Factors Affecting Health Related Quality of Life in Adult with Beta – Thalassemia Major in Nineveh Province

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ABSTRACT

Background: Thalassemia is a common genetic disorder on a global scale. Individuals with transfusion dependent beta-thalassemia experience a range of physical, psychological, and social disorders that ultimately result in a diminished quality of life.

Aim of study: To assess Health related quality of life in adult patients with transfusion dependent beta-thalassemia and to correlate it with various clinical parameters.

Methods: This study is designed as a case-control study involving 80 adult individuals transfusion dependent beta-thalassemia at Ibn Al-Atheer Hospital in Mosul city, along with 80 individuals who are considered healthy controls. To assess the Health-Related Quality of Life score, the researchers utilized the RAND Short Form 36 Questionnaire.

Results: In this study 32 men and 48 women were included. The mean age was 23.8 years ±5.9. After reviewing the patients' quality of life, it was observed that the mean score for physical function was 79.7±15.7%, role of limitations due to physical health problem was 66.9±30.9%, role of limitation due to emotional problem was 73.3±43.3%, energy fatigue was 65.5±29.4%, social function was 77.27±18.6%, pain was 72.2±18.6%, general health was 54.1±22.4%, emotional wellbeing was 58.2±26.8% and overall mean was 67.9±17.8% were significantly lower than healthy controls. The role of limitation due to emotional problem and emotional wellbeing were significantly lower in male compared to female patients with p-value of (0.004, 0.02) respectively.

Conclusion: In this study the Health Related Quality of Life in adults with transfusion dependent beta-thalassemia was reduced in all domains of short form 36 scores.

Keywords: Health, Quality, life and thalassemia.

العوامل المؤثرة على نوعية الحياة المتعلقة بالصحة لدى البالغين المصابين بمرض بيتا – الثلاسيميا الكبرى في محافظة نينوى

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الخلاصة

الخلفية : الثلاسيميا هو اضطراب وراثي شائع على نطاق عالمي. يعاني الأفراد المصابون بمرض بيتا ثلاسيميا الكبرى من مجموعة من الاضطرابات الجسدية والنفسية والاجتماعية التي تؤدي في النهاية إلى انخفاض نوعية الحياة .

الهدف من الدراسة : تقييم نوعية الحياة المتعلقة بالصحة لدى المرضى البالغين المصابين بالثلاسيميا بيتا المعتمدة على نقل الدم وربطها بمختلف المؤشرات السريرية.

الطُرق :تم تصميم هذه الدراسة كدراسة حالة-شواهد تشمل ٨٠ فرداً بالغاً تم تشخيص إصابتهم بمرض بيتا ثلاسيميا الكبرى في مستشفى ابن الأثير في مدينة الموصل، إلى جانب ٨٠ فرداً يعتبرون من الأصحاء. لتقييم درجة جودة الحياة المتعلقة بالصحة للاستبيان RAND 36 باستخدام

النتائج : شملت هذه الدراسة ٣٢ رجلاً و٤٨ امرأة. وكان متوسط العمر ٢٣.٨ سنة ±٥.٩. بعد مراجعة نوعية حياة المرضى، لوحظ أن متوسط درجة الوظيفة الجسدية كان ٧٩.٧±١٥.٧%، وكان دور المحدودية بسبب مشكلة الصحة البدنية ٦٦.٩±٣٠.٩%، وكان دور المحدودية بسبب مشكلة عاطفية ٧٣.٣±٤٣.٣ ٪، كان إرهاق الطاقة ٦٥.٥ ± ٢٩.٤٪، وكانت

الوظيفة الاجتماعية ٧٧.٢٧ ± ١٨.٦٪، وكان الألم ٧٢.٢ ± ١٨.٦٪، وكانت الصحة العامة ٥٤.١ ± ٢٢.٤٪، وكانت الرفاهية العاطفية ٥٨.٢ ± ٢٦.٨٪ وكان المتوسط العام ٦٧.٩ ± ١٧.٨٪ بشكل كبير. أقل من الضوابط الصحية. كان دور القصور الناتج عن المشكلة العاطفية والرفاهية العاطفية أقل بشكل ملحوظ عند الذكور مقارنة بالمرضى الإناث حيث كانت القيمة (p 0.004، 0.02) على التوالي.

الخاتمة : في هذه الدراسة، تم انخفاض نوعية الحياة المتعلقة بالصحة لدى البالغين الذين يعانون من بيتا ثلاسيميا المعتمدة على نقل الدم في جميع المجالات ذات الشكل القصير ٣٦ درجة.

الكلمات المفتاحية : الصحة ، الجودة ، الحياة والثلاسيميا.

INTRODUCTION

Beta thalassemia are heterogeneous autosomal recessive hereditary anemia characterized by reduction or absent in synthesis of the beta globin chains of the hemoglobin (Hb) tetramer. Thalassemia disorders were clustered in the Mediterranean region and the Middle East, sub-Saharan Africa, and East and Southeast Asia, but the emigration have altered the epidemiology of thalassemia to include Northern and Western Europe, North America, and Australia. 200 mutations and more have been reported, the large majority is point mutations in functionally important regions of the beta globin gene [1,2.]

Three main forms of beta thalassemia (major, also referred to as “Cooley’s anemia” and “Mediterranean anemia” which is severe transfusion dependent; Intermedia which is milder; and minor, called “beta thalassemia carrier,” “beta thalassemia trait,” or “heterozygous beta thalassemia.”) [3]. Individuals diagnosed with thalassemia major typically come to medical attention during the period spanning from 6 to 24 months of age. The primary clinical manifestations encompass stunted growth, paleness, yellowing of the skin (jaundice), skin pigmentation changes, weak musculature, knock-knee deformity (genu valgum), enlargement of the liver and spleen (hepatosplenomegaly), leg ulcers, the emergence of masses due to extramedullary hematopoiesis, and alterations in the skeletal structure as a result of bone marrow expansion[3].

Patients with transfusion dependent thalassemia typically presented with low hemoglobin (Hb) levels, decreased mean corpuscular volume, and reduced mean corpuscular Hb. Examination of their peripheral blood smear reveals microcytosis and hypochromia, as well as variations in cell size (anisocytosis), irregularly shaped red blood cells (poikilocytosis) such as spiculated, tear-drop, and elongated cells, and the presence of nucleated red blood cells (erythroblasts). In the bone marrow, there is a significant increase in cellularity due to pronounced erythroid hyperplasia, leading to a reversal of the normal myeloid/erythroid ratio from around 3 or 4 to 0.1 or lower. Qualitative and quantitative analysis of hemoglobin (Hb) composition, conducted through methods like cellulose acetate electrophoresis or high-performance liquid chromatography, may vary depending on the specific type of beta thalassemia. In cases of beta 0-thalassemia, characterized by the absence of beta-globin chain synthesis, HbA is not present, HbF accounts for approximately 95-98%, and HbA2 comprises 2-5% of the hemoglobin content. Mutations in the beta-globin gene, which are commonly observed, can be detected using polymerase chain reaction (PCR)-based techniques [2].

After the age of 10–11 years, affected patients are at risk of developing severe complications related to post transfusion iron overload like: Diabetes mellitus, hypogonadism, hypothyroidism, hypoparathyroidism, adrenal glands insuffency [3,4]. ardiac complications including atrial and ventricular arrythmia, and/or congestive heart failure , myocardiopathy and pericarditis, countng on their compliance with chelation therapy . Hepatitis B and C virus and human immune deficiency virus (HIV) also happened [3].

Deferoxamine (DFO) subcutaneously or intravenously is used as iron chelation, but the difficulty of its uses there are alternative, including orally active iron chelators like Deferasirox (DFP) [5].

Profoundly low levels of calcium leading to tetany require the intravenous administration of calcium (Ca) while closely monitoring the patient's electrocardiogram (ECG). Subsequently, oral vitamin D may be administered. Individuals with diabetes mellitus (DM) will necessitate daily subcutaneous insulin injections. In cases of intense iron chelation therapy employing DFO and DFP, there may be an observed enhancement in glucose intolerance, encompassing improved glucose levels and insulin secretion. This effect is particularly noticeable in patients in the early stages of glucose intolerance. Hypogonadism can be addressed through sex hormone replacement therapy and regular physical exercise is advisable. Oral Ca supplements needs to be used with caution because of the risk of renal. Bone marrow transplantation (BMT) is the only definitive cure currently available for patients with thalassemia. [5].

Health-Related Quality of Life

Health-related quality of life (HRQoL) is defined as (patients' appraisal of their current level of functioning and satisfaction with it compared with what they perceive to be ideal). The World Health Organization (WHO) describes 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. It is a broad ranging concept affected the way by the person's physical health, psychological state, personal beliefs, social relationships and their relationship to salient features of their environment. Apart from the physical issues and limitations in functionality, these patients also grapple with marital difficulties, challenges in education, and the pursuit of a suitable job. These factors not only contribute to a decline in their quality of life but also negatively impact their psychological well-being [6,7].

Health status has specific instruments but the most commonly used instruments is the generic such as the Medical Outcome Study 36- Item Short Form (SF-36), which has important clinical uses as giving information and monitoring outcome in clinical encounters, also used for monitoring population health, estimating the burden of different conditions and with benefits of use across diseases and populations, allows comparison on the same metric across diseases, level of health, and age ranges. On the other hand its use may not be sensitive to change as disease specific measure and may be it does not provide a single summary. The RAND 36 item Short Form Health Survey (SF-36) was used to assess the HRQoL, by self-administration or face-to-face inter-views (for illiterate persons or those with other difficulties).

The SF-36 is suitable for self-administration. It consists of 36 questions that are grouped into eight health status scales: : physical functioning, bodily pain, role limitations due to physical health problems, role limitations due to personal or emotional problems, emotional well-being, energy/fatigue, general health and social functioning. This survey was suitable for self-administration by people 14 years of age and above, or be administered by telephone [8].

Aims of Study

1. To determine the life quality in adults with transfusion dependent beta-thalassemia compared to healthy persons.
2. To correlate various clinical parameters with health related life's quality.

MATERIALS AND METHODS

This study was conducted at Thalassemia center in Ibn Al-Atheer Hospital in Mosul city in Iraq during a period between February 2020 and August 2020. Eighty patients diagnosed as transfusion dependent beta-thalassemia, 18 years and above, registered at the thalassemia center in Nineveh Province were enrolled, .excluding any patients with cognitive impairment and with history of bone marrow transplantation. Eighty healthy, 18 years old or above, age and sex matched to the patients were recruited from Ibn_Al Atheer visitors.

A full history have been taken from all patients regarding the following: age, sex, education, job of the patient, monthly income, age at diagnosis, age of starting first transfusion, date of last transfusion, frequency of transfusion in the last year and any other feature or comorbidity that may be of relevance in the history, then checked for height and weight, the following features were particularly noticed by physical examination: Facial skeletal changes, whether absent, mild, moderate or severe, history of splenectomy, splenomegaly/hepatomegaly and patient information from their records:

Quality of Life

It was performed using the medical outcome study "MOS-SF (version 1.0)" so called RAND 36-item health survey as shown in table 1 [9].This was administered by an interview Arabic -translated version of this questionnaire was used. The outcome of survey was eight health status scales namely: physical functioning, role-limitation due to physical health, role-limitation due to emotional problem, vitality (energy/fatigue), emotion well-being, bodily pain, social functioning and general health.

At first, the numerical values were recorded in each scoring. The ranges of scores from 0 to 100, the higher scores indicating a higher quality of life, better functioning, less limitation or less pain. In second step, items in the same scale were averaged to make the eight scale scores, then determine the median of all elements as shown in table 2 and 3 [9].

Table 1: the RAND 36-item health survey (generic)[9]

Patient Name: _____

1. In general, would you say your health is: (Circle One Number)
   Excellent 1 | Very Good 2 | Good 3 | Fair 4 | Poor 5

2. **Compared to one year ago**, how would you rate your general health right **now** ? (Circle One Number)
   Much better than one year ago 1 | Somewhat better than one year ago 2 | About the same 3 | Somewhat worse now than one year ago 4 | Much worse now than one year ago 5

The following items are about activities you might do during a typical day: Does **your health now limit you** in these activities? If so, how much? (Circle One Number on Each Line)	Yes, Limited A Lot	Yes, Limited A Little	No, Not Limited at All
3. **Vigorous activities**, such as running, lifting heavy objects, participating in strenuous sports	1	2	3
4. **Moderate activities**, such as moving a table pushing a vacuum cleaner, bowling or playing golf	1	2	3
5. Lifting or carrying groceries	1	2	3
6. Climbing **several** flights of stairs	1	2	3
7. Climbing **one** flight of stairs	1	2	3
8. Bending, kneeling or stooping	1	2	3
9. Walking **more than a mile**	1	2	3
10. Walking **several blocks**	1	2	3
11. Walking **one block**	1	2	3
12. Bathing or dressing yourself	1	2	3

During the **past 4 weeks**, have you had any of the following problems with your work or other regular daily activities **as a result of your physical health** ?: (Circle One Number on Each Line)	Yes	No
13. Cut down the amount of time you spend on work or other activities	1	2
14. Accomplish less than you would like	1	2
15. Were limited in the kind of work or other activities	1	2
16. Had difficulty performing the work or other activities (for example, took extra effort)	1	2

During the **past 4 weeks**, have you had any of the following problems with your work or other regular daily activities **as a result of any emotional problems** ?: (depressed, anxious) (Circle One Number on Each Line)	Yes	No
17. Cut down the amount of time you spend on work or other activities	1	2
18. Accomplish less than you would like	1	2
19. Didn't do work or other activities as carefully as usual	1	2

20. During the **past 4 weeks**, to what extent has your physical health or emotional problems interfered with your normal social activities with family, friends, neighbors or groups? (Circle One Number)
    Not at all 1 | Slightly 2 | Moderate 3 | Quite a bit 4 | Good 5

21. How much **bodily** pain have you had during the **past 4 weeks**: (Circle One Number)
    None 1 | Very Mild 2 | Mild 3 | Moderate 4 | Severe 5 | Very Severe 6

22. During the **past 4 weeks**, how much did pain interfere with your normal work (including both work outside the home and housework ? (Circle One Number)
    Not at all 1 | Slightly 2 | Moderately 3 | Quite a bit 4 | Extremely 5

These questions are about how you feel and how things have been with you **during the past 4 weeks**. For each question, please give the one answer that comes closest to the way you have been feeling.

How much of the time during the past 4 weeks . . . (Circle One Number on Each Line)	All of the Time	Most of the Time	A Good Bit of the Time	Some of the Time	A Little of the Time	None of the Time
23. Did you feel full of pep?	1	2	3	4	5	6
24. Have you been a very nervous person?	1	2	3	4	5	6
25. Have you felt so down in the dumps that nothing could cheer you up ?	1	2	3	4	5	6
26. Have you felt calm and peaceful?	1	2	3	4	5	6
27. Do you have a lot of energy?	1	2	3	4	5	6
28. Have you felt downhearted and blue?	1	2	3	4	5	6
29. Did you feel worn out?	1	2	3	4	5	6
30. Have you been a happy person?	1	2	3	4	5	6
31. Did you feel tired?	1	2	3	4	5	6

32. During the **past 4 weeks**, to what extent has your **physical health or emotional problems** interfered with your normal social activities like visiting with family, friends, relatives, etc.? (Circle One Number)
    All of the time 1 | Most of the time 2 | Some of the time 3 | A little of the time 4 | None of the time 5

How TRUE or FALSE is each of the following statements for you? (Circle One Number on Each Line)	Definitely True	Mostly True	Don't Know	Mostly False	Definitely False
33. I seem to get sick a little easier than other people	1	2	3	4	5
34. I am as healthy as anybody I know	1	2	3	4	5
35. I expect my health to get worse	1	2	3	4	5
36. My health is excellent	1	2	3	4	5

Comments: _____

Patient Signature: _____ Date _____

Continue

Table 2: Step 1 scoring the RAND 36-Item Health [9]

Item number	Change original response category	To record value of:
1,2,20,22,34,36	1	100
	2	75
	3	50
	4	25
	5	0
3,4,5,6,7,8,9,10,11,12	1	0
	2	50
	3	100
13,14,15,16,17,18,19	1	0
	2	100
21,23,26,27,30	1	100
	2	80
	3	60
	4	40
	5	20
	6	0
24,25,28,29,31	1	0
	2	20
	3	40
	4	60
	5	80
	6	100
32,33,35	1	0
	2	25
	3	50
	4	75
	5	100

Table 3: Step 2 averaging Items to Form Scales [9]

Scale	Number of item	After recording per table 1 Average the following item
Physical function	10	3,4,5,6,7,8,9,10,11,12
Role of limitation due to physical health	4	13.14.15.16
Role of limitation due emotional problem	3	17.18.19
Energy/ fatigue	4	23,27,29,31
Emotional well being	5	24,25,26,28,30
Social functioning	2	20,32
Pain	2	21,22
General health	5	1,33,34,35,36

Statistical Analysis

The statistical analyses were done using the Statistical software program version 24 (SPSS Inc., Chicago, IL, USA), using mean values ± standard deviations, Independent t- test was used to make comparisons between the scores in different groups of patients relating number of complications, P-values ≤ 0.05 were considered as statistical significant.

Ethical Consideration

This study was approved by an official permission by the ethics committee at Iraqi board medical Specialties commission and by the directorate of health of Mosul. Informed consent was obtained from all recruited patients.

RESULT

The age distributions of majority of patients were between 18-30 years (88.7% of patients) (Figure 1). The mean age of the patients was 23.8 years±5.9), and included 32 males (40%) with mean age (25.04 years) and 48(60%) females with mean age (23.25years). Majority of the patients were literate 65/80 (81.2%), parent consanguinity were 51(63.8%). (Table 4).

Patients' mean age at diagnosis was at 13.4 months. Spleen was palpable in 22/80 (27.5%), while the remaining patients had been splenectomized. Thalassemic faces were noted in 31/80 (38.8%) with moderate or severe changes, while no or mild changes including 49/80 (61.2%). All patients were on chelation therapy, most with the oral deferasirox or Deferoxamine, most patients were good compliant with therapy 53/80 (65.7%)(Table 4)

Overall mean HRQoL of 36 item score in the 80 enrolled adults with transfusion dependent thalassemia was 67.94% ±17.88%, which was significantly lower than that among 80 age and matched controls (Mean96.78±3.69 % (P-value < 0.001). The most affected domain of the eight SF36 domains was the general health 54.13 % and the emotional wellbeing at 58.26%. While the least affected was the physical function at 79.77%. Among these eight domains, all were significantly reduced as compared to their respective scores in the controls (P-value< 0.001) (Table 5).

Gender differences in sex domains of HRQoL were insignificant except for overall mean (p-value 0.02), role of limitation due to emotional problem and emotional wellbeing which were significantly lower among males (P value =0.04) (Table 6).

There were no significant differences in HRQoL overall means and of any of the eight domains among educational background, splenectomy state and compliance for chelation therapy. Significant

differences between patients with severe thalassemic facies in physical function, role of limitation due to physical health, energy fatigue and social function with p-value (0.02,0.005,0.03 and 0,03) respectively (Table 7).

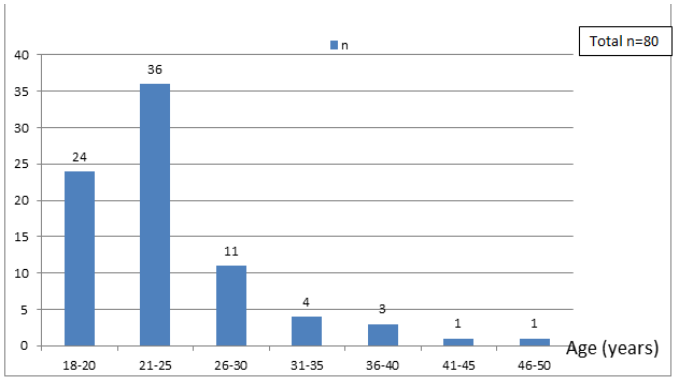

Figure 1: Age distribution of patients with transfusion dependent beta-thalassemia

Table 4: Demographic, social and clinical features of patients with transfusion dependent beta-thalassemia

Category	N	% or mean ± SD
Gender Male Female	 32 48	 40% 60%
Parent consanguinity Yes No	 51 29	 64.7% 36.2%
Education Illiterate Primary school Secondary school High school	 15 45 8 12	 18.8% 56.2% 10% 15%
Age of diagnosis(months)	80	13.40±17.91
History of splenectomy Yes No	 58 22	 72.5% 27.5%
Thalassemic Faces Sever Not sever	 31 49	 38.8% 61.2%
Chelation therapy Regular Irregular	 53 27	 65.7% 34.3%

Table 5: RQoL (SF36) score (%) comparison between with transfusion dependent beta-thalassemia and healthy controls, patients n=80 and control n=80.

HRQoL	Patients (Mean±SD)%	Controls (Mean±SD)%	P- value
Physical function	79.77± 15.74	96.49± 2.88	*<0.001
Role of limitation due to Physical health problem	66.97± 30.98	97.01± 8.16	*<0.001
Role of limitation due to emotional problem	73.35±43.34	98.50± 6.95	*<0.001
Energy fatigue	65.52±29.40	99.55±1.89	*<0.001
Social function	77.57±18.69	99.25±2.98	*<0.001
Pain	72.27±18.52	98.54±5.04	*<0.001
General Health	54.13±22.42	98.88±4.15	*<0.001
Emotional well being	58.26±27.81	88.77±6.97	*<0.001
Overall mean	67.94±17.88	96.78±3.69	*<0.001

*=significant ≤ 0.05

Table 6: Comparison between female and male in patients with transfusion dependent beta-thalassemia concerning parameters of HRQoL (SF36) score (%).

Category	HRQoL (Mean±SD)								
	Physical function	Role of limitation due to Physical health problem	Role of limitation due to emotional problem	Energy fatigue	Social function	Pain	General health	Emotional well being	Mean
Male	78.07± 19.18	57.69± 32.99	62.78± 40.09	59.61± 27.12	72.01± 21.52	68.75± 16.82	51.23± 23.94	48.61± 27.05	61.81± 19.94
Female	80.87± 13.43	72.18± 28.51	80.39± 28.97	68.50± 25.39	80.77± 16.04	73.88± 19.19	55.12± 21.07	67.70± 27.09	71.92± 15.39
P-value	0.48	0.06	*0.04	0.18	0.06	0.27	0.49	*0.02	*0.02

*=significant ≤ 0.05

Table 7: RQoL and its relevance to some clinical and social parameters of patients with transfusion dependent beta-thalassemia.

Category	Physical function	Role of limitation due to Physical health	Role of limitation due to emotional problem	Energy fatigue	Social function	Pain	General health	Emotional well being	Mean
Literate Illiterate P-value	80.72± 13.69 75.41± 23.30 0.29	67.50± 29.71 64.58± 37.62 0.77	75.42± 32.06 75.41± 23.30 0.29	68.36± 24.94 52.50± 30.03 *0.05	78.07± 17.98 74.70± 22.06 0.57	74.0 ± 18.18 64.1 ± 18.62 0.09	55.85± 22.52 46.25± 21.11 0.18	60.50± 26.89 48.00± 30.79 0.16	69.07 ± 18.15 62.18 ± 1527 0.22
Splenectomy Not splenectomy P-value	81.83± 12.93 74.16± 21.09 0.07	67.60±32.06 65.27±28.61 0.78	72.76± 34.32 74.98± 35.36 0.81	65.81± 25.68 64.72±29.02 0.88	76.38± 19.35 80.41± 16.60 0.43	71.7 ± 18.58 73.6 ± 18.92 0.74	56.06± 23.11 48.88± 20.11 0.24	58.69± 26.54 57.11± 30.73 0.83	67.26 ± 19.01 69.40 ± 14.19 0.66
Thalassemic facies Not severe Severe P-value	83.29± 11.59 74.23± 19.68 *0.02	75.30± 25.69 53.84± 34.41 *0.005	79.24± 29.55 64.08± 39.63 0.07	70.85± 21.79 57.11± 30.98 *0.03	82.79± 15.20 69.07± 20.67 *0.03	73.7 ± 19.07 69.9 ± 17.72 0.40	56.09± 20.84 51.03± 24.83 0.37	60.97± 26.10 54.00± 30.34 0.32	71.20 ± 16.49 62.54 ± 18.72 *0.05
Compliance for Chelation Good Not good P-value	81.02± 16.55 77.39± 14.13 0.37	69.03±19.71 63.04± 33.59 0.45	76.87± 34.14 66.62± 34.46 0.24	67.61± 25.11 61.52± 28.85 0.37	77.39± 18.97 77.60± 18.38 0.96	74.8 ± 17.40 67.3 ± 19.94 0.11	53.40± 22.38 55.2± 22.96 0.71	60.63± 28.66 53.73± 26.11 0.33	69.21 ± 18.93 65.20 ± 15.34 0.38

DISCUSSIONS

Recently it is common to measure the HRQoL in adults with hemoglobinopathies. HRQoL in adult with transfusion dependent beta-thalassemia, it forms an important assessment of care and was related to variable extents to disease outcomes, burden and assess the impact of management options. Moreover, the increased survival rates of patients have brought about a significant focus on their quality of life, which has become a central concern for both healthcare policymakers and medical practitioners.(Dominick et al, 2002 [10]. In this study we use SF-36 item to compare with healthy controls and patient own complication as by Panepinto et al, 2012 [11] and Ali , 2019 [12] in north of Iraq.

The majority of patient's age distributions were between 18-30 years (88.7% of patients) (figure1). The mean age of the patients was 23.8±5.9 years, included 32 males (40%) and 48(60%) females. Which was near the mean of Ali H, 2019 22.7±5.07 years in north of Iraq [12], while mean age by Haghpanah S. et al in southern Iran, 2017 [13] was 19.5±4.4 years.

Parent first degree consanguinity was 63.8% similar to Al-Gazali et al, 2006 [14] which found 25-60 %.

Patients' mean age at diagnosis was at 13.4 months while the age of diagnosis in Basrah by Abdul-Zahra et al, 2014 [15] was at 10.92 months and mean age of 8 months in Caocci G,et al, 2012 [7].

Spleen was palpable in (27.5%) of patients in this study while in Ali , 2019 was 14.5% [12]. The remaining patients had been splenectomized were 72.5% while 31.7% of beta thalassemia major patients were splenctomized in Haghpanah et al in Iran [13]. This high percentage of splenectomized patients may be due to complications of hypersplenisim.

Severe thalassemic facies were noted in (38.8%), while Ali , 2019 [12] was 75.9% may be due to that patients in this study are younger age group.

Overall mean and eight domains of HRQoL were significantly reduced as compared to their respective scores in the controls (P-value< 0.001). Among these, the most affected domain was the general health (54.13 %). General health was mostly affected in Ali , 2019 [(12)] and Amani et al, 2015 in Iran [16] .

While energy Fatigue was the most reduced in a study from Saudi Arabian transfusion dependent thalassemia patients by Amoudi , et al, 2014 [17].

The least domain affected was the physical function at 79.77% as in Amani et al 2015(79.8%) [16], while Caocci , et al, 2012 [7] reported a physical function of 68.4% in a cohort of thalassemia patients from Syria.

Gender differences in sex domains of HRQoL were not significant except for role of limitation due to emotional problem, emotional wellbeing and overall mean, which were significantly lower among males (P value 0.04, 0.02 and 0.02) which are similar to Baraz , et al, 2016) [18]. Role of emotional scores were also significantly lower among females. In a study by Hadi , 2009 [6], women had a better quality of life in SF domain whereas studies conducted in Thailand and Malaysia had shown that sex is not effective on the quality of life domains Thavorncharoensap , 2010 [19].

Significant differences between patients with severe thalassemic facies in physical function, role of limitation due to physical health, energy fatigue and social function with p-value (0.02,0.005,0.03 and 0,03) respectively. While study by Ali , 2019 in north of Iraq [11] had no significant differences.

CONCLUSIONS

1. This study was done for the first time in thalassemia center in Nineveh Province by measuring Health Related Quality of Life in adult with thalassemia major compared with healthy control.
2. There was reduction in all domains of short form 36 scores.

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