortality in thalassemia, our results agree with study done by [8], entitled (Pain over time and its effects on life in thalassemia). This could explain the bad QOL scour in present study 11-15 cohort.

This finding was males were infected with beta thalassemia at a rate close to that of females in the current study, because this condition causes both males and females to inherit the relevant gene mutations equally because it follows an autosomal pattern of inheritance with no preference for Sex, Our results agree with study done by [9], entitled (A descriptive study to assess the health related quality of life in terms of knowledge and attitude among patients with thalassemia attending in selected hospitals at Meerut with a view to develop information booklet). The genetic origin of this illness is clear in our result where it has high correlation with family history.

The same regarding QOL where there was no significant difference between male and female. However, a research shows that QOL in particularly bone disease and it consequences resulting from thalassemia was in male patients more frequently than females, Our results agree with study done by [10], entitled(Sex differences in the prevalence and severity of bone disease in thalassaemia). Similarly other study show that female affected more by depression resulting from thalassemia.

Several reasons have been proposed for this Sex difference, including genetic predisposition, psychological factors, and increased vulnerability to stress, Our results agree with study done by [11], entitled (Examining depression and quality of life in patients with thalassemia in Sri Lanka. International Journal of Noncommunicable Diseases). Controversially a research shows that female patients had a better quality of life than males significantly, our results agree with study done by [12], ,Education level of thalassemia patients in our study appear not influences QOL of these patients. A study mentions that higher education may enhance QOL of thalassemia patients.

The poor education concluded to worsen the QOL of thalassemia patients our results agree with study done by [13], entitled (Quality of life in thalassemia major). The reason that may affect our result regarding similarity of

QOL that we have divided the participant in two main categories educated and uneducated and also may resulting from small number of uneducated participants.
The recommended treatment for thalassemia major involves lifelong regular blood transfusions, usually administered every two to five weeks, to maintain the pretransfusionhemoglobin level above 9–10.5 g/dl, Our results agree with study done by [14], entitled (Guidelines for the Clinical Management of Thalassaemia). In our study, QOL of thalassemia patient who's received blood transfusion every two weeks or every three weeks was similar which indicate good analytical approach to estimate the requirement of blood transfusion.
Most of resent study patient use chelation therapy (83.3%) to reduce accumulation of Iron. This element overload β-thalassemia major who receive frequent blood transfusions. The iron excess is resulting in life-threatening complications, namely cardiopathy, liver and endocrine dysfunction and reduced patient's survival. Iron excess also increases cell concentration of iron-binding proteins such as ferritin and hemosiderin complexes in lysosomes. The use of chelating therapy extend lifespan mainly by reducing heart disease Our results agree with study done by [15], entitled (Iron chelation therapy in thalassemia syndromes).
Many patients with thalassemia major require splenectomy. However, optimal clinical management from the time of diagnosis may delay or even prevent hypersplenism, thereby increasing the efficiency of transfusion therapy and reducing the need for splenectomy. Throughout the care of the patient with thalassaemia, the size of the spleen should be carefully monitored on physical examination and as needed, by ultrasonography; Our results agree with studies done by [14, 16], entitled (Guidelines for the clinical management of thalassaemia) and trail for nurses . That's explaining why most of our thalassemia patients participate in survey had a splenectomy.
In conclusion the present study was conducted to investigate the effect of β- thalassemia on QOL. The results suggested that β-thalassemia had a significantly negative effect on physical health status, so assessing QOL was recognized as an indispensable indicator for a general evaluation of patients with thalassemia through which valuable evidence could be provided to improve treatments and to make effective decisions. Increase longevity and life expectancy in these patients could be achieved via therapies such as splenectomy and access to the drug such as chelating agents which have possible improvement of QOL. The lower QOL could be due to inappropriate evaluation of QOL.

CONCLUSION AND RECOMMENDATION

Thalassemia is a chronic disease that negatively impacts children's health-related quality of life (HRQoL) and muscle strength. Children with thalassemia are less active, have decreased muscle strength and flexibility, and experience muscle pain. This condition affects their emotional, social, and school functioning, leading to impaired HRQoL. Thalassemia is inherited and can run in families, with those with relatives from certain parts of the world having a higher risk. Treatment includes regular blood transfusions and splenectomy. Iron overload can cause hyperpigmentation, and pallor evaluation is recommended for anemia detection.
Thalassemia patients require regular blood transfusions to maintain a high level of hemoglobin and prevent symptoms like poor growth and bone deformities. Iron deposition in organs can lead to complications, such as heart muscle weakness and liver and sex gland issues. Marriages with beta thalassemia can be unsafe, and iron-containing medicines should be avoided. Despite regular blood transfusions, patients can study and work normally with proper opportunities.

Declarations

Ethical approval and consent to participate

We obtained ethical clearance from the Ethical Review Committee of the AL-Kut University College.

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Consent for Publication

Not applicable.

Conflict of Interest

The author declares the absence of any other conflict of interest.

Author's contribution

EAK wrote a manuscript, data, designed a study, supervised the data collection and reviewed the manuscript.

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