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Quality of life among adolescents with hemoglobinopathies

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***Abstract*---**Background: Hemoglobinopathies constitute the commonest recessive monogenic disorders worldwide, and the treatment of affected individuals presents a substantial global disease burden. Objectives: The purpose of this study is to assess the quality of life among adolescents with hemoglobinopathies in Karbala City/ Iraq. Methods: The cross-sectional study with quantitative approach was carried out on 200 participants who were attending hereditary blood disease center in Karbala teaching hospital for children. The questionnaire's dependability was established through a pilot study, and it was subsequently presented to experts for validation. The information was collected through the use of interview techniques and evaluated using descriptive and inferential statistical analysis. Results: the results of current study indicate that physical function related to quality of life was demonstrate that (42.5%) of adolescent are experienced moderate, emotional function related to quality of life was demonstrated that (36.5%) of adolescent are experienced moderate, and quality of life among Hemoglobinopathies adolescents was demonstrate that (60.5%) are experienced moderate quality of life with average of M ± SD=47.63±24.38. Conclusion: The study concluded that two fifth of adolescent have moderate physical function, One third of adolescent have moderate emotional function, Two fifth of adolescent have moderate social function.

***Keywords*---**Quality of Life, Adolescent, Hemoglobinopathies.

Introduction

Hemoglobinopathies (a collective term for severe inherited blood disorders) are the most common life threatening, monogenic disorders in the world, with

approximately 5% of the world's populations are carriers for hemoglobinopathies. The most common causes of inherited hemolytic anemia are sickle cell disease (SCD) and thalassemia. SCD and thalassemia are inherited blood disorders, passed from parents to children through altered hemoglobin genes [1].

Correspondingly, [2] added that Hemoglobinopathies are anemias that result from a structural or quantitative abnormality involving hemoglobin. These alterations range in severity from clinically insignificant to profound and life-threatening disease. Sickling disorders are hemoglobinopathies in which a structural alteration in the hemoglobin molecule causes red blood cells to assume a sickle shape under certain circumstances. Hemoglobin S is an example of a structurally abnormal hemoglobin. Its presence instead of the normal hemoglobin A is associated with the most common sickling disorder, hemoglobin SS disease.

In addition to, thalassemia is a complex heterogeneous group of diseases ranging from serious anemia to clinically asymptomatic individuals. It occurs due to diminution generation of hemoglobin. There are two fundamental sorts of thalassemia, Alpha thalassemia occurs when the imperfection occur in alpha globin gene while Beta thalassemia happened when the defect occur in beta globin gene .β-Thalassemia is characterized by the reduced or absent production of β-globin chains in the hemoglobin molecule leading to an excess of α-globin chains. α-Thalassemia is characterized by the reduced or absent synthesis of α-globin chains (3).

Hemoglobinopathies has a negative impact on quality of life due to the effects of the disease and its treatment, not only affecting children physical function but also their social, emotional, and school function, leading to an impaired quality of life. The most commonly affected domains previously reported were feelings of depression, anxiety, psychological problems, emotional burden, hopelessness, difficulty with social integration, and school problems [4].

Thalassemia and sickle cell disease are important public health problems in Middle Eastern countries, including Iraq. These hemoglobinopathies are either due to reduction in the globin chain synthesis or to the production of abnormal chains (e.g. sickle cell dis-orders). In Iraq, β-thalassemia is rather uniformly distributed throughout the country with an average carrier rate of around 4% and ~ 15,000 registered patients with thalassemia major/intermedia. Sickle cell disorders, on the other hand, are less uniformly distributed with carrier rates varying from 0 to 16.0%, and they cluster in the extreme north and south of the country [5] .

The World Health Organization has reported that, Thalassemia has affected up to 270 million people globally, and that the Mediterranean region has 15 to 25 million individuals carrier [6]. About 6-10% of the population in Iraq had hemoglobinopathy of which Thalassemia is a major part [7].

Methods

Cross-sectional descriptive study design is conducted in Karbala City among adolescent with haemoglobinopathies to investigate Impact of Hemoglobinopathies

upon Quality of Life of Adolescents. By a non-probability (purposive) samples of 200 subjects who attending hereditary blood disease center.

Instruments
The instruments underlying the study phenomenon deals with the following:

Socio-demographic characteristics
A-This include age, gender, marital status, occupation and income.
This section includes data about the child's age, gender, class, residency ,the child's birth order in the family…
B- Parent's demographic data
This part concerned with the parent 's demographic data; parent 's educational level, occupation, family income, family number, and residence …)

Quality of Life Questionnaire
Using specific scales to measure the variable of the present study quality of life questionnaire. The authors created a specific tool based on an extensive literature analysis. Three domains make up the assessment instrument:
PedsQL is tool recommended for quality of life assessment because it fulfills several criteria for such a valid tool. It has high validity and reliability (has been proven in several studies in children with malignancies, diabetes and heart disease), is available in generic and for cancer, asthma, diabetes, rheumatic heart disease, cerebral palsy, epilepsy and thalassemia), can be filled by children (self-report) or parents/guardians (proxy report), has been translated to several languages to facilitate usage, and is available for various age group: 2-4 years (proxy report), 5-7 years (self and proxy report), 8-12 years (self and proxy report), as well as 13-18 years (self and proxy report). This tool consists of several versions (version 1.0 until 4.0) with 23 questions in 4 domains of assessment: physical, emotional, social and school domains. The 4.0 version is a generic measures tool. In PedsQL, the quality of life of children with chronic illness may be compared to healthy/normal children as a control. In this part there are (23) items questions which included (4) domains of the quality of life such as the physical, emotional , social, and school of adolescent children with haemoglobinopathies, which described as the following:

Validity

Validity was given to a panel of 15 arbitrators, including nursing science experts. Arbitrators were asked to offer their opinions and suggestions on each of the study questionnaire's components in terms of language appropriateness, association with the dimension of study variables to which it was assigned, and suitability for the study population. These experts were asked to review the questionnaire for content clarity, relevancy and adequacy; their responses indicated that minor changes should be done to few items. Such changes were made according to their suggestions and valuable comments.

Reliability

Reliability was known as the scope by which a consistently measures of concepts throughout instrument (Burns& Grove 2010). The pilot research was carried out

to determine the reliability of the research tool. Cronbach's Alpha was performed to measure the reliability of the present study instrument by the use of the Statistical Package for Social Science Program (IBM SPSS) version 25.0. The reliability of instrument was (r = 0.843) .The result of the Cronbach's Alpha reliability of the study tool was statistically high, indicate that the study tool was reliable and has equal measurability.

Statistical Analysis Approach

The SPSS version 20.0 software application was used to conduct statistical analysis (SPSS). The information was evenly spread.

Results

Table1: Overall Quality of Life related to Physical Functioning among Hemoglobinopathies Adolescent

Physical Function	Freq.	%	*M ± SD*
Poor (*M=0-10*)	45	22.5	*16.57±8.62*
Moderate (*M=11-21*)	85	42.5	
Good (*M=22-32*)	70	35.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of physical function related to quality of life was demonstrate that (42.5%) of adolescent are experienced moderate with average of *M ± SD=16.57±8.62*.

Table 2: Overall Quality of Life related to Emotional Functioning among Hemoglobinopathies Adolescent

Emotional Functioning	Freq.	%	*M ± SD*
Poor (*M=0-6*)	63	31.5	*9.49±6.55*
Moderate (*M=7-13*)	73	36.5	
Good (*M=14-20*)	64	32.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of emotional function related to quality of life was demonstrate that (36.5%) of adolescent are experienced moderate with average of *M ± SD=9.49±6.55*.

Table 3: Overall Quality of Life related to Social Functioning among Hemoglobinopathies Adolescent

Social Functioning	Freq.	%	*M ± SD*
Poor (*M=0-6*)	42	21.0	*10.66±5.98*
Moderate (*M=7-13*)	84	42.0	
Good (*M=14-20*)	74	37.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of social functioning related to quality of life was demonstrate that (42%) of adolescent are experienced moderate with average of *M ± SD=10.66±5.98*.

Table 4: Overall Quality of Life related to School Functioning among Hemoglobinopathies Adolescent

School Functioning	Freq.	%	*M ± SD*
Poor (*M=0-6*)	44	22.0	*10.91±5.81*
Moderate (*M=7-13*)	86	43.0	
Good (*M=14-20*)	70	35.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of school functioning related to quality of life was demonstrate that (43%) of adolescent are experienced moderate with average of *M ± SD=10.91±5.81*.

Table 5: Overall Quality of Life related among Hemoglobinopathies Adolescent

	Freq.	%	*M ± SD*
Poor (*M=0-42*)	71	35.5	*47.63±24.38*
Moderate (*M=43-85*)	121	60.5	
Good (*M=86-128*)	8	4.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of quality of life among Hemoglobinopathies adolescents was demonstrate that (60.5%) are experienced moderate quality of life with average of *M ± SD=47.63±24.38*.

Discussion

Table-1: Overall Quality of Life related to Physical Functioning among Hemoglobinopathies Adolescent

The analysis of physical function related to quality of life was demonstrate that (42.5%) of adolescent are experienced moderate with average of M ± SD=16.57±8.62.

This finding in the same line with study that carried out by [8] who reported that Nearly three quarters of the children (71.0%) had ”low” QOL regarding physical functioning.

This finding may be due anemia and complications of iron overload.

Table2-: Overall Quality of Life related to Emotional Functioning among Hemoglobinopathies Adolescent

The analysis of emotional function related to quality of life was demonstrate that (36.5%) of adolescent are experienced moderate with average of M ± SD=9.49±6.55.

This findings in contrast with [9] who carried out a study about determination of quality of life for thalassemia adolescent and illustrated that the overall emotional QoL is (pass). This is probably because thalassemia adolescents feel different from their counterparts and develop a negative feelings about their life and their future, this bad feeling may come from the negative impact of the treatment and the disease itself, and the concern of overall health status or indications of recent deterioration in health.

Table3: Overall Quality of Life related to Social Functioning among Hemoglobinopathies Adolescent.

The analysis of social functioning related to quality of life was demonstrate that (42%) of adolescent are experienced moderate with average of M ± SD=10.66±5.98.
This findings in congruent with [4] who carried out a cross- sectional study , and reported that social domain the quality of life of thalassemic children was significantly lower than the control group.

Table4: Overall Quality of Life related to School Functioning among Hemoglobinopathies Adolescent

The analysis of school functioning related to quality of life was demonstrate that (43%) of adolescent are experienced moderate with average of M ± SD=10.91±5.81.
in similarity with [10] who carried out a study entitled Health related quality of life in Malaysian children with thalassemia who their result indicated that that school functioning is lower in adolescent than in children under 12 years age , this may be attributed to the cumulative effects of the disease and its treatment on the school functioning level.

Table5: Overall Quality of Life related among Hemoglobinopathies Adolescent
The analysis of quality of life among Hemoglobinopathies adolescents was demonstrate that (60.5%) are experienced moderate quality of life with average of M ± SD=47.63±24.38. this finding in the same line with [11] who reported that lower physical, social, emotional, school functioning, and total QoL scores compared with their matched healthy peers

Conclusions

The study concluded that Two fifth of adolescent have moderate physical function, One third of adolescent have moderate emotional function, Two fifth of adolescent have moderate social function, Two fifth of adolescent have moderate school function, and More than half of adolescent have moderate quality of life.

Recommendation

1- Healthcare services can be modified by overcoming underuse, overuse and misuse of services and by controlling disparities in quality. In addition, a more

effective and convenient regimen of iron chelation is essential to improve HRQoL

2- Rehabilitation programs should be held for the adolescents with thalassemia and their caregivers to teach them stress management technique as progressive relaxation techniques, meditation and mindfulness.

3- The health care team members particularly nurses should assume the responsibility to develop adequate social support from a diversity of source as caregivers, relatives, friends and members of self-help group with thalassemia to help adolescents with thalassemia to cope with stressors related to the disease and reduce their stigma felt.

Financial disclosure: There is no financial disclosure.

Conflict of interest: None to declare.

Ethical Clearance: "Official approval was received from the Karbala health directorate and directed to the center for training and development in the Karbala health directorate, the primary approval took to facilitate the research mission and collect the data from hereditary blood disease center in Karbala teaching hospital for children, and after the meeting of research commitment of training and development center the final decision of Approval to conduct research had obtained".

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